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Editorial Office

Department of Anatomy,
Faculty of Medicine,
University of Ruhuna,
Sri Lanka

Correspondence regarding editorial matters, articles, reviews and news items regarding the membership should be addressed to the: Assistant Editor, SLAJ, Dr. S.K.Y.I. Kodikara, Department of Anatomy, Faculty of Medicine, University of Ruhuna, Sri Lanka.

E-mail-anatomicalsocetysrilanka@gmail.com

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Editorial

Embryological Descent, Remnants and Implications for Thyroid Surgery

The thyroid gland is the first of the body's endocrine glands to develop, on approximately the 24th day of gestation. The gland originates as a proliferation of endodermal epithelial cells on the median surface of the developing pharyngeal floor. Descent is complete at 7th gestational week. The thyroid is functional by the 8th week of gestation.

In this descent there are several anomalies which give rise to clinical conditions. In addition, there are some remnants of this embryological process which have critical significance in thyroidectomy.

There are four embryological remnants that are recognized and are very important in thyroidectomy

They are:

- Thyroglossal duct remnants (TGDR)
- Pyramidal lobe
- Tubercle of Zuckerkandl
- Thyrothymic remnants /rests

While some standard text books of Anatomy and Embryology mention tract remnants and the pyramidal lobe, very little emphasis is given to the tubercle of Zuckerkandl and the thyrothymic remnants /rests.

A persistent thyroglossal duct courses through the base of the tongue from the foramen caecum. It then passes inferiorly, anterior to, and rarely through, the hyoid body, and often has a diverticulum that hooks below and behind the hyoid, before it courses towards the thyroid gland. TGDRs may present at any age as a cyst, abscess, sinus, fistula or tumour, anywhere along the embryological course of the thyroid gland.

The common anomaly of the thyroglossal duct remnant is a thyroglossal cyst and the close relationship of the embryological tract to the hyoid bone must be borne in mind during surgery and the excision of the central part of the hyoid is essential to minimize the risk of recurrence.

The pyramidal lobe is a narrow lobe of the thyroid gland that arises from the upper border of the isthmus and extends upward, sometimes as far as the hyoid bone; it marks the point of continuity with the thyroglossal duct. The pyramidal lobe may arise from the right or left side of the isthmus. Most studies have shown that the pyramidal lobe exists in 40-60% of the people. Failures to excise the pyramidal lobe will result in a pyramidal lobe recurrence. Rarely the main enlargement of the thyroid is confined to the pyramidal lobe.

The tubercle of Zuckerkandl (TZ) is an embryological remnant of thyroid gland which first described by Austrian anatomist Emil Zuckerkandl in 1902 as the "processus posterior glandulae thyroideae", is as a distinguishable thickening or nodule at posterolateral aspect of the gland. Its close relationship to the recurrent laryngeal nerve (RLN) makes it an important surgical landmark in thyroid surgery. Though it was described in 1902, it was forgotten for more than 100 years, hence called the 'forgotten tubercle'. This is primarily because subtotal thyroidectomy was considered the main technique of thyroid surgery during those 100 years.

With the emergence of the era of total thyroidectomy and more reoperative thyroid surgery being undertaken by endocrine surgeons the significance of the tubercle has been emphasized in the last two decades. The TZ when present is said to "point" to the position of the RLN and the superior parathyroid like an arrowhead. These facts are critical for thyroidectomy. It is common site for goiter recurrence.

There are some embryological remnants of thyroid tissue located in the line of the thyrothymic ligament, referred to as thyrothymic remnants (TTR) or rests. Several studies have shown that rests of thyroid tissue within the thyrothymic area are relatively common (50%). They are often mistaken for lymph nodes or parathyroids. Eighty percent of identified rests are attached to the thyroid proper by a pedicle of thyroid tissue. Twenty percent are entirely separate and most rests are small, with 88% being less than 1 cm in diameter. These need to be identified during thyroidectomy to ensure completeness of surgery which is crucial in preventing recurrence and also very important in thyroid cancer surgery.

These facts regarding embryological descent remnants needs greater emphasis in the teaching of Anatomy and Embryology and must be included in the standard text.

Professor Ranil Fernando
MS, FRCS (Edin) FRCS (Eng) FCPS (Pakistan) FAIS (India) FCS (Sri Lanka) PhD (Col)
Professor of Surgery & Head Department of Surgery, Faculty of Medicine,
University of Kelaniya
Consultant Surgeon, North Colombo Teaching Hospital, Ragama
Past President and Director Examinations College of Surgeons
International Advisor to RCS Edin.

Invited article

Selecting the Best Imaging Investigation for Your Patient with Abdominal Pain

H. Dissanayake[1]

Imaging investigations have come to the forefront in present day patient management. Abdominal pain is a chief complaint in clinical practice. Differential diagnosis of "abdominal pain" includes a broad spectrum of clinical entities that range from benign self-limiting conditions to illnesses associated with high morbidity and mortality. It is often difficult on the basis of history, physical examination and biochemical investigations alone to separate the patients those who require urgent intervention or surgery. Increased availability and use of imaging investigations have dramatically changed the management and outcome of the patients presenting with "abdominal pain".

In United States, of all patients who present to the emergency department with abdominal pain, about one-third never have a diagnosis established, one-third have appendicitis, and one-third have some other definitive pathology. In this "other" category, the most common causes include acute cholecystitis, small bowel obstruction, pancreatitis, renal colic, perforated peptic ulcer, cancer, and diverticulitis (1,2,3,4).

If fever is also present, the need for quick, definitive diagnosis is considerably important. With the history and the clinical examination the problem has to be narrowed down before embarking in to the imaging investigations as to decide the most appropriate. In our setup availability and accessibility will influence the selection of the Imaging Investigation.

ACUTE ABDOMINAL PAIN WITH FEVER

The range of pathology that can produce abdominal pain with fever is very broad. It includes appendicitis, pneumonia, hepatobiliary disease, pancreatitis with or without complications, pyelonephritis, gastrointestinal perforation or inflammation, bowel obstruction or infarction, intra abdominal pus collections, abscesses, pelvic inflammatory disease, some of the tumors etc.

In general, computed tomography (CT) is the most important modality of evaluating patients with abdominal pain with fever. The use of contrast agents greatly increases the spectrum of detectable pathology. However the allergic history and the state of renal function are important factors to consider.

Some authors have found that CT is superior to clinical evaluation for finding the cause of abdominal pain and the use of CT in patients with acute abdominal pain increases the emergency department clinician's level of certainty and reduces hospital admissions by about 24% (4,5,6).

Plain radiographs may provide useful information about bowel gas pattern or free air, but they offer no additional information if CT is to be performed. In gut perforation, while radiographs are sensitive to small volumes of free air, CT is more sensitive to even smaller volumes and can detect additional loculated air or air in the mesenteric root (6,7).

Ultrasonography (US) may be useful in

[1]Faculty of Medical Sciences, University of Sri Jayewardenepura.

selected conditions, like cholecystitis, cholangitis, liver abscess, appendicitis and US may be able to detect abscess or malignancy (such as lymphoma). The diagnostic yield in US is poor in the presence of increased bowel gas or free air. The shortcomings of US are partially offset by its lack of ionizing radiation, particularly in younger patients and in women of child bearing age.

Magnetic resonance imaging (MRI) offers imaging without ionizing radiation and has been shown to provide clinically useful information. The draw backs are the time taken for imaging and lack of free availability in our setup.

ACUTE PANCREATITIS

Imaging is done for various reasons in patients with pancreatitis; to detect the cause, for detection and classification of the severity of the process and to see its complications.

CT is the only imaging study that has consistently shown clinical value in predicting not only the severity but clinical outcomes as well. The CT severity index, as described by Balthazar in conjunction with clinical scoring systems is one basis for patient decision-making. The decision of when to perform CT depends on the overall clinical presentation and should be based on clinical assessment (1,8).

US is often performed in the evaluation of patients with acute pancreatitis since it has a high sensitivity in detecting gallstones. However, patients may not have gallstones but another etiology for their pancreatitis.

The use of MRI in evaluating patients with acute pancreatitis is gaining acceptance. It offers several advantages, especially with heavily T2-weighted sequences for assessing biliary and pancreatic ducts in its entirety, and duct disruption can often be assessed easily, compared to other noninvasive imaging modalities.

Figure 1: Axial CT scan with intravenous contrast show oedematous pancreas, indistinct pancreatic margins and surrounding retroperitoneal fat stranding, suggestive of acute pancreatitis.

Situation where IV contrast can be administered, it can be very helpful in assessing the presence of necrosis (6,9,10).

The disadvantages of MRI are; it is often not readily available in an acute setting and the acquisition times are considerably longer than with CT.

In acute pancreatitis situation following are to be remembered:

In the acute setting, imaging should be performed only if clinically indicated.

- Initial imaging with CT may underestimate the severity of the disease.
- CT with IV contrast gives best overall assessment of the pancreas and complications related to pancreatitis.

- US is primarily used to assess gall stones.
- MRI with IV contrast and MRCP have the potential to be an all-inclusive examination for assessing pancreatitis.

ABDOMINAL PAIN WITH JAUNDICE

The most common causes of obstructive jaundice are neoplasms of the pancreas, ampulla of Vater or biliary tract, choledocholithiasis, pancreatitis, and iatrogenic strictures of the biliary tree.

The methods used in evaluating the jaundiced patient today include US, CT, magnetic resonance cholangio-pancreatography (MRCP), percutaneous transhepatic cholangiography (PTC), and endoscopic retrograde cholangiopancreatography (ERCP). These are effective to varying degrees in assessing both the cause and the site of obstruction; ERCP can also be therapeutic as well.

US is the least invasive and cheapest imaging technique available for evaluating obstructive jaundice. US determine the presence of obstructive jaundice by detecting dilated bile ducts. Inability to see the extrahepatic biliary tree (often because of interposed bowel gas) and the absence of biliary dilation in the presence of obstruction are drawbacks. US is less effective than CT or direct cholangiography (either PTC or ERCP) in determining the site and the cause of obstruction.

CT is more sensitive and specific than US in detecting biliary obstruction. In addition, the ability to determine the site and the cause of obstruction is greater with CT than with US. CT is strongly recommended as the primary modality for evaluating patients with suspected malignant biliary obstruction, both for diagnosis and for staging (3,6).

MRI can demonstrate both site and the cause of biliary obstruction. MR cholangiography has been shown to be useful in depicting the three-dimensional (3D) anatomy of the biliary and pancreatic ducts. For detection of ductal calculi, MRCP is the most sensitive of all noninvasive techniques.

LIKELIHOOD OF BENIGN BILIARY OBSTRUCTION

Patients in this category present with jaundice and acute abdominal pain. There may be a prior history of gallstones documented by sonography or of prior biliary surgery. Sonography is an accurate and the least expensive method for detecting dilated intrahepatic bile ducts and common hepatic duct at the hepatic hilum. Biliary ductal calculi are not detected with the same sensitivity as gallbladder calculi. The sub hepatic common duct may or may not be visible due to overlaying bowel gas. In addition, intrahepatic bile ducts may not be dilated in the early phase of acute obstruction or in patients with partial obstruction. Despite recognized limitations, sonography is recommended as the initial diagnostic test in patients with suspected calculus obstruction of the common duct.

LIKELIHOOD OF MALIGNANT BILIARY OBSTRUCTION

Patients in this category typically present with insidious development of jaundice and associated constitutional symptoms (weight loss, fatigue, etc). Mechanical biliary obstruction can be confirmed by sonography. Malignant obstruction is most commonly due to pancreatic carcinoma but may be secondary to cholangiocarcinoma

of either proximal or distal duct or to periductal nodal compression. A contrast-enhanced CT examination with multiplanar reformation has high sensitivity in lesion detection and in discriminating resectable and unresectable tumour including important information in tumor staging, tumor contiguity or invasion of the superior mesenteric and portal vein, peripancreatic tumor extension, regional adenopathy and hepatic metastases (6,8).

MRI and MRCP are also accurate in tumor detection and staging.

CT is generally more available and more frequently used, while MRI/MRCP reserved for patients with contraindications to CT.

In summary, the diagnostic approach for adults presenting with jaundice depends to a large extent on whether

a) the jaundice is obstructive or non obstructive;
b) the most likely cause, benign or malignant;
c) the patient is an operative candidate, once the diagnosis is made.
d) Lastly, the availability of each modality and the expertise with which it is offered.

RIGHT LOWER QUADRANT PAIN – SUSPECTED APPENDICITIS

Both CT and US may be effective in detecting suspected appendicitis and alternative etiologies of right lower quadrant abdominal pain.

CT is the most accurate study for evaluating patients without a clear clinical diagnosis of acute appendicitis.

The use of CT to evaluate appendicitis has shown to decrease overall cost and has decrease the negative appendectomy rate (3,10).

Several factors are unique in children, including increased radiosensitivity to ionizing radiation and smaller body size and less body fat, favoring initial use of US.

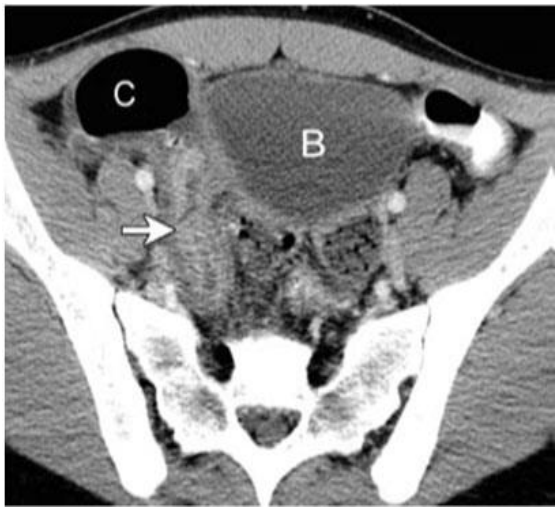

Figure 2: Axial CT images of a patient clinically suspected of having acute appendicitis. CT scan obtained after intravenous and rectal contrast material administration shows appendicitis: a distended appendix with thickened wall (arrow) and surrounding infiltration. B-bladder, C-.cecum. Appendicitis was confirmed at surgery and histopathologic analysis (6).

RIGHT UPPER QUADRANT PAIN

US is the usual initial imaging investigation of choice for patients with right upper quadrant pain, suspected acute cholecystitis (AC), for variety of reasons-availability, lack of ionizing radiation, morphologic evaluation, confirmation of the presence or absence of gallstones, evaluation of intrahepatic and extrahepatic bile ducts, and identification or exclusion of alternative diagnoses. Complications of AC include gangrene, empyema and perforation can also be evaluated with US (1,4). Other clinical conditions that can simulate AC can present with acute right upper quadrant pain are chronic cholecystitis, peptic ulcer disease,

pancreatitis, gastroenteritis, bowel obstruction and many others.

Figure 3: US image of a patient with 4 day history of right upper quadrant pain, nausea, and vomiting shows a thickened gallbladder wall (arrowheads) and an obstructing gallstone (arrow) (6).

If US is negative for AC and an alternative diagnosis is not identified, CT is the next preferred imaging examination for the identification of these disorders. Although not advocated as a primary imaging examination for acute right upper quadrant pain, in equivocal cases on US, CT can confirm or refute the diagnosis of AC and demonstrates complications of AC, including gangrene, gas formation, and perforation (8).

ACUTE ONSET FLANK PAIN – SUSPICIOUS OF RENAL STONE DISEASE

In renal calculus disease, treating physician wants to know the size of the calculus, location and its effect on renal function.

Patients with suspected diagnosis of renal colic have traditionally been evaluated with urinalysis, abdominal radiography of the kidney- ureter - bladder (X-ray KUB), or intravenous urography (IVU). More recently, US, and CT have been used.

Radiography of the abdomen may be sufficient to diagnose urolithiasis in patients with known stone disease. The sensitivity of the X-ray KUB for diagnosing urolithiasis in other patients is poor.

Since the introduction of the use of helical (spiral) Non Contrast CT (NCCT) it is confirmed to be the study with the highest sensitivity and specificity for urolithiasis. Virtually all stones are radio-opaque, and stone size can be measured accurately in cross-section, aiding in predicting outcome. Stone location, accurately depicted by NCCT, has also been associated with spontaneous stone passage rates, with the more proximal stones having a higher need for intervention (11,12).

Figure 4: Ureteric stone in a patient who presented with acute left flank pain. Axial unenhanced CT scan shows a 4mm stone in the left distal ureter (arrow) (11).

Secondary signs such as ureteral dilatation and perinephric stranding allow CT to make the diagnosis of recent passage stone. NCCT is rapid and safer than IVU since it uses no contrast media.

If there is uncertainty about whether a calcific density represents a ureteral calculus or a phlebolith at NCCT, intravenous contrast can be administered and excretory phase images obtained for definitive diagnosis.

Figure 5: Axial CT scan without intravenous contrast shows a calculus in the left mid ureter (arrow).

US is a safe, noninvasive imaging modality that can be used to study the urinary tract effectively. The diagnosis of obstructive urinary tract calculi depends on identification of the offending calculus and concomitant pelvicaliectasis and ureterectasis extending to the obstructing site. Because it may take many hours for hydronephrosis and hydroureter to develop, US will miss some of the acute obstructions caused by a ureteral stone in patients who are not specifically hydrated for the study.

US has been found to be very sensitive for signs of obstruction (hydronephrosis, ureteral dilatation). However, the sensitivity of US as compared to NCCT for detecting renal calculi is quite low, and is especially poor for small stones (6).

US can also evaluate the presence and type of ureteral jet.

LEFT LOWER QUADRANT PAIN

Appropriate imaging for patients with suspected diverticulitis (i.e., left lower quadrant pain) should address two major clinical questions:

1) what are the differential diagnostic possibilities in this clinical situation
2) what information is necessary to make a definitive management decision.

Some patients with acute diverticulitis may not require any imaging, notably those with typical symptoms of diverticulitis (e.g., left lower quadrant pain and tenderness, fever) or those who are diagnosed history of diverticulitis who present with clinical symptoms of recurrent disease. Some patients with diverticulitis require surgery because of associated abscesses, fistulas, obstruction, or perforation. As a result, there has been a trend toward greater use of radiologic imaging tests to confirm the diagnosis of diverticulitis, evaluate the extent of disease, and detect complications before treatment (6,10).

CT is now widely advocated as the imaging test of choice for evaluating patients with suspected sigmoid diverticulitis because of its high sensitivity and specificity and its ability to diagnose other causes of left lower quadrant pain that mimic diverticulitis (eg, genitourinary and gynecologic abnormalities) that have a similar clinical presentation.

CT also has a major role in determining disease extent; this assessment is rarely possible with contrast enema.

A variety of contrast media have been used for CT to optimize the sensitivity and specificity of the examination, including oral, rectal and intravenous contrast agents.

Transabdominal US has limited use.

Transvaginal US is particularly of value when left lower quadrant pain occurring in women of childbearing age. In this setting, gynecologic problems such as ectopic pregnancy and pelvic inflammatory disease are also important diagnostic considerations. US is therefore an excellent choice for the initial imaging of these patients, because it is more sensitive in detecting gynecologic abnormalities that cause left lower quadrant pain (6).

SUSPECTED SMALL BOWEL OBSTRUCTION

There is no single generally accepted approach for evaluating patients with suspected small-bowel obstruction (SBO).

Radiography has been the traditional starting point for imaging evaluation of suspected SBO.

In such a setting, gastrointestinal contrast studies (small-bowel follow-through [SBFT], enteroclysis, and barium enema) are controversial due to problems like intravascular volume depletion, electrolyte imbalance, barium impaction etc (13).

CT is useful in suspected high-grade SBO in identifying the cause of obstruction. Patients with suspected high-grade obstruction may not require any oral contrast medium since the fluid in the bowel provides adequate contrast. Low-grade obstruction is a relative "blind spot" for standard CT (7).

CT is very useful for detecting complications of bowel obstruction such as ischemia and strangulation. CT has been useful in effectively triaging patients into operative versus non operative treatment groups. In the pediatric age group, US have proven benefit in evaluating intussusceptions, midgut volvulus and other causes of SBO.

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CORRESPONDENCE

Dr. Harsha Dissanayake
Department of Anatomy,
Faculty of Medical Sciences,
University of Sri Jayewardenepura.

E mail-harshadis@yahoo.com

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Post Mortem Artefacts that Mimic Deliberate Physical Violence and Ante Mortem Lesions - Revisited

J. Warushahennadi[1], P. R. Ruwanpura[2]

INTRODUCTION

Post mortem artefact is any change caused or a feature introduced in a body after death that is likely to lead to misinterpretation of medico legally significant ante mortem findings (1). Reddy states all events that happen after death which may simulate ante mortem injuries can be grouped together under the heading of post mortem artifacts (2). These artifacts are not related to actual patho-physiological mechanism of death and often lead to misinterpretation of medico legal important ante mortem findings which may be responsible for the cause, manner and the mode of death. Artefacts are commonly observed at autopsies which present in a form of an injury, post mortem change, pathological lesion, histological finding, bio-chemical finding, histo-chemical finding or as an error in photography, radiography, ultrasound and other ancillary investigations.

It is important to differentiate ante mortem injuries from the postmortem artefacts, keeping in mind the different changes which take place after the death. Lack of ability to differentiate ante mortem injuries from the post mortem artefacts may lead to incorrect assessment of time since death, wrong cause of death, miscarriage of justice, and waste of time and resources due to misleading findings (3).

Objective of this paper is to highlight certain types of artefacts authors during over 5000 autopsy examinations and discuss medico-legal issues of their interpretation and differential diagnosis with reference to available literature sources.

TYPES OF ARTEFACTS AND THEIR ORIGINATION

The postmortem artefacts are highly versatile and classified according to their origin. Iatrogenic artefacts are ante-mortem or peri mortem in origin and caused by various medical and surgical manipulations. Some examples are haemorrhage around the injection marks appear as contusions or snake bites, abrasions on the skin in a linear pattern after removal of the medical tape, discoid bruises on upper arms due to neurological stimulation which may be misinterpreted as injuries due to physical abuse, incisions for intercostal tubes imitating stab wounds etc.

Resuscitation artefacts accounts for reckoning portion of lesions of peri-mortem origin usually caused by terminal therapeutic procedures of the medical staff. Some of the common resuscitation artifacts are chest contusions and rib fractures due to cardio pulmonary resuscitation, contusion of lips due to tracheal intubation, circumscribe patterned burn marks on the chest due to defibrillation, redness and edema of larynx due to intubation, haemorrhage into the myocardium due to intra-cardiac adrenalin injection. The peri mortem origin of these lesions leads to formation of vital reaction around the lesions which make it extremely difficult to distinguish from ante

[1]Department of Forensic Medicine, Faculty of Medicine, University of Ruhuna.
[2]JMO office, Teaching Hospital, Karapitiya.

mortem mechanical injuries. Effects of heat causes multitude of artifacts that may interfere with recognition of essentially ante-mortem lesions responsible for volitional activities and death. A pugilistic attitude due to heat stiffening of skeletal muscles being long misinterpreted as appealing volition of a murder victim. Other heat related post mortem appearance include blister formation on the skin due to burning after death, heat lacerations on skin with underlying muscles, haematoma in the extradural space, fractures of the skull and long bones, presence of coagulated blood in respiratory passages, soot particles in the oral cavity and larynx etc.

Post mortem changes are responsible for vast spectrum of artifacts, commencing from blood stained discharges at nostrils, presence of coffee-brown material in the stomach due to conversion of terminal erosions by gastric hydrochloric acid, milky spots on the epicardium due to early autolytic changes, Tache-Noire due to drying of sclera, shedding and blistering of epidermis due to decomposition which may be mistaken for thermal burns, prolapsed uterus due to the pressure following collection of gases produced in the abdomen mimicking criminal abortion, autolytic rupture of gastric mucosa (gastromalasia) may be mistaken for penetrating injury, protrusion of tongue misinterpreted as a sign of pressure on the neck.

A submersion tends to the appearance of different changes on dead bodies leading to conflicting opinions even among well experienced pathologists. The distinction between the facts and the artifacts are vital in determining whether the victim was alive at the time of entering into the fluid media. The artifacts mainly caused by submersion are rather during the process of death due to drowning. The commonly observed immersion artifacts are cutis anserine or 'Goose-Flesh' appearance of the skin due to agonal contraction of erector pilae muscle, `washer woman's hand due to wrinkling of the skin, unusual distribution of hypostasis on face and hands, sunburns of exposed surfaces of floating bodies, discharges at natural orifices, presence of sand in upper respiratory tract and at times positive Diatom tests.

Artefacts caused by animal activities are essentially post mortem and the knowledge about the biodiversity and behavioural patterns of the fauna in the particular region is very essential in recognizing and interpretation of these lesions. These lesions appear as skin abrasions, contusions, lacerations, incised wounds, stabs, firearm injuries, fractures, disfiguring and mutilation of dead bodies. Ants are observed on the body shortly after the death. The feeding actions of ants cause irregular scalloped areas of superficial skin loss or small punctate lesions on uncovered areas of the body. Usually these injuries do not associated with bleeding but sometimes a considerable bleeding can take place where removal of the skin occurs in congested parts of the body. Rats, dogs and cats destroy exposed soft tissues which have nibbled margins. The paws of large animals like crocodiles cause multiple parallel tears on soft tissues mimicking cuts. The sharp, long canines may cause injuries mimicking penetrating injuries on the soft tissues and the bones.

Artefacts due to post mortem handling of the body by undertakers and mortuary attendants consist of facial abrasions due to shaving, fracture of laryngeal cartilages

during evisceration, multiple punctures, cuts and haemorhages due to embalming procedures, ligature marks due to tying and pulling of the body etc. Injuries like abrasions and lacerations on the skin can be produced by the manipulations of the body during handling, transportation and storage.

Artefacts are not limited to gross physical lesions. They can be present in a form of histological finding in the ancillary investigations. Some of the common histo-pathological artifacts are autolysis of cells resembling necrosis, blood in the renal tubules mimicking acute tubular necrosis, leucocytic reaction and pavementing of white blood cells even after cessation of circulation, presence of colonies of microorganisms in internal organs mimicking infections etc. A migration of foamy macrophages into the alveolar spaces is believed to be a post mortem phenomenon.

Biochemical artifacts include post mortem redistribution and diffusion of toxic substances, high albumin content in the tissues from hypostatic areas, low sugar levels in the vitreous humor, high K^+ in post mortem blood, post-mortem production of alcohol and cyanide in the blood vessels etc.

DISCUSSION

In the context of Forensic Pathology an artifact can be defined as a spurious postmortem presentation which stimulates a finding which would be significant in the course of ante-mortem events (4). These artefacts may be introduced before death, at the time of death or after the death and labeled as therapeutic artifacts, agonal artifacts and post-mortem artefacts (5). Artefacts are not illusions. It is always an objective finding observed on a body due to other than path-physiological evolution of a disease process and death. The same fact which is considered to be an artifact in relation to one particular medico legal issue could be a valuable finding in relation to another medico-legal aspect of criminal investigation. For an example; the present of a patchy abrasion due to postmortem sunburns on back aspect of the body is an artifact in relation to violent mode of death and blunt force injuries, but at the same time the fact indicates that the body was left face down position on an open area during the daytime for several hours.

In contrast to livor mortis, rigor mortis, autolysis, and putrefaction that are frequently observed in daily practice, more uncommon postmortem changes that do occur occasionally and under specific intra-individual or environmental conditions may be interpreted falsely by the inexperienced pathologists. For an example the hypostasis in the muscles located in the lateral submalleolar region and the thener eminence may mimic antemortem bruises (6). Tachiea noire may be misinterpreted as subconjunctival haemorrhege. In a retrospective study of 230 autopsies requested by the coroner's office the postmortem artifacts were mistaken for traumatic lesions in 18 reviewed cases (7). Therefore the ignorance and misinterpretation of such post mortem artefacts leads to arrive wrong cause of death, wrong manner of death, under suspicion of criminal offence, a halt in the investigation of criminal death, unnecessary spending of time and effort as a result of misleading findings or even and miscarriage of justice (8).

Resuscitation and emergency care incorporated vigorous medical intervention is another aspect in Forensic Pathology

which cause practical difficulty to differentiate ante-mortem and post-mortem injuries. It is because in most instances the resuscitation occurs around the time of death with some degree of maintenance of circulation movements which leads to extravasation, infiltration and coagulation of blood and developing vital reaction mimicking ante-mortem injuries (9). An autopsy based survey revealed that resuscitation or iatrogenic artefacts were present in 12.84% cases, with artefactual rib fractures were found in 15% cases (10). The displaced rib fractures may inflict lung contusion, anterior mediastinal and myocardial contusions and even penetrating injuries on the lungs and heart (11). Defibrillator paddles produce hyperemic oval or ring shape burns clustered over the precordium. The Gray has described a rare artefactual injury from application of defibrillator simulating human bite (12). The other rare injuries caused by the closed chest compression are liver lacerations. The pathologist should be careful in interpretation of injuries in areas of the body where resuscitation has taken place.

Heat related artefacts are well documented in the forensic literature. The key medico-legal issue of investigation of a burnt body is to determine whether the victim was alive at the time of onset of fire, and distinguish heat related artefacts from ante mortem injuries. The most important signs of vitality in burned bodies are soot deposits in the distal respiratory tract, esophagus and the stomach as well as elevated CO-Hb values in the blood. Analysis of 88 burnt victims revealed that about 3% of the cases the question whether there was an ante-mortem heat exposure could not be answered (13). Apart from burns the external findings may include leathery consolidation and tightening of the skin and the presence of partly long splits and so-called pugilistic attitude[10] which is due to shortening and heat contractions of relatively stronger flexor muscles and does not demonstrate volition by the decedent to fight back the fire immediately before death (14). When superficial injuries are distorted with severe degree of burning, it can be extremely difficult, if not impossible to distinguish facts from artefacts. But even in charred bodies internal injuries are well preserved which helps to assume the type of external injuries (15). Heat extradural hematoma and heat fractures with chipping fragments are usually observed in skulls subjected to extensive post-mortem burns. Comminuted skull fractures with burning of fractured margins are indicative of secondary burns of already fragmented head. Burnt skull bones has a grey white colour showing a fine superficial network of heat fractures on its cortical surface which may crumble even on handling (16). The heat haematoma can resemble an extra dural haemorrhege of ante-mortem origin. The usual site at the vertex or occipitut and absence of fractures crossing the grooves of the middle meningeal arteries and the chocolate brown colour honey comb appearance helps to distinguish the heat haematoma from the extra dural haemorrhage (16,17). The boiled and shrunken brain is a common finding of burns. The brain may appear swallon with widening and flattening of the gyri and obliterating the sulci due to contraction of the coagulating dura against the brain (15). The shrunken dura may split with herniation of the brain matter into the extra dural space (17). Immersion artefacts present on bodies are irrespective of whether death was due to

Immersion artefacts present on bodies are irrespective of whether death was due to drowning or the person was dead on entering the water. Therefore, immersion artifacts do not contribute to the proof of death by drowning (18). Human body, once retained under the fluid media is subjected to many physical effects. These changes are varying in line with quality and temperature of the water stream. General putrefactive changes are delayed in the water which may lead to errant estimation of time since death (15,19). Many investigators argue that the goose flesh appearance of skin is not a sign of ante mortem drowning but due to post mortem rigor mortis of erector-pile muscle of hair bulb. The presence of sand in upper trachea of a drowning victim is another point of non-consensus. An appearance of washer woman's hands is agreeable post mortem phenomenon due to prolonged immersion probably caused by post mortem vaso constriction (20). The important vital features of ante-mortem injuries could be washed off by effect of water, promoting post mortem outlook.

There is no unanimous opinion about the presence of diatoms in bone marrow as a confirmatory evidence of drowning, and the positive diatom test needs to be carefully evaluated on case by case basis (21,22). Furthermore, diatoms can be present when people dye due to other non-drowning causes of death (23). The macroscopical and microscopical signs in the fresh drowned victim are non-specific and moreover putrefaction will varnish these autopsy findings quite rapidly (24). Finally what influences the decision making in favour or against the death due to drowning is neither autopsy nor diatom test alone. It is a combination of thorough autopsy in light with the right information by the police together with all other ancillary investigations (25). Dead bodies are attracted by animal population soon after death, known as taphonomic processes of corpses. The impact of animal activities on corpses may result from mere post mortem skin wounds to complete distortion of the body. The diagnosis of postmortem lesions inflicted by animals generally presents no problems; however, it can be difficult when the body is in an advanced state of putrefaction (26). Ants attack the bodies in early post-mortem interval causing abrasion like lesion. Drying of post mortem ant bites can give the impression of bruises or chemical burns (27,28). Sometimes there may be significant post mortem bleeding from the ant bite giving an impression of ante-mortem trauma. As ants attack the uncovered areas of the body, ant bites can frequently give rise to suspicion especially if located on the neck mimicking pressure on the neck. They are also occasionally misinterpreted as patterned abrasion due to the imprinted effect of a blunt or offending object (27). Decomposition and action of necropagous species like maggots of flies result in penetrating defects resembling firearm wounds (5). Otherwise, maggots could alter existing penetrating wounds destroying key features of the original lesion. Rodent attacks, especially by mice and rats inflict superficial patchy skin loss with ragged margins with no bone damage. We have observed patterned cuts like lesions are produced in attack of corpses by large monitors. These injuries are caused by pointed nail tips of the animal.

PRINCIPALS OF DIFFERENTIAL DIAGNOSIS

According to a Canadian autopsy survey, artifact misinterpretation occurred in

7.83% cases of all requested autopsies and 35.29% in decomposed cases (29). A study conducted in Pakistan reveled that out of 780 autopsies, 229 showed postmortem artifacts which included decomposition in 78, animal and insects producing changes in 45, injuries due to rough handling in 40, breaking of rigor mortis and shifting of postmortem lividity during transportation in 37, iatrogenic fracture of skull during opening in 15, fractured ribs during resuscitation after death in 8 and exhumation producing fractures of bones in 6 bodies. In fact some degree of artefactual changes is present in almost all dead bodies.

Most of the post mortem artifacts are avoidable, or at least can be minimized with the availabilities of well-equipped mortuary room with enough storage provisions, with chemical and histo-pathological laboratory facilities. Trained mortuary attendants and the law enforcement who are engaged in the medico legal work will avoid infliction of artefacts. Photograph of dead bodies at recovery, on arrival to the mortuary will helpful to understand and interpret the origin of different artifacts.

Systematic post mortem examination which includes detailed history from all possible sources, overall assessment of the statue of the body, scene visit, detailed study into the examination and necessary ancillary investigation findings enables the forensic pathologist to overcome the difficulties of the identification and interpretation of the artefacts and conclude the correct decision.

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CORRESPONDENCE
Dr. Janaki Warushahennadi
Department of Forensic Medicine,
University of Ruhuna, Sri Lanka.

E mail- janakiwh@gmail.com

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Knee Joint Alignment in the Indigenous People of Sri Lanka

H. A. Amaratunga[1], S. B. Adikari[1], M. R. Ileperuma[2], G. H. M. D. N. Chandrasekara[2], M. S. Chandrasekara[1], H. J. Suraweera[2]

ABSTRACT

Objective: The objective of this study was to measure the knee joint alignment of the indigenous people of Dambana using the goniometer and compare with a control group of Sinhalese individuals.

Materials and Methods: Hundred adult volunteers, from the population of "veddas" living in Dambana and 100 adult Sinhalese volunteers above the age of 25 were recruited for the study. Weight and height of the subjects were measured and the BMI calculated using the Quetelets index. The knee joint alignment was measured using the goniometer according to the method described by Kraus *et al*. (2005).

Results: Indigenous people included 46 females and 54 males with an age range of 26 to 75 years. The control sample consisted of 50 females and 50 males with an age range of 25 to 65 years. The BMI of indigenous people was 21.19 and that of the control population was 22.5. Indigenous people had a mean knee alignment angle of 182.4° and the control sample had a mean of 180.9°. None of the over 50 years population had clinical evidence of osteoarthritis (OA) in the indigenous group while in the control group, all females above 50 years had mild knee pain and 2 had clinically detectable OA.

Conclusions: Knee joint alignment in the indigenous population is closer to the normal range than in the control sample. Clinically detectable OA appears to be absent among the indigenous people.

Key words: knee joint alignment, knee osteoarthritis, indigenous people

INTRODUCTION

The alignment of the knee joint has generated much interest in the recent past due to its link with osteoarthritis (OA) (1). It is described as the angle formed by the femoral shaft and the tibial shaft at the knee. The exact factors that influence the alignment of the knee have not been elucidated to date. However, it is postulated that genetic factors, body mass index (BMI), laxity of the joint and the life style may play a role (2). A full length anteroposterior weight bearing limb radiograph is considered as the gold standard for measuring the alignment of the knee (3). Goniometry is an alternative and simpler method to physically measure the alignment and matches well with the gold standard measurement (4,5). Being a physical examination method it does not expose the patient to radiation and can be easily incorporated into the knee examination.

The indigenous population of Sri Lanka, are commonly known as the "veddas" live in a relatively closed community, with many inter marriages. They have a way of life which causes them to squat, climb trees, carry heavy weights and walk long distances. This type of life style leads to strain on the knee joints. Strain on the knee

[1]Department of Anatomy, University of Peradeniya, Sri Lanka.
[2]Orthopedic Unit, Teaching Hospital Peradeniya, Sri Lanka.

joints has been associated with development and progression of OA (6,7). Due to this life style and the high fiber content of their food obesity is relatively uncommon in their population.

Therefore, the objective of this study was to measure the knee joint alignment of the indigenous people and compare with a group of urban Sinhalese individuals and to detect OA of the knee joint by clinical examination.

MATERIALS AND METHODS

Hundred adult volunteers above the age of 25 years, from the population of indigenous people living in Dambana were included in the study. The other group consisted of 100 adult Sinhalese volunteers from the University population above the age of 25 years.

In both groups weight and height were measured in kilograms and meters using standard measuring scales and the BMI was calculated using the Quetelets index (8). The knee joint alignment was measured using the goniometer according to the method described by Kraus *et al.* (5). Each subject was carefully questioned with regard to the symptoms of knee joint OA. Both knees were examined for the presence of swelling, deformity, tenderness and crepitus. A diagnosis of OA was made using the clinical criteria laid down by the American College of Rheumatology. The criteria being, knee pain with morning stiffness lasting 30 minutes or less and crepitus on motion (9).

RESULTS

The sample from the indigenous people consisted of 100 individuals who included 46 females and 54 males with an age range of 26 to 75 years. The control sample consisted of 50 females and 50 males with an age range of 25 to 63 years. Therefore 200 knees of the indigenous people and 200 knees of the control sample were measured. The mean BMI of the indigenous people was 21.19 (±3.95) and that of the control group was 23 (±3.2). The observed difference in the mean BMI was statistically significant. Indigenous people had a mean knee alignment angle of 182.4° (±3.45) and the Sinhalese sample had a mean of 180.9° (± 4.7) (Figure 1). However the difference observed in the means was not statistically significant. In the indigenous people highest number of knees were in the 181° to 185° range while in the control the highest number was seen in the range of 176 to180°. The variance was lower than the Sinhalese in the indigenous population (Figure 2). None of the over 50 years population had clinical evidence of OA in the indigenous group while in the Sinhalese group, all females above 50 years (12) had mild knee pain and 2 had clinically detectable OA.

DISCUSSION

In studies done in the West the normal range for knee joint alignment angle as measured by the goniometer is considered as 182 to 184° (5). It was observed that the knee angle range in the indigenous people was very close to this. However in the control population the range was several degrees less. The active life style of the indigenous people maybe resulted in an increased strength in the ligaments and muscle in the lower limbs, thereby maintaining the angle within the normal range. The lower BMI of the indigenous people is by reducing the load on the knees maybe maintaining the alignment closer to the normal range. The fact that the knee alignment angle is concentrated to a narrower range with low variance in the

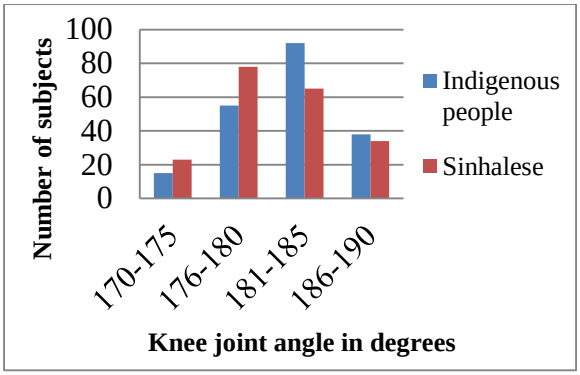

Figure 1: Distribution of the knee joint alignment in the indigenous and the Sinhalese population

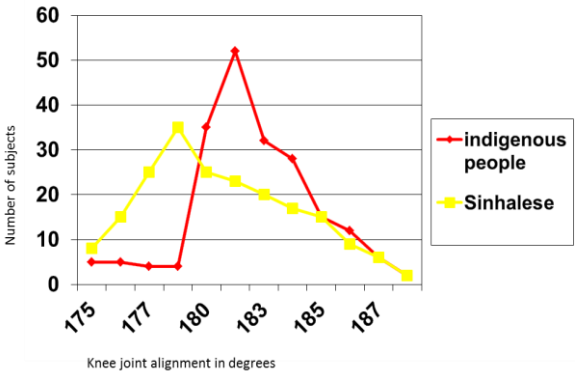

Figure 2: Variance in the knee joint alignment in the indigenous people and the Sinhalese group

indigenous people could be due to the closeness of the community and inter marriages leading to a restricted gene pool (10). Knee joint alignment in the indigenous population is closer to the normal range than in the control sample. However there is no significant difference between the mean knee alignment in the two groups. Clinically detectable OA appears to be absent among the indigenous people.

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CORRESPONDENCE

Dr. H. A. Amaratunga
Faculty of Medicine,
University of Peradeniya, Sri Lanka.

Email: himanipeiris@yahoo.com

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Assessment of Renal Length and Serum Creatinine Values in Chronic Kidney Disease - A Hospital Based Study

S. K. Y. I. Kodikara[1], D. T. K. Gamage[2], L. Y. D. Dulmini[3], B. G. Nanayakkara[1], I. Ilayperuma[1]

ABSTRACT

Introduction: Chronic kidney disease (CKD) is a global & local health problem. Serum creatinine (Scr) is an indicator of renal function. Ultrasound is a freely available safe, reliable imaging modality, commonly used to assess kidneys in CKD. Association of renal length and Scr in CKD is evaluated in this study, as such data is lacking for Sri Lankan population.

Methodology: A descriptive cross sectional observational study was conducted at the Radiology Unit of the Base Hospital Tissamaharama from March to August 2017 (as a preliminary study of a large ongoing study), using 41 patients with normal Scr (control group) and 74 patients diagnosed with CKD. The Scr values done within the last three months were evaluated. Absolute renal length was measured ultrasonically by an experienced Radiologist, who was blind to the patient's serum creatinine value, with Mind-ray ultra sound scanner.

Results: Study population (n=115) consists of control group (n=41) and CKD patients (n=74) where 40% were females and 60% of were males. Mean age was 58.3 years & mean renal length in control group was 9.6cm and CKD group was 9.0 cm. A significant difference observed in renal length in control and CKD groups (<0.001) & a weak negative correlation between renal length and Scr value in CKD group (r=0.328).

Conclusion: Absolute renal length in CKD was significantly lower compared to the control group. Weak negative correlation was established between absolute renal length and the Scr value, thus with the renal function. Further studies are needed to evaluate the relationship between relative renal length and serum creatinine values and to assess above parameters according to etiological categories of CKD.

Key words: chronic renal failure, serum creatinine, ultrasound scan, renal length.

INTRODUCTION

Chronic kidney disease (CKD) is defined as reduced renal function for three or more months (1). CKD is a global health problem with a prevalence of 13·4% (11·7–15·1%) (2). It is becoming a major health problem in Sri Lanka over last two decades and a significant burden to the health sector.

Ultrasound (USS) is a safe imaging modality freely available in developing countries like Sri Lanka, which does not involve with ionizing radiation. Sonographical measurement of renal length is an easy, reproducible and non invasive method. According to the available literature, no significant inter and intra observer renal measurement variability with USS, thus is a reliable imaging modality for assessment of renal length (3). Other available imaging

[1]Department of Anatomy, Faculty of Medicine, University of Ruhuna,
[2]Consultant Radiologist, Base hospital Tissamaharama,
[3]Medical officer, Base hospital Tissamaharama.

modalities like computer tomography, provide promising answer for renal measurements, with the cost of ionizing radiation exposure. For said reasons, renal USS is of value in the assessment of renal size in CKD patients (3,4,5,6). Population specific analysis of renal size is important, as renal size varies with ethnicity (7).

Serum creatinine (Scr) value is an indicator of renal function and cut off values are established to evaluate renal function and to diagnose renal impairment (8).

The present study was aimed at evaluating the association between absolute renal length and serum creatinine values in CKD and to compare with that of healthy population.

MATERIALS AND METHOD

Present descriptive cross sectional observational study was approved by the ethical review committee of the Faculty of Medicine, University of Ruhuna and was conducted at the Radiology Unit of the Base Hospital Tissamaharamaya from March to August 2017 as a preliminary study of a large ongoing study. Present study included 41 patients with normal serum creatinine as control group and 74 patients diagnosed with CKD. Study subjects were above 18 years of age. The serum creatinine reports that were done within the last three months, at the day lab of the Base Hospital Tissamaharamaya were included in this study. Following patients were excluded from the study; history of previous renal surgeries, acute renal insufficiency or on renal replacement therapy (haemodialysis, peritoneal dialysis, renal transplantation), fatty liver or any other liver disease and unwilling patients. All USS were performed by an experienced Radiologist, who was blind to patients serum creatinine value, with Mind-ray ultra sound scanner, using standard grey scale imaging with 3.5 MHz curved array transducer. Grey scale amplification and the time gain compensation curve were adjusted to acquire the best quality images. Single focus point was adjusted at the level of the kidney, while routinely using tissue harmonic imaging. All patients were well hydrated and are with full bladder at the time of imaging. Maximum pole to pole length (Figure 1) of each kidney was measured and recorded as the renal length (absolute).

Descriptive analysis of data was done with mean, standard deviation, minimum and maximum values. Student's t-test was used to evaluate the differences between two groups. Correlation between the variables was assessed with Pearson correlation co-efficient.

RESULTS

Study population (n=115) consisted of a control group (n=41) and diagnosed CKD patients (n=74). Forty six (40%) of them were females and 69 (60%) were males. Average age was 58.3 years (range 20 – 89 years). Average renal length in the control and CKD groups are shown in Table 1. The results of Student t-test comparing the renal lengths of control and CKD groups are presented in Table 2. Serum creatinine values of respective groups are shown in Table 3. Figure 2 demonstrates the relationship between the renal length and serum creatinine values. Pearson correlation co-efficient comparing serum creatinine values with renal length in CKD group shows weak negative correlation, r value was -0.328.

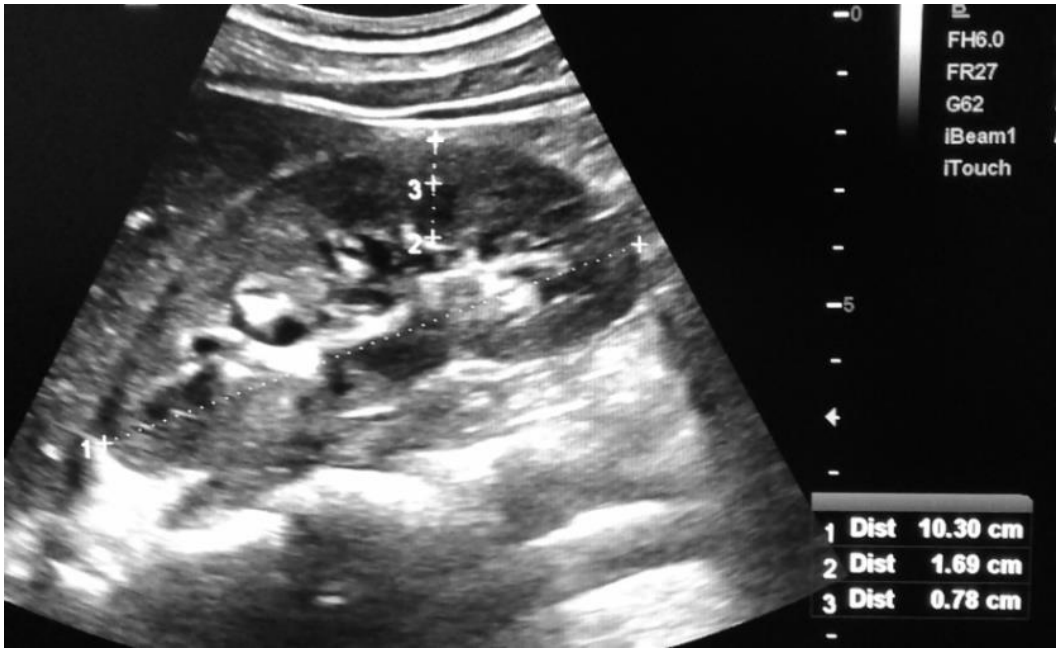

Figure 1: Renal length measurement using Ultrasound
1-Renal length, 2-Medullary thickness, 3-Cortical thickness

Table 1. Average renal length in the control and CKD groups

Renal length (cm)	Control group	CKD
Right renal length. mean ±SD	9.98 ± 0.87	8.97 ± 1.3
Right renal length: range	6.5 – 11.1	6.8 – 11.4
Left renal length: mean ±SD	9.94 ± 0.75	9.0 ±1.3
Left renal length: range	8 – 11.2	8.2 – 11.2

Table 2. Student t-test results comparing renal lengths of control and CKD groups

	T value	P value	Sig/ N.sig
RK, control Vs CKD	5.29	<0.00001	Significant
LK, control Vs CKD	4.89	<0.00001	Significant

P value < 0.01 is considered as significant. RK– right kidney; LK – left kidney

Table 3. Serum creatinine values of control & CKD groups

Serum creatinine (mg/dL)	Control group	CKD
Mean serum creatinine ± SD	0.83 ± 0.15	1.89 ± 0.55
Serum creatinine range	1.1-1.6	4.0-1.4

DISCUSSION

Natural history of CKD demonstrates a progressive decline in renal size with progressive loss of renal function (6,5). Present study analyzed the relationship between the sonograhically measured absolute renal length and serum creatinine value. Serum creatinine, an indicator of renal function, used to compare renal function and renal length in CKD and control groups. The results of the present study established a statistically significant deference in renal length between the two study groups, CKD and control groups. The renal length of the CKD group was

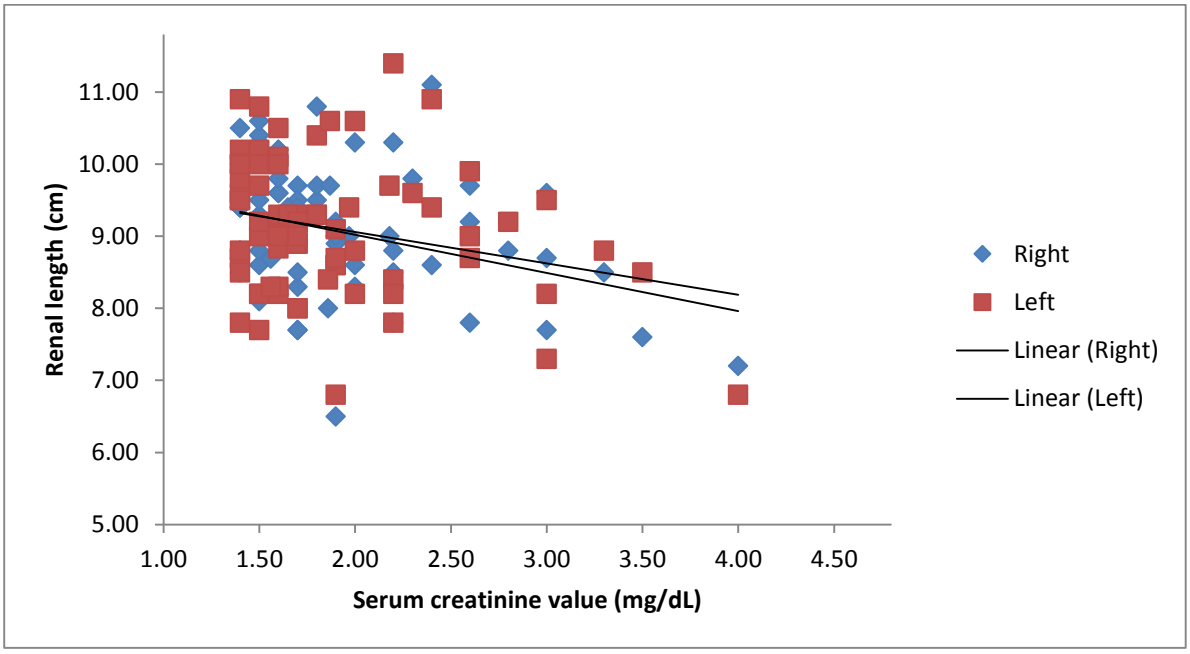

Figure 2: Serum creatinine value Vs renal length

significantly lower than that of the control group. This finding is in par with the results of previous similar studies (4). There was no significant difference in the renal length between right & left kidneys either within the control or within the CKD groups. A weak negative correlation (r- 0.328) was established between the renal lengths and Scr values.

Jovanovic *et al*. reported a strong negative correlation (-0.69) of the same parameters. They studied 49 CKD patients and 46 healthy adults as a control group (9). As in the present study, they have also not

catagcategorized CKD patients according to the etiology of renal disease, therefore a similar methodology. Some authors have described an etiology related variation in renal length in CKD patients (4). Kokoris *et al*. have studied 126 patients with chronic kidney diseases with different etiologies, including Balkan endemic nephropathy, diabetic nephropathy, autosomal dominant polycystic kidney disease, primary glomerular disease and compared renal length with a control group. They have established a significant difference in renal lengths with different etiologies. Diabetic nephropathy (DMN) is recognized as a common cause of CKD in Sri Lanka (10), therefore it is assumed that our study sample must have a significant number of DMN. In DMN, the renal size initially increases (4). Therefore, the established weak negative correlation of renal length and Scr in the present study could be partially due to this etiological variation. However, considering the fact that Jovanovic *et al*. have reported strong negative correlation in above parameters with similar methodology a population specific variability of renal length might play a role in the findings of the current study.

Absolute renal length was measured in the present study as maximum pole to pole to length of the kidneys. Previous studies have established a positive relationship of renal size with the height of the patient (4,9,11,12). In their studies, the relative renal length was calculated by dividing the absolute renal length (in millimeters) by the body height (in centimeters). They concluded that relative renal length as a better indicator of renal length compared to absolute renal length. Therefore, it will be of value to assess the relative renal length in order to investigate the association of renal length and renal function.

CONCLUSION

Findings of the current study established a significant reduction in absolute renal length in CKD patients when compared to the control group.

Weak negative correlation was elicited between the absolute renal length and the serum creatinine value in CKD, therefore with the renal function.

Further studies are recommended to evaluate the relationship between the relative renal length and serum creatinine values and to assess above parameters according to etiological categories of CKD.

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CORRESPONDENCE
Dr. S. K. Y. I. Kodikara
Department of Anatomy,
University of Ruhuna.

E-mail-iroshani.kodikara@gmail.com

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Early Feeding after Laparoscopic Assisted Pancreatico-Duodenectomy with Pancreatico-Gastric Anastomosis

K.B. Galketiya[1], V.Pinto[2], W.Rathnaweera[3], R.Nirmalasingham[3], R.M.I.M. Senevirathna[4], W.E.W.M.G.W.D.N. Wijerathna[4]

ABSTRACT

Introduction: Laparoscopic assisted pancreatico-duodenectomy (LAPD) reduces morbidity of open surgery. Enhanced recovery protocols has shown benefits from oral feeding from first post operative day. The study was done to evaluate out come with early oral feeding in patients undergoing LAPD and pancreatico-gastrostomy.

Method: Twenty two patients undergoing LAPD and pancreatico-gastrostomy were evaluated for out come after starting early oral feeding.

Results: In 22 patients studied 86% tolerated feeding from the first post operative day and the rest were fed by the third day. In 77% IV fluids were omitted by the fifth day. One patient had a prolonged drain output which settled after three weeks. One patient got readmitted after two weeks with a gastric bleed, underwent laparotomy, but died in the ICU with multi organ failure.

Conclusions: Early feeding was well tolerated after LAPD and pancreatico-gastrostomy.

Key words: laproscopy, pancreatico-duodenectomy, pancreatico-gastrostomy, ERAS

INTRODUCTION

The curative treatment for carcinoma of the peri ampullary region and head of pancreas is pancreatico-duodenectomy (PD). Considerable morbidity and occasional mortality is reported with the procedure. Long incision, handling of bowel and prolonged use of retractors can result in post operative respiratory inadequacy due to pain. Ileus can compound the problem. Blood loss is often significant. Laparoscopic assisted pancreatico-duodenectomy (LAPD) will minimize post-operative complications, which allow early discharge from hospital (1,2,3,4,5,6).

The pancreatic anastomosis may be to jejunum (pancreatico-jejunostomy-PJ) or the stomach (pancreatico-gastrostomy-PG). In addition there will be a choledocho/ hepatico jejunostomy and a gastro-jejunostomy. In LAPD all or some of anastomoses are performed through a mini laparotomy (3,6). PG is technically easier to perform through a mini laparotomy than PJ (7). PG is equally good or better than PJ when post operative outcomes are compared (8,9).

Traditionally oral feeding is delayed after PD for about five days. However with enhanced recovery protocols oral feeding from first post operative day one has shown benefits in many studies (10,11). However there is a need for further studies (10).

OBJECTIVE

The objective of the study was to evaluate the feasibility and out come with early feeding in patients who are undergoing LAPD and PG.

METHOD

A retrospective analysis of twenty two patients who underwent LAPD at the Professorial Surgical Unit Teaching Hospital Peradeniaya from February 2014 to April 2017 was done. The resection was completed laparoscopically. in three patients hepatico-jejunostomy was performed laparoscopically and pancreatico-gastrostomy and gastro-jejunostomy performed with mid line mini-laparotomy. In others, all three anastomosis were done by mini laperotomy. Oral liquids at 30ml/hour, was introduced from first day after abdominal examination. If tolerated, fluids were increased by doubling the amount daily and IV fluids were omitted when free oral intake was established. Soft solids were introduced on the third day for the patients tolerating oral liquids well. Solids were commenced by the sixth day for suitable patients. The volume of drainage from drains was measured daily. Abdominal drains were removed when drainage settled.

RESULTS

A total of 22 patients were operated, 12 being females. Age ranged from 36 to 74. There were 12 periampullary, 1 duodenal, 4 pancreatic head, 1 uncinate process and 4 distal CBD carcinomas. In nineteen (86%) feeding was established from next day, in two (9%) from 2nd and in one (5%) from 3rd day. In 17 (77%) IV fluids were gradually tailed off over 5days and in other 5 (23%) by the seventh day. None were started on total parenteral nutrition. In 19 cases drains were removed by the 7th day. In two cases, by the 9th and 10th day. In one case, drainage persisted for 3weeks. Oral feeding didn't increase drainage volume in any patient. One patient got readmitted with haematemesis after 2 weeks. Laparotomy revealed an unhealthy area with bleeding in the gastric fundus which was excised and sutured but the patient died in ICU after three days with multi organ failure. This brought a mortality of 4.5%

DISCUSSION

PD is associated with a considerable morbidity specially with regards to pancreatic fistulas. Many techniques of pancreatic anastomoses are described including a debate about PJ and PG as well as the technique of anastomosing (7,8,9). Traditionally oral feeding after PD is delayed due to multiple upper GI anastomoses. However Enhanced Recovery After Surgery (ERAS) protocols have shown that early oral feeding is well tolerated (10,11).

At Professorial Surgical Unit, Teaching Hospital Peradeniya, following LAPD PG is the preferred method of pancreatico anastomosis. In twenty two patients performed during a three year period were studied about the outcome with early oral feeding. Oral liquids were started from the first post operative day after evaluating the patient and gradually increased with careful monitoring. Majority, (19/22) tolerated oral liquids from post operative day one and rest by the third day. Non required TPN or restarting IV fluids. There was a readmission after two weeks who had no problems with early feeding and was on a normal diet on discharge.

CONCLUSION

Early oral feeding was well tolerated after laparoscopic assisted pancreatico-

duodenectomy with pancreatico-gastric anastomosis

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CORRESPONDENCE

Professor K. B. Galketiya
Department of Surgery,
Faculty of Medicine,
University of Peradeniya.

Email- kbgalketiya@yahoo.com

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The Position and Morphology of the Vermiform Appendix in Sri Lankans: A Study on Autopsies

P. M. N. S. Ekanayake[1], H. A. Amaratunga[1], A. N. Vadisinghe[2], P. Senasinghe[3], S. B. Adikari[1]

ABSTRACT

Introduction: The vermiform appendix is a vestigial structure showing extreme variability in its position. Varying symptoms of acute appendicitis reflect this variability making the clinical presentation of appendicitis highly inconsistent.

Objective: To observe the anatomical variations of the position of the tip of the appendix and its base, length, details of meso-appendix and appendicular artery in adult medico-legal autopsies.

Methods: A sample of medico legal autopsies carried out on those over 18 years by forensic pathologists at the Teaching Hospital Peradeniya was included in this study.

Results: Of the 60 autopsies, 39 were male, with an age range of 19-88 years. Fifty eight appendixes were healthy, 1 appendicular mass and 1 absent appendix. Positions of the appendices were: 29 (50%) post-ileal, 20 (34.48%) retro-caecal, 7 (12.07%) pelvic and 2 (3.45%) para-caecal. The base was at the postero-medial wall in 36 (62.07%), lower pole in 18 (31.03%) and postero-lateral wall of the ceacum in 4 (6.0%) subjects. The length was 3cm - 14 cm (mean 8.2 cm). The distance between ileo-caecal valve and base of the appendix was 1 cm - 6 cm (mean 2.8 cm). The distance between the edge of the meso-appendix and the tip was 0 cm - 7 cm (mean 1.23 cm). In 51.7 % of cases meso-appendix continued to the tip. The appendicular artery continued to the tip in 37 (63.8%) cases.

Conclusions: Post ileal position is the commonest position of the appendix, with the base situated commonly on the postero-medial wall of the caecum. In the majority of cases meso-appendix and the appendicular artery continued to the tip.

INTRODUCTION

The Vermiform appendix is a vestigial structure in humans and is represented by a blind ended muscular tube, situated at the confluence of the three Taenia coli of the caecum. The appendicular base is described to have a relatively constant position in the postero-medial wall of the caecum (1), although it can sometimes be found on the postero-lateral wall and the lower pole of the caecum (1). The appendix is described to be of variable length but usually ranges from 7.5–10 cm in adults (1). The appendix is known to show extreme variability in its position and morphology. The classical positions described in the text books include retrocaecal, pelvic, pre-ileal, post-ileal, para-caecal, sub-caecal, and promonteric (2, 3).

Even though it's a vestigial structure with no proven function, it still qualifies attention due to the fact that it gets inflamed rather commonly, resulting in acute appendicitis, one of the most common cause for an acute abdomen in surgical casualties. Though a classical

[1]Department of Anatomy, Faculty of Medicine, University of Peradeniya.
[2]Department of Forensic Medicine, Faculty of Medicine, University of Peradeniya.
[3]Medico-Legal Unit, Teaching Hospital Peradeniya.

sequence of symptoms is described for acute appendicitis may vary making the clinical presentation of appendicitis highly inconsistent (4,5). The reason for this inconsistency could be due to the variability of its relationship with the surrounding structures. Notwithstanding the advances in modern radiographic imaging and laboratory investigations, the diagnosis of appendicitis remains essentially clinical, requiring adequate knowledge of the anatomical variations (6,7). Furthermore appendicitis may result in surgical removal of the appendix which requires a clear knowledge about the anatomy and relationships to other structures (8). Even though many studies have reported different positions of the appendix, racial differences may exist.

The appendicular position in the Sri Lankan population has not been documented to date. Therefore it was the aim of our study to observe the anatomical variations, position of the tip of the appendix & its base, length, extent of the meso-appendix and the distribution of the appendicular artery, during medico-legal autopsies.

METHODS

A random sample of autopsies done by Forensic Pathologists from November 2011- November 2012 at the Teaching Hospital Peradeniya was included in the study. Putrefied bodies, bodies with abdominal pathologies and abdominal trauma, surgical interventions or procedures necessitated to interfere with the position of the appendix, deformities which may affect the anatomy of the appendix (lower spinal abnormalities), microscopically abnormal appendices and any condition which could have affected length were excluded. Subjects whose age was below 18 years were excluded from the study. Ethical clearance was obtained from the ethical review committee of the Faculty of Medicine, University of Peradeniya.

The anatomical details of the appendix were observed immediately after opening the abdominal cavity; before any manipulation the position of the tip of the appendix was noted. When the appendix was not visible the ceacum was mobilized and shifted carefully to observe the appendix. Next, the ilioceacal valve and the base of the appendix were identified. The exact position of the base on the wall of the caecum was noted. Then the length between the ileoceacal valve and the base of the appendix was measured using a calibrated metal Vernier caliper. The appendix was carefully unfolded and held straight. The length from the base to the tip was measured using the Vernier caliper. The mesoappendix was then studied carefully and the edge of the structure was located. Then the length between the edge of the mesoappendix and the tip of the appendix was measured with the Vernier caliper. All details were recorded in two diagrams specially designed for the purpose (figure 1 and 2).

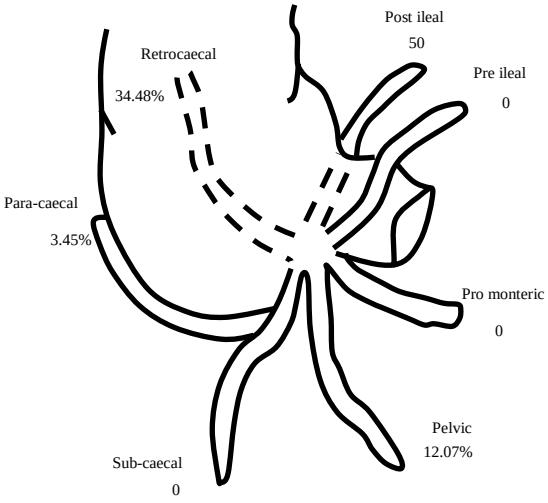

Figure 1: positions of the tip of the appendix

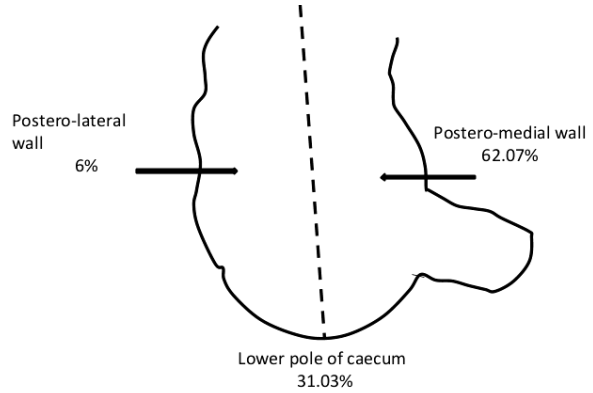

Figure 2: Position of the base of the appendix

RESULTS

Of the 60 autopsies, 39 were males and 21 were females. Age of these subjects varied from 19-88 years. Mobile healthy appendices were noted in 58 cases, and one appendicular mass was found. The appendix was absent in one case with no history of appendisectomy. Of the appendices, 29 (50%) were post-ileal, 20 (34.48%) retro-caecal, 7 pelvic (12.07%), and 2 (3.45%) were para-caecal (Figure 1). No pre-ileal, sub-caecal or promonteric appendices were noted. The base of the appendix was found on the postero-medial wall of the caecum in 36 (62.07%), on the lower pole of the caecum in 18 (31.03%) and on the postero-lateral wall in 4 (6.0%) of the cases (Figure 2). The length of the appendix varied from 3cm to 14 cm with a mean value of 8.2 cm. The distance between the ileo-caecal valve and the base of the appendix varied from 1 cm to 6 cm with a mean value of 2.8 cm. The distance between the edge of the meso-appendix and the tip of the appendix varied from 0 cm (meso appendix continues to the tip of the appendix) to 7 cm with the average being 1.23 cm. In 30 cases, (51.7 %) the meso-appendix continued to the tip of the appendix. All appendicular arteries studied were end arteries. The appendicular artery continued to the tip of the appendix in 37 (63.8%) cases while the continuation was not observed in 21 (36.2%). In all cases where the mesoappendix continued to the tip, the appendicular artery continued to the tip as well. In 7 of the cases the appendicular artery continued to the tip even though the mesoappendix ended before reaching the tip.

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DISCUSSION

Human vermiform appendix shows extreme anatomical variations and this has implications on the diagnosis and management of appendicitis (1). The post-ileal position was the commonest position seen among this group of Sri Lankan adults with the retroceacal position being the next commonest position. Most studies report the retrocaecal and retrocolic position as the commonest, while the post ileal position is reported as the second commonest (9-13). However some studies report the pelvic appendix as the second commonest position (10). In studies done on appendectomy patients the retrocaecal position is described as being the commonest (11). In the current study the base of the appendix was commonly situated on the postero-medial wall of the caecum. This is the classical position of the base described in books and do not give details about the other positions of the base. We found that 30% of the cases had the base at the lower pole of the ceacum. Some studies report the lower pole as the commonest position of the base of the appendix (9). The distance between the ileocaecal valve and the base of the appendix was 2.8 cm and is consistent with published data (3,9,10,12). The length of the appendix is reported as being between 2- 25 cm with the average being around 9 cm (2,13). Similar results were

obtained by us, with the average length being slightly less at 8.2 cm. In about 50% of the cases meso-appendix and the appendicular artery continued to the tip of the appendix. However in the rest the mesoappendix ended at an average of 1cm before reaching the tip. In 7 of the cases the artery traveled over the wall to reach the tip. Similar findings have been reported by other researches (13,14). Appendices that have an appendicular artery that does not reach the tip maybe at a higher risk of ischaemic necrosis during inflammatory processes.

It is apparent that the appendicular position and its morphology and blood supply is highly variable. Therefore a good knowledge about the appendix and its morphology is necessary for the clinicians to make a clear diagnosis and to perform a surgery with least complications.

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CORRESPONDENCE

Dr. H. A. Amaratunga
Department of Anatomy,
University of Peradeniya.

E mail-himanipeiris@yahoo.com

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Difficulties in the Diagnosis of Myocarditis –Case report

R. H. A. I. Rathnaweera[1]

ABSTRACT

Myocarditis is an elusive illness to study, diagnose and treat because the clinical presentation may range from nearly asymptomatic to overt heart failure. A 61 years old female developed fever with chills and rigors during a pilgrimage. After returning, she developed shortness of breath, with retrosternal chest pain and admitted to Teaching Hospital Karapitiya. An E.C.G. (Echocardiography) showed ST elevation in V1 – V6 and ST depression in II, III and AVF leads. The patient was treated as having an acute anterior ST elevation MI (myocardial infarction) treated with heparin and anti anginal drugs. The patient died about six hours after the admission. At the post-mortem, heart appeared flabby with 30% occlusion of the left coronary artery. Histopathology examination revealed acute myocarditis. Many secondary causes can lead to inflammation of the myocardium and therefore the diagnosis of myocarditis cannot be made by evidence of inflammation of the myocardium alone. Therefore, unless multiple, florid foci are found with myofibril necrosis, bland mononuclear foci that are purely interstitial and not involving muscle fibers must be disregarded.

Keywords: myocarditis, post-mortem histopathology

INTRODUCTION

Myocarditis is an elusive illness to study, diagnose and treat because the clinical presentation may range from nearly asymptomatic to overt heart failure and sudden death (1). It may even resemble acute coronary ischemic syndrome. Sometimes misdiagnosis of myocarditis is made and sometimes we mis the diagnosis of myocarditis.

CASE REPORT

Sixty one years old female, married, mother of six children, developed fever with chills and rigors for two days during a trip to Rathnapura. After returning home, on the following day morning, she developed shortness of breathing, together with retrosternal chest pain. No fever was present at that time. She was admitted to the Teaching Hospital Karapitiya, immediately and was taken to the E.T.U. At the E. T. U. an urgent E.C.G. was taken and it showed ST elevation in V1 – V6 together with ST depression in II, III, and AVF leads. She was treated as for anterior ST elevation myocardial infarction, with heparin and anti anginal drugs. But she deteriorated gradually and died about six hours after admission to the E.T.U.

At the post-mortem, heart appeared flabby, with 30% occlusion of the anterior descending branch of the left coronary artery. All the other organs appeared unremarkable. Histological examination revealed diffuse mixed inflammatory cell infiltrates rich in lymphocytes. There was focal necrosis of myocardium around which inflammatory infiltrate was prominent (Figure 1 and 2). The above features are compatible with acute myocarditis, most probably viral in origin.

[1]Department of Forensic Medicine, Faculty of Medicine, University of Ruhuna.

DISCUSSION

Myocarditis refers to a process of diffuse inflammatory infiltrate and focal areas of necrosis of the myocardium. Under this category, inflammatory processes of the myocardium that result in injury to cardiac myocytes are grouped (2). However, the presence of inflammation alone is not diagnostic of myocarditis, because inflammatory infiltrate may also be seen as a secondary response in conditions such as ischemic injury (2). In myocarditis, by contrast, the inflammatory process is the cause of rather than a response to myocardial injury (2).

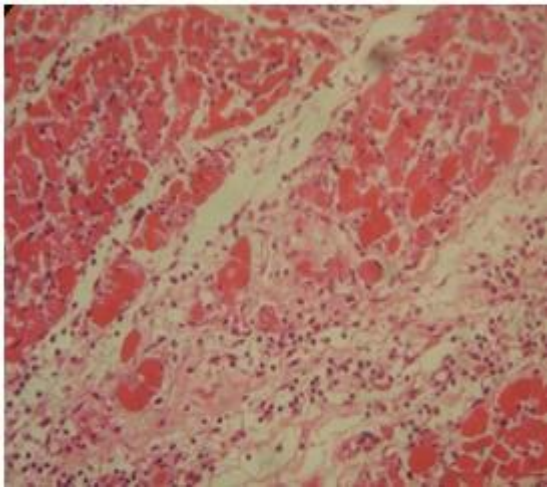

Figure 1: Lymphocyte rich inflammatory cell infiltrate with focal myocardial necrosis-lower (x10) magnification

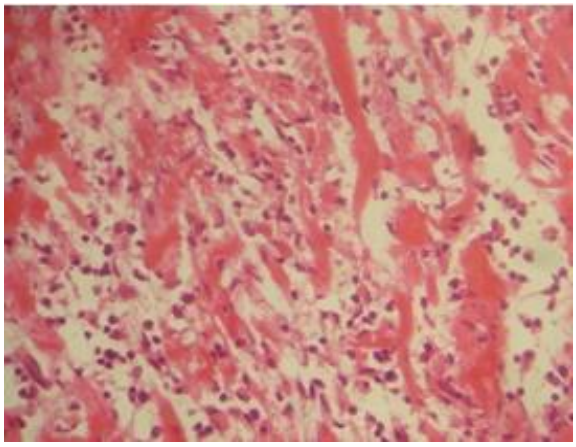

Figure 2: Lymphocyte rich inflammatory cell infiltrate with focal myocardial necrosis–higher (x40) magnification

Infections and particularly viruses are the most common cause of myocarditis. Coxackie viruses A and B and other entero viruses account for most of the myocarditis cases. Viral myocarditis, as in this case, is characterized by mononuclear inflammatory infiltrate composed largely of lymphocytes and associated myocyte injury (2). Other less common etiologic agents include Chlamydia, Rickettsia, Bacteria, Fungi and Protozoa. A morphologically distinctive form of myocarditis of uncertain cause, called giant cell myocarditis, is characterized by a wide spread inflammatory cellular infiltrate containing multinuclear giant cells interspersed with lymphocytes, eosinophils, plasma cells and macrophages and having at least focal but frequently extensive necrosis. These giant cells are either of macrophage or myocyte in origin. These variant cells carry a poor prognosis (2). Hypersensitivity myocarditis is characterized by an interstitial inflammatory infiltrate composed largely of eosinophils and mononuclear inflammatory cells predominantly localized to perivascular and large interstitial spaces. This form of myocarditis is associated with drug hypersensitivity.

The Dallas criteria were proposed in 1986 and provided a histopathological categorization by which the diagnosis of myocarditis could be established (3). According to the Dallas criteria myocarditis requires an inflammatory infiltrate and associated myocyte necrosis or damage not characteristic of an ischemic event. Borderline myocarditis requires a less intense inflammatory infiltrate and no light microscopic evidence of myocyte destruction (4).

These criteria have been used exclusively by American investigators over the last two decades. Sampling error, variation in expert interpretation, variance with other markers of viral infection and immune activation in the heart and variance with treatment outcomes all suggest that the Dallas criteria are no longer adequate. It was demonstrated by biopsies done on post-mortem hearts of patients who had died with myocarditis that, from a single endomyocardial biopsy, histological myocarditis could be demonstrated in only 25% of samples (5). With more than 5 biopsies, Dallas criteria myocarditis could be diagnosed in approximately two thirds of subjects. This point outs the importance of taking multiple sections of the heart in any complete examination of the heart for heart disease.

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CORRESPONDENCE

Dr. R. H. A. I. Rathnaweera
Department of Forensic Medicine,
University of Ruhuna, Sri Lanka.

E mail-ajithrathnaweera@gmail.com

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Metastatic Melanoma Presenting as a Subcutaneous Nodule with Complete Regression of the Primary Lesion

T. G. Liyanage[1], T. W. Wijesiri[1], P. Weerasiri[2]

INTRODUCTION

Melanoma is a skin cancer arising from the malignant transformation of melanocytes. Melanoma comprises less than 5% of malignant skin tumors and it accounts for almost 60% of lethal skin cancers (1). Skin metastases are not uncommon among patients with melanoma and frequently are the first sign of extra-nodal metastatic disease (2). In 2 to 9% of the patients presenting with cutaneous melanoma, the primary site of the tumor cannot be detected (3,4,5,6). This can be due to complete regression of the primary lesion.

Here a rare case of metastatic melanoma with complete regression of the primary lesion is described.

CASE REPORT

A 32-year old woman presented with a gradually enlarging nodule on her lower back. She claimed that there was a small nevus adjacent to the nodule which was there from birth. The nevus gradually enlarged recently with change in colour and regressed with time.

Physical examination revealed a solitary, mobile, soft to firm lump measuring 5x3.5x3.5 cm which was not attached to the skin. Overlying skin showed a pale blue discoloration without any significant lesions. The initial clinical diagnosis was a lipoma.

Due to the suspicious nature of the lesion during surgery, the lesion was completely excised including the overlying skin. The specimen was submitted for histo-pathological assessment. Macroscopically there was a fairly circumscribed lesion within the subcutaneous fat. Cut surface revealed a solid brownish black tumour with focal whitish areas. Overlying skin was macroscopically unremarkable.

Microscopical examination revealed a tumour within the subcutaneous fat sparing the dermis and epidermis. The tumour was composed of solid sheets, nests and proliferated fascicles of atypical melanocytes. The cells had nuclei with mild to moderate nuclear pleomorphism with prominent eosinophilic nucleoli. Melanin pigment was seen in the cells with variable melanin distribution. The entire skin ellipse was sampled and examined. There was no evidence of atypical melanocytic proliferation with junctional activity. An area of the dermis resembling the regressed area of the primary lesion was identified. This area showed prominent collections of melanophages associated with fibrosis of the papillary dermis and subepidermal perivascular collections of lymphocytes. Based on the histopathological features, a diagnosis of a metastatic deposit of a malignant melanoma was made. With the absence of skin involvement the usual Clark's or Breslow's staging for melanoma was precluded. The diagnosis was confirmed by positivity for Melan A by immune-histochemistry.

Additional wide local excision was later carried out, including adjacent skin and subcutaneous tissue. It was negative for residual malignancy. As there were no

Department of Pathology, Faculty of Medicine, University of Ruhuna[1], DH Tissamaharama[2]

other detectable lesions, and no regional lymphadenopathy confirmed by USS, the decision was made for close clinical and radiologic surveillance of the patient.

Figure 1: Cut surface of the tumour showing whitish and blackish areas. Black areas represent tumour cells containing melanin pigment.

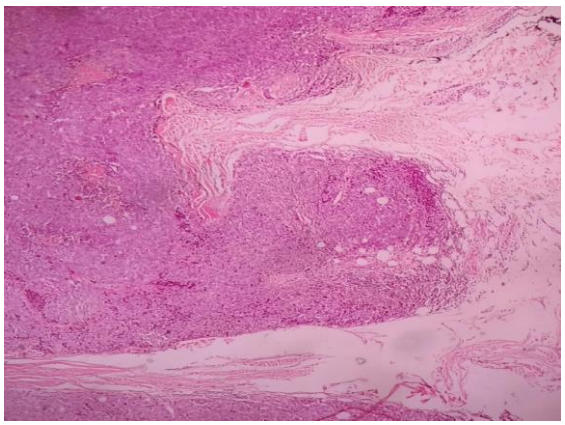

Figure 2: Tumour within the subcutaneous fat (H&E X100)

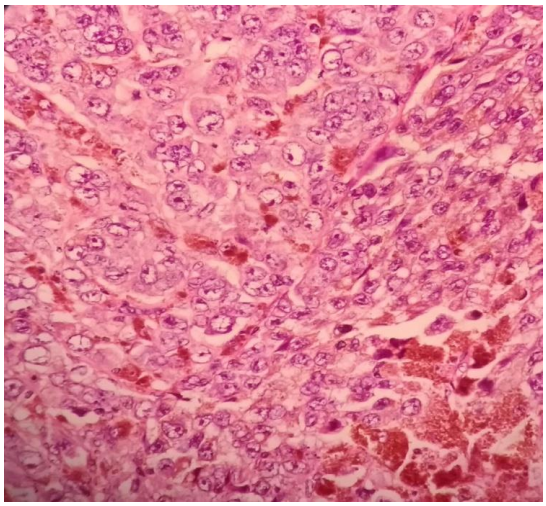

Figure 3: Tumour cells showing prominent eosinophilic nucleoli and intracytoplasmic melanin pigmemt (arrow) (H&E X400).

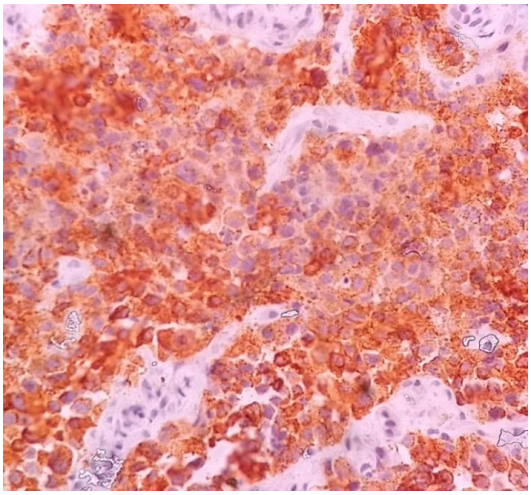

Figure 4: Tumour cells showing strong cytoplasmic Melan A positivity (Immunohistochemistry X400).

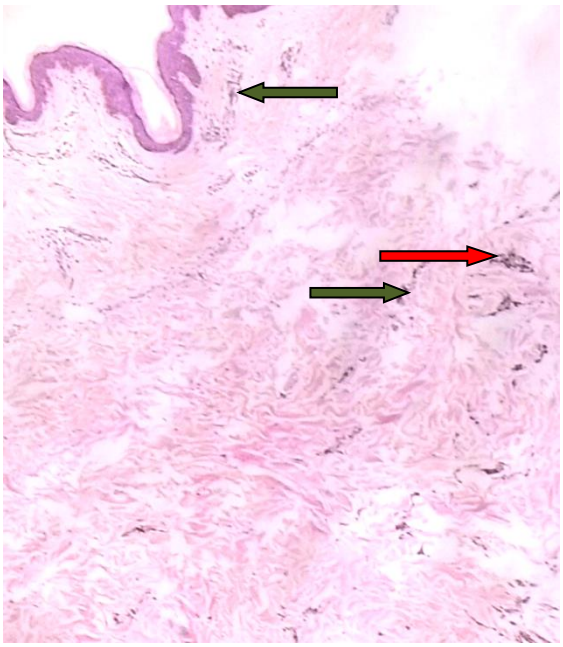

Figure 5: Adjacent dermis showing collections of melanophages (red arrow), perivascular lymphocytes (green arrow) and papillary dermal fibrosis. There are no atypical melanocytes in the epidermis or dermis (H & E X100)

DISCUSSION

Melanoma is a lethal skin cancer arising from the malignant transformation of melanocytes. It accounts for 5.2% all new cancer cases and 1.6% of all cancer deaths (7). Melanoma is reported as the 19th most common cancer worldwide, with estimated

age-standardized incidence rates of 2.8–3.3 per 100,000 (7). Cutaneous melanoma is the most common melanoma subtype, accounting for around 90% of cases of melanoma (8). The commonest presenting symptom of a melanoma is a pigmented lesion of changing size, followed by a change in colour and pain or tenderness. On clinical examination they usually show papules, nodules or masses in approximately 45% and as flat topped plaques, macules or patches in approximately 45% (9). Majority of cutaneous melanomas are associated with sunlight exposure but around 34% melanomas arise from the malignant transformation within a benign nevoid lesion (9).

Melanomas commonly metastasize to any organ or tissue in the body. Patients with metastatic melanoma generally have a poor prognosis. The duration of survival for metastatic melanoma is less than a year and the median survival is nearly 6 to 8 months (10). Skin metastases are relatively frequent in the natural history of melanoma and can develop in early as well as in late stages of the disease. Around 10-17% melanoma patients develop cutaneous and subcutaneous metastasis during the disease course (11). Melanoma presenting solely as solitary lesions involving the dermis and/or subcutaneous tissue varies from 0.51% to 0.92% (12). In 2 to 9% of the patients presenting with cutaneous melanoma, the primary site of the tumor cannot be detected (3,4,5,6). This can be explained by complete regression of the primary lesion.

In our patient melanoma presented as a subcutaneous nodule and due to the absence of dermal involvement after extensive sampling of the overlying skin, it was diagnosed to be a secondary deposit from a melanoma. Inspite of thorough clinical examination and radiological evaluation a primary site could not be identified. Considering the patients history, clinical and histopathological evidence, the skin lesion that was present adjacent to the subcutaneous melanoma deposit was assumed as the primary lesion which has regressed completely.

Spontaneous regression of malignant melanoma is defined by the disappearance of melanocytic neoplastic cells partially or completely. In contrast to the partial form, complete regression of primary malignant melanoma is a rare phenomenon, with an incidence of 0.22% to 0.27% (13). In our patient also the primary lesion completely regressed leaving the subcutaneous metastatic deposit. Complete regression is commonly seen with metastasis disease and only one case is reported in the literature with complete regression without metastasis (14). Clinically regressing melanoma usually show depigmentation within or around the lesion. Histopathological features of tumour regression include diminishing or disappearing of tumor melanocytes, appearance of newly formed blood vessels, collections of melanophages, lymphocytes in dermis and papillary dermal fibrosis.

The significance of primary regression on the prognosis of melanoma patients is controversial. In early days, studies have demonstrated that melanomas with regression has poor prognosis than melanomas without regression and some studies have failed to demonstrate such a relation (14). According to the current literature, regression in melanoma is less likely to be associated with sentinal lymph node metastasis and therefore now it is regarded as a good prognostic factor (15). However, the incidence of metastatic

disease in the setting of complete regression of the primary lesion is estimated to range from 4% to 10% (16). The regression phenomenon is presumed to be the result of a host immune response. It is proved that natural tumor-specific cytotoxic T cell response play an important role in primary regressing human melanoma (17).

CONCLUSION

This case report presents a rare diagnosis of a secondary deposit from a melanoma with complete regression of the primary lesion. The appearance of a solitary subcutaneous metastasis without an evident primary tumor can be explained by spontaneous regression and complete disappearance of the primary melanoma after dissemination to a secondary site. It also should be remembered that in the presence of a melanoma in subcutaneous fat, the clinician needs to perform a meticulous search for a primary lesion and for signs of metastasis in regional lymph nodes and in common metastatic sites. In the setting of localized metastatic disease with complete regression of the primary lesion, it is important of have close clinical and radiological follow up of patients, to identify widely metastatic disease.

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CORRESPONDENCE

Dr. T. G. Liyanage
Department of Pathology,
University of Ruhuna

E mail-thusharieliyanage@yahoo.com

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Sudden Death of an Infant due to Endocardial Fibroelastosis - Case Report

R. H. A. I. Rathnaweera[1]

ABSTRACT

Endocardial fibroelastosis (EFE) is an idiopathic disorder of the tropical and subtropical regions of the world that is characterized by the development of restrictive cardiomyopathy. It is characterized by proliferation of both elastic and collageneous fibers within the endocardium, causing diffuse or localized thickening. A three month old baby was admitted to a local hospital in Galle, with a history of sudden onset shortness of breathing for one day. Baby was found to be in a state of heart failure and died within few hours after admission. At the post-mortem examination, heart was found to be enlarged with dilated left ventricle. The endocardium of both ventricles was markedly thickened. Histopathology showed a thick layer of collagenous fibrous tissue and elastin fibres confirmed by Vangisen Elastin stain. The cause of death was given as endocardial fibroelastosis. The underlying mechanisms of myocardial fibrosis remain unclear. It is hypothesized that infectious, inflammatory and nutritional processes are involved in this process. This case highlighted the importance of special histology staining methods when arriving at the cause of death.

Key words: infant, sudden death, endomyocardial fibroelastosis, restrictive cardiomyopathy

INTRODUCTION

Endocardial fibroelastosis (EFE) is an idiopathic disorder of the tropical and subtropical regions of the world that is characterized by the development of restrictive cardiomyopathy (1). It is characterized by proliferation of both elastic and collageneous fibers within the endocardium, causing diffuse or localized thickening. This leads to endocardial stiffness and severely reduced contractility of the involved ventricles.

This is a condition that continues to arouse interest because of its unfavorable prognosis. The cases of endocardial fibroelastosis reported in the literature are generally of small children (2).

Its presentation in newborns is considered rare. Number of cases reported in Sri Lanka is very few.

CASE REPORT

A three month old baby was admitted to a local hospital in Galle, with a history of sudden onset shortness of breathing for one day. He was apparently well prior to this incident. No history of fever, cough or refusal of feeding. No significant ill health in the past as well. No consanguinity among the parents of this family. This baby was the first child of this family and it was a normal vaginal delivery when the mother was 26 years old. Antenatal history was uncomplicated. Baby was found to be in a state of heart failure on admission and despite ward management, his condition deteriorated rapidly. Cause for the heart failure was not diagnosed and few hours later, baby died. An inquest was held and a post-mortem was ordered.On general examination, no congenital abnormalities

[1]Department of Forensic Medicine, Faculty of Medicine, University of Ruhuna.

were seen and the face was found to be congested. No injuries were seen. Body weight was 5.3kg. Brain appears unremarkable. Both lungs were congested and squeezing reveals a fine froth. Kidneys and liver also found to be congested. Heart was enlarged with dilated left ventricle. The endocardium of both ventricles was markedly thickened with the involvement of the papillary muscles and the chordae tendinae (Figure 1,2,3). The myocardium was normal. The microscopic sections done from several areas of both the right and the left ventricle showed a thick layer of collagenous fibrous tissue and elastin fibers confirmed by Verhhoeff Elastin stain (Figure 4, 5, 6). The cause of death was given as endocardial fibroelastosis.

Figure 1: Thickened endocardium of the right ventricle

Figure 2: Thickened endocardium of the left ventricle

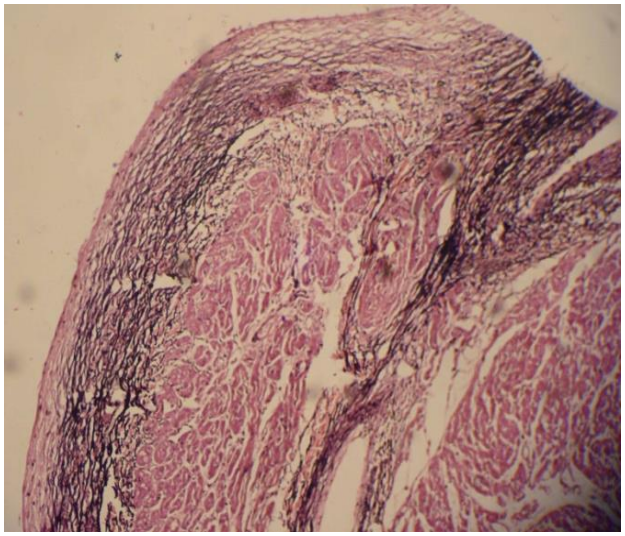

Figure 3: Verhhoeff Elastin stain of the endocardium- lower (x 10) magnification

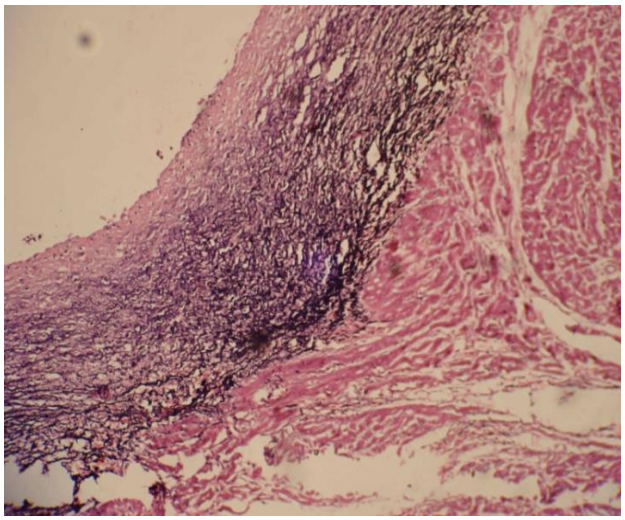

Figure 4: Verhhoeff Elastin stain of the endocardium-higher (x40) magnification

DISCUSSION

Endocardial fibroelastosis is a rare disorder in newborns, accounting for no more than 1-4% of total congenital heart disease. It occurs one in every 5000-6000 live births (1). Around 80% of patients present with congestive heart failure within the first year of life. The distinction into primary and secondary endocardial fibroelastosis has recently been challenged and according to more recent thinking, EFE is simply a reaction to chronic myocardial stress (3). This stress may be

due to congenital disorders of the vessels and or valves, to viral agents, or to genetic causes determining dysfunction of the myocardium (4). In general, fibrosis in cardiac tissue has been mainly linked to increased level of a cytokine, transforming growth factor-β1. The underlying mechanisms of myocardial fibrosis in this specific entity remain unclear. Hypotheses include infectious, inflammatory and nutritional processes as causes (3). EFE is frequently associated with concomitant parasitic infections (eg, helminthes) and their attendant eosinophilia, although the role of parasitic infections and/or the eosinophil remains speculative. The development of EFE as a sequel to toxoplasma-related myocarditis has also been described, as has a relationship of malarial infection to development of EFE. However, no specific organism has been consistently associated with EFE (5).

The endocardial fibrosis develops in the foetus too, the endocardium is the layer most sensitive to injury, being the last to be supplied by the coronary arteries. Many studies have shown that endocardial smooth muscle cells exposed to chronic stress, proliferate and undergo a transition to fibroblasts, producing both collagen and

elastin (6). It is not yet clear how this occurs, but it would seem to be similar to occurrences in other organs. Patchy fibrosis of the endocardial surface of the heart leads to reduced compliance and, ultimately, restrictive physiology as the endomyocardial surface becomes more generally involved. Endocardial fibrosis principally involves the apices of the right and left ventricles and may affect the atrioventricular valves mainly by tethering the papillary muscles, leading to tricuspid and mitral regurgitation. Ventricular stiffness along with atrioventricular valvular regurgitation results in atrial enlargement, which has been linked to atrial arrhythmias such as atrial fibrillation. Fibrosis also reduces conduction velocity, impairs activation pattern and may provide the substrate for wave breaks and reentry (7).

The earliest changes of EFE are not well described because most patients do not present with symptoms until relatively late in the clinical course. Olsen described 3 phases of EFE (8). The first phase involves eosinophilic infiltration of the myocardium with necrosis of the sub endocardium and a pathologic picture consistent with acute myocarditis. This is reportedly present in the first 5 weeks of the illness. The second stage, typically observed after 10 months, is associated with thrombus formation over the initial lesions, with a decrement in the amount of inflammatory activity present. Ultimately, after several years of disease activity, the fibrotic phase is reached, when the endocardium is replaced by collagenous fibrosis. This patho-morphologic schema is not observed uniformly and has not been consistently supported by other investigators.

EFE is most frequently observed in the socially disadvantaged and in children and young women. These groups frequently have malnutrition and in regions of sub-Saharan Africa where the disease is most prevalent, the typical diet is high in a tuber called cassava, which contains relatively high concentrations of the rare earth element Cerium (Ce). The combination of high Ce levels and hypomagnesemia has been shown to produce EFE-like lesions in laboratory animals (9). This case highlighted the importance of special histological staining methods when arriving at the cause of death.

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CORRESPONDENCE

Dr. R. H. A. I. Rathnaweera
Department of Forensic Medicine,
University of Ruhuna, Sri Lanka.

E mail-ajithrathnaweera@gmail.com

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Anaesthetic Management of a Laparoscopic Pancreatico-Duodenectomy - Case report

W. M. A. S. B. Wasala[1], K. B. Galkatiya[1], M. V. G. Pinto[1]

ABSTRACT

A pancreatoduodenectomy involves the removal of a tumour of the peri ampullary region and head of the pancreas. It is a technically difficult procedure requiring experienced anaesthesia and surgical teams. We report the peri-operative management of a patient undergoing laparoscopic pancreaticoduodenectomy.

Key words: laparoscopic, whipple, pancreatico-duodenectomy, anaesthesia, epidural

INTRODUCTION

Whipple procedure or pancreatico-duodenectomy is the surgical treatment of choice for carcinoma of the peri ampullary region and head of pancreas. The open procedure is associated with considerable morbidity and occasional mortality (1). Recently, laparoscopic pancreatico-duodenectomy has started to gain wider acceptance as surgeons become more comfortable with laparoscopic technology and skills. The anesthetist plays a key role in peri operative management and can significantly influence patient outcome. Epidural analgesia, prevention of hypothermia, careful fluid balance, close intra and post operative monitoring are key strategies that improves outcome of these patients (2).

CASE REPORT

A 53 year old lady presented with obstructive jaundice and examination revealed a palpable gallbladder. The patient was a diabetic for the last 10 years. A contrast enhanced computed tomography (CT) scan revealed dilatation of the intra and extra hepatic biliary ducts with a carcinoma of the head of the pancreas, which was confirmed by Magnetic Resonance Cholangio Pancreatography (MRCP). The patient was scheduled for laparoscopic Whipple procedure.

Pre operative laboratory findings included a haemoglobin level of 12g/dl, platelet count of 235×109, INR of 0.86, bilirubin of 88umol/l, AST of 67 IU/l, alkaline phosphatase of 291 IU/l, normal renal function and serum electrolytes. On the day of the surgery blood sugar was 7.8mmol/l. Echocardiography revealed sclerosis of the mitral valve, functional mitral regurgitation and ejection fraction of 60%, mild obstruction in lung function test, FEV1>80%. ECG revealed no significant changes.

Pre operative vital signs were as follows; blood pressure, 107/54 mmHg, pulse rate 116 bpm, oxygen saturation of 98% while breathing room air. Lungs and airway assessment were clinically normal. Anaesthetic plan of general anaesthesia with epidural analgesia was explained and consent

[1]Faculty of medicine, Uuniversity of Peradeniya.

was taken. Patient was minimally fasted and hydrated adequately over night. Standard monitoring of vital signs was started in the theatre, epidural catheter was inserted at T8 -T9 level. After pre oxygenation with 100% oxygen, general anaesthesia was induced with 100ug of fentanyl, 150 mg propofol, 40 mg of atracurium. The patient was intubated with 7.5 cuff endotracheal tube. Tube placement was confirmed and a nasogastric tube was inserted to decompress the stomach. Co-amoxiclave 1.2g and morphine 6mg was given before starting the procedure. General anaesthesia was maintained with 1.2-1.5% Isofluraine in 50% oxygen and air. A 16F, 18F, 18F, 3-lumen central line was inserted in the right internal jugular vein and a 20gauge arterial line was inserted in the left radial artery. A forced air warming blanket at 43°C was used. All fluids were heated using a fluid warming device. The urine output was measured via an indwelling urinary catheter.

The patient was placed in reverse-Trendelenburg 20 degrees and rotated to the left by 30 degrees with legs abducted to 60 degrees. Pneumoperitoneum was created after 30 minutes of induction, CO_2 was insufflated at pressure of 14 mmHg. After creating the pnumopertonium central venous pressure rose from 9cm H_20 to 16cmH_20, peak airway pressure from 16mmHg to 27mmHg, $ETCO_2$ from 28mmHg TO 39mmHg. Ventilation was adjusted to maintain the $ETCO_2$ between 35-40mmHg and peak airway pressure was kept below 30mmHg. There was a drop of 3cm H_20 CVP and 5mmHg airway pressure after releasing the pnumopertonium. Serial glucose measurements remained between 163 and 230 mg/dL and did not require pharmacologic intervention.

The patient was haemodynamicly stable through out the surgery. Mean arterial blood pressure was maintained between 70mmHg to 80mmHg. Urine out put remained more than 0.5ml/kg hour(total of 400ml) with slight reduction during the period of pnumopertonium. Ringer's lactate 2000ml, hetarstarch 200 ml and 450ml of fresh frozen plasma were used during the procedure. The time for resection was 390 minutes with a blood loss of 400ml.Total amount of CO_2 used was 498 liters. Repeated arterial blood gas measurements showed good oxygenation, adequate control of $PaCO_2$ and minimal metabolic derangements (PH, K^+, Na^+, Lactate).

Epidural was top up with 10ml of 0.1% bupivacaine with 2ug ml fentanyl before leaving the theatre. Patient was transferred to the ICU and haemo-dynamicaly stable over night. The patient was extubated and epidural analgesia was continued during the ICU stay. Octeotride 250ug 8 hourly subcutaneous dose was started. The patient required a minimum amount of morphine during her ICU stay and sent to the ward on post operative day 4, discarded from the hospital on day 10.

DISCUSSION

Pancreatic tumour is one of the commonest cause of cancer related deaths (4). Currently, the only curative treatment for pancreatic cancer is surgical resection (4). Whipple procedure, or pancreatico-duodenectomy is one of the technically challenging major abdominal procedure occasionally results in increase morbidity

and mortality(4). Due to the advanced age, associated co morbidities and complexity of the surgery, both surgeon and anaesthesiologist play a crucial role in peri operative evaluation and management. These patients are referred early to tertiary care centres where multidisciplinary team involvement is available.

Most striking clinical symptom in these patient is painless jaundice. Ultra sound scan is the first line imaging test and has a sensitivity of more than 90% of site of the obstruction. Dilatation of the common bile duct and pancreatic duct are seen in patients with a pancreatic head tumour. CT and MRI scan have high sensitivity of detecting small tumours. Magnetic Resonance Cholangio pancreatography provides detail imaging of hepatobiliary and pancreatic systems(4).

Despite current developments in operative techniques and postoperative care, pancreatic surgery is associated with high morbidity and mortality. Potential complications and information about the therapeutic procedures and recovery should be informed to patient. According to the current evidence, routine use of long acting preoperative anxiolytics, optimisation of nutrition in malnourished have shown mild to moderate evidence (3). Association between renal disfunction and obstructive jaundice, is well established (5,6). Minimum fasting and adequate hydration during the perioperartve period reduce the incidence of renal failure (3).

Post operative mortality and morbidity depends on many factors. Cardio respiratory diseases are the most commonly observed co morbidities, age related medical problems an increasing incidence of diabetes decrease patients ability to respond to surgical stress. Insulin resistance and hyperglycaemia are strongly associated with post operative mortality and morbidity (3). Treatment of hyperglycaemia with intravenous insulin during the peri operative improves outcome but hypoglycaemia remains a risk (3).

Almost all Whipple procedures are done under general anaesthesia with or without epidural analgesia. In our unit thoracic epidural is routinely used for laparoscopic Whipple procedures. Evidence in epidural analgesia is limited in laparoscopic Whipple procedures, but it is superior to opioid only in accelerating the return of bowel functions and dietary intake while providing better pain relief (3). Timely administration of antibiotics reduces the surgical site infection and should be used in a single dose manner initiated 30-60 min before skin incision (13,14,15).

Serum bilirubin of this patient was four times of normal limit and the anaesthetists should be aware of the impact of the hyper bilirubinaemia on anaesthetic drugs. Prolong procedure, changing positions, pnumoperitonium and associated co morbidities make it necessary for close monitoring such as central vents pressure and intra arterial blood pressure.

Several meta-analyses and randomized control trials have demonstrated that preventing inadvertent hypothermia during major abdominal surgery reduces the prevalence of wound infections, cardiac complications, bleeding and transfusion requirements, as well as the duration of post anaesthetic recovery (16,17,18). Hence, the use of active warming is highly recommended to reduce postoperative morbidity and enhance recovery (16,17,18).

Laparoscopic procedure requires significantly less amount of fluids both

during and after the procedure. Excessive overload of salt and water in the peri operative period increases postoperative complication rates and delays the return of gastrointestinal function. (7,8,9). Near-zero fluid balance as well as avoiding overload of salt and water improves outcome. Perioperative monitoring of stroke volume with trans-oesophageal Doppler, LidCO to optimize cardiac output with fluid boluses improves outcomes (11). Balanced crystalloids should be preferred to 0.9% saline (12).

Close observation in the intensive care setting is mandatory to monitor for complications and provide adequate pain relief in early phase of the recovery. Somatostatin and its synthetic analogues (e.g., octreotide) reduce splanchnic blood flow and the release of pancreatic exocrine secretion (19).The rationale for its use is to reduce the risk of pancreatic anastomotic fistulas by decreasing the volume of pancreatic exocrine secretions. The most recent meta-analysis involved 17 trials with 1457 patients undergoing Whipple procedure and 686 undergoing distal or other resections. The authors concluded that the use of somatostatin analogues reduced the crude rate of pancreatic fistulas, but that the rate of clinically significant fistulas as well as the overall major morbidity and mortality remained unchanged (20).

CONCLUSION

There is a significant development of laparoscopic surgical skills in the recent past. Laparoscopic pancreatico-duodenecomy is one of the technically challenging procedure. However, immediate post operative outcome has significantly improved with laparoscopic procedure. Multidisciplinary team approach, pre operative optimization and optimum intra, post operative management can minimize the associated complications in these procedures.

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CORRESPONDENCE

Dr. W. M. A. S. B. Wasala
Faculty of Medicine,
University of Peradeniya.

E-mail- wasalaasb@yahoo.com

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Left Vertebral Artery Arising Directly from the Aortic Arch

H. A. Amaratunga[1], S. P. M. Peiris[2], H. R. Gunasena[1], J. K. Dissanayake[1]

ABSTRACT

Objective of this article is to report a case of the vertebral artery arising as the 3rd branch of the arch of the aorta and ascending up into the neck to enter the foramen transversarium of the 3rd cervical vertebra. This knowledge regarding variations is beneficial for planning aortic arch surgeries or endovascular interventions.

Key words: vertebral artery, variation

INTRODUCTION

The vertebral artery arises as the first branch of the subclavian artery at the root of the neck. In 6% of individuals the left vertebral artery may arise from the arch of the aorta itself (1). Such variations warrant mention due to its importance in thoracic surgeries and vascular investigations. Here we report a case of the left vertebral artery arising from the aortic arch detected during cadaveric dissections.

CASE REPORT

During a routine dissection procedure on a cadaver of an 83 year old male conducted at the Department of Anatomy, 4 branches were detected arising from the aortic arch (Figure 1). Further dissections revealed the first branch to be the right bracheocephalic trunk, the second to be the left common carotid artery, the third to be the left vertebral artery and the fourth the left subclavian artery. The left vertebral artery was further dissected to expose its course up into the neck. When the vertebral artery arises from the subclavian artery it enters the foramen transversarium of the 6thcervical vertebra (2). However in this case the vertebral artery travelled parallel to the left common carotid on its left and then turned posteriorly to enter the foramen of the 3rd cervical vertebra. No other abnormality of the blood vessels or the heart was detected in the cadaver.

The arch of the aorta can have 4 branches arising from it and the left vertebral artery can arise from this site. This knowledge regarding variations is beneficial for planning aortic arch surgeries or endovascular interventions.

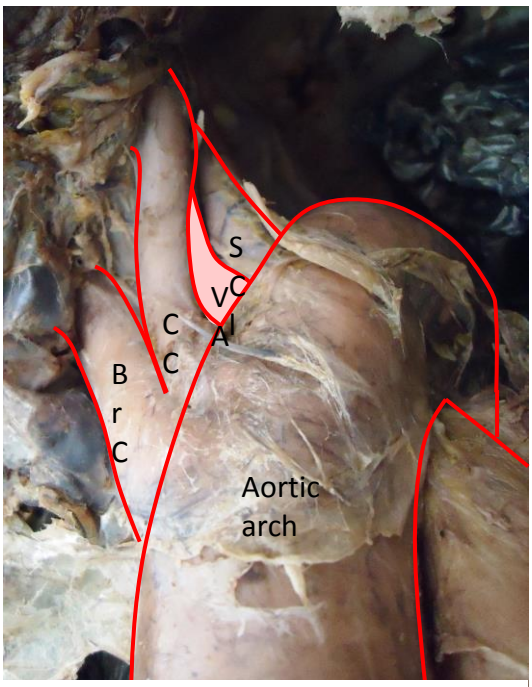

Figure 1: The four branches arising from the arch of the aorta. **BrC**– Brachiocephalic artery, **CC** – Common Carotid artery, **VA** – Vertebral artery, **SCl**- Subclavian artery

[1]Department of Anatomy, University of Peradeniya, Peradeniya, Sri Lanka.
[2]Department of Surgery, University of Peradeniya, Peradeniya, Sri Lanka.

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CORRESPONDENCE
Dr. H. A. Amaratunga
Department of Anatomy,
University of Peradeniya.

Email- himanipeiris@yahoo.com

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Men as Victims of Intimate Partner Violence - A Case Report

R. H. A. I. Rathnaweera[1]

ABSTRACT

Intimate partner violence (IPV) is an important worldwide problem. Several studies on IPV found equivalent rates of assault perpetrated by men and women. The victim was a 30 years old three wheel driver, married with two children. He was assaulted by his wife with a club over the head and was admitted to the Teaching Hospital Karapitiya. A CT scan of the brain revealed a left parieto-occipital large extra dural haemorrhage (EDH), left parietal skull fracture with midline shift of 2cm and cerebral oedema. Left tempero-parietal craniotomy and evacuation of EDH was done.

On examination, apart from the surgical incision, he had a sutured laceration 8cm in length over the left side of the head. Left sided 3rd cranial nerve palsy was also present. The idea of women being violent is a hard thing for many people to believe as it goes against the stereotype of the passive and helpless female. This, in spite of the fact that women are known to be more likely than men to commit child abuse and child murder. Findings of this case suggest that women use IPV against their male partners. Given the potentially serious physical and mental health consequences this can have, particularly for victims, research in this area needs to move beyond the argument over who perpetrates more IPV and who suffers more as a consequence of IPV.

Key words: intimate partner violence, male victims, female perpetrators, Sri Lanka

INTRODUCTION

Intimate partner violence (IPV) is an important worldwide problem (1,2). It is a type of domestic violence defined by the World Health Organization as " any behavior within an intimate relationship that causes physical, psychological or sexual harm to those in the relationship (1). The patriarchal model of the society supports the idea that IPV is a gender issue, perpetrated by men towards women (3,4). Meanwhile, several studies on IPV found equivalent rates of assault perpetrated by men and women (5,6).

CASE REPORT

The victim was a 30 years old three wheel driver, married with two children. For the last couple of months, wife started to suspect him regarding an extramarital relationship which he strongly refused. On 5th of December 2011 around 6.30pm, when he was sleeping on the floor (after a bout of alcohol), he felt severe pain over the left side of the head. When he raised his head, he saw his wife standing near him holding a club. When he shouted for help she ran away and he had an episode of loss of consciousness. By that time few neighbors gathered around the house and immediately, he was brought to the Teaching hospital, Karapitiya.

On admission to ETU, he had been semiconscious with GCS 08/15. He complained of severe pain over left side of the head. Immediate CT scan of brain was performed.

[1]Department of Forensic Medicine, Faculty of Medicine, Karapitiya, Galle.

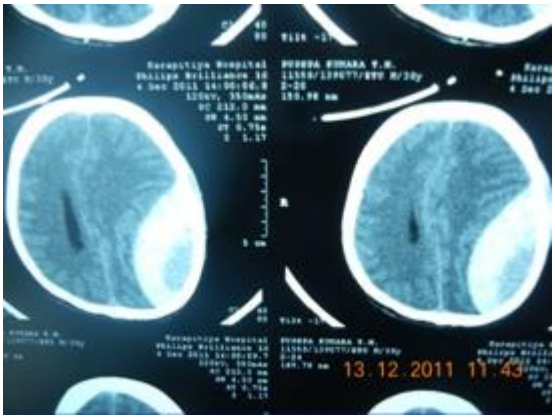

Figure 1: Left parieto-occipital extra dural haemorrhage (CT scan image)

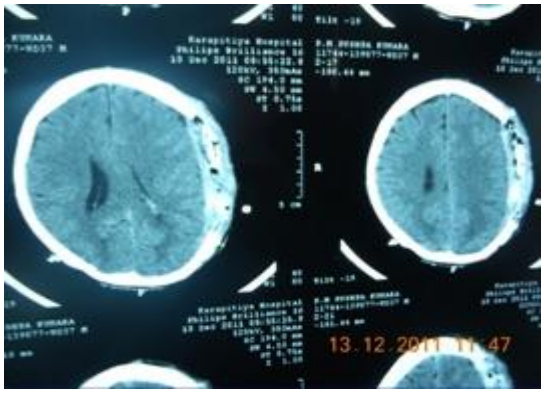

Figure 2: Repeat CT with extra dural haemorrhage and mid line shift (CT scan image)

It revealed (Figure 1), left parieto-occipital, large extra dural haemorrhage (EDH) (thickness 4cm), left parietal skull fracture, midline shift of 2cm to the right side and cerebral oedema. Ipsilateral lateral ventricle compressed. Left tempero-parietal craniotomy and evacuation of EDH was done. On 6/12/2012, repeat CT done and it revealed (Figure 2) an EDH of 3x8cm with mid line shift of 7cm. Re-evacuation of EDH done on the same day. After that he recovered gradually.

On examination, apart from the surgical incision, he had a sutured laceration 8cm in length over the left side of the head, obliquely placed, lower end is placed 6cm from midline and 9cm from top of the head (Figure 3). Left sided 3rd cranial nerve palsy was also present (Figure 4). He was discharged 14 days after the incident.

Figure 3: **Sutured laceration over the left side of the head**

Figure 4: Left sided 3rd cranial nerve palsy

DISCUSSION

In the case under discussion, the victim had a fresh laceration with a large underlying EDH. No other injuries present over any part of the body. According to the history he was assaulted while he was lying down, using a club. The injury pattern is compatible with such history. The other two possibilities were either a fall or RTA. In both of these situations, it is highly unlikely to receive a single laceration over the head. The circumstantial evidences also are supportive for the history given by the victim. The fresh injuries suffered by this victim are; laceration over the head, skull fracture and extra dural haemorrhage with

mid line shift. In this case, emergency craniotomy was performed to evacuate the EDH. Unless prompt and proper care was given, this patient would die. So the category of hurt would be " fatal in the ordinary cause of nature" . Assailant can be charged for attempted murder.

In 1977, Suzanne Steinmetz released results from several studies showing that the percentage of wives who have used physical violence is higher than the percentage of husbands, and that the wives' average violence score tended to be higher, although men were somewhat more likely to cause greater injury. She also found that women were as likely as men to initiate physical violence, and that they had similar motives for their violent acts. Steinmetz concluded that "the most unreported crime is not wife beating - it's husband beating" (7).

The idea of women being violent is a hard thing for many people to believe. It goes against the stereotype of the passive and helpless female. This, in spite of the fact that women are known to be more likely than men to commit child abuse and child murder (8).

In 1988, an investigation of spousal homicide between 1978 and 1982 found that 7.8% of murder victims were husbands murdered by wives, and 8% were wives murdered by husbands (9).

More recently, in a study of spousal homicide in the period from 1986 to 1995, it was found that there was an overall ratio of 1.3:1.0 of murdered wives to murdered husbands (10).

In 1996, for instance, in a critique of the Curtis report, Wilt & Bannon wrote that "nonfatal violence committed by women against men is less likely to be reported to the police than is violence by men against women; thus, women assaulters who come to the attention of the police are likely to be those who have produced a fatal result" (10).

Results like these are greeted with great suspicion by those who see domestic violence as a political issue to be exploited rather than a social problem to be solved. In a book on domestic violence, Roger Langley and Richard C. Levy conclude a chapter on battered husbands by saying, "Husband abuse should not be viewed as merely the opposite side of the coin to wife abuse. Both are part of the same problem, which should be described as a one person abusing another person. This problem must be faced and dealt with not in terms of sex but in terms of humanity" (11).

CONCLUSION

Severity of the injuries received by the victim in this case was very significant when compared to the cases reported in other countries. Findings of this case suggest that women use IPV against their male partners. Given the potentially serious physical and mental health consequences this can have, particularly for victims, there are compelling reasons why research in this area needs to move beyond the argument over who perpetrates more IPV and who suffers more as a consequence of IPV.

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CORRESPONDENCE
Dr. R. H. A. I. Rathnaweera
Department of Forensic Medicine,
University of Ruhuna, Sri Lanka.

E mail-ajithrathnaweera@gmail.com

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An Omphalocele, Epispedias, Cleft Palate, Cranial deformity and Facial Dysmorphism: A Case with Midline and Laterality Defects

L. B. L. Prabodha[1], T. S. D. Amarasena[2], I. Ilayperuma[1], B. G. Nanayakkara[1]

ABSTRACT

A 7 year old male child with cleft soft palate, omphalocele, epispedias, posterior prominence of the skull, prominent forehead with high anterior hair line, dextraposition of the heart, right sided inguinal hernia, mental retardation, generalized hypotonia and flexion deformity of both toes and fingers presented to the paediatric clinic, Teaching Hospital Karapitiya, for the follow up management. Furthermore, the child had subtle dysmorphic features including, broad nasal bridge, hypertelorism and low set ears. He was the second child of the family and there were no other family history of congenital anomalies. The karyotype was 46XY. Mutations in chromosome bands 3p12-21, *ZIC3* gene in human X chromosome and Wolf-Hirschhorn syndrome involving heterozygous deletion of 4p16.3 region (4p syndrome) can be presented with above clinical features and it is necessary to investigate the patient further for the genetic involvement.

Key words: omphalocele, epispedias, cleft palate, cranial deformity, facial dysmorphism, *ZIC3* gene, 4p syndrome

INTRODUCTION

Disturbances of the normal asymmetric position of organs or situs inversus, have been described as laterality defects. Congenital anomalies of midline structures such as, esophageal defects, anal defects, neural tube defects, cleft lip and palate are considered as midline defects. Animal model studies and human birth defect registries suggest that the midline and laterality defects are etiologically related (1).

All vertebrates have a body configuration with a midline and left and right halves that are symmetric for most external features. Many internal organs have asymmetric placement in the body with respect to the midline. The heart is positioned in the left side of the thorax with its apex pointed towards the left. The spleen and stomach are placed on the left side, liver in the right side of the abdomen. Alterations of the left/right (LR) asymmetry may cause alteration of the position of organs (heterotaxia or situs ambiguous) or they may produce reversal of LR organ position (situs inversus). Disturbances of LR asymmetry may also be limited to a single asymmetric organ such as the heart in individuals with isolated dextrocardia or dextropositon of the heart. Collectively, any abnormalities of LR asymmetry are referred to as a laterality defect (1). It has been recognized that there is a genetic basis for some disturbances of LR asymmetry in humans. Although most cases are considered sporadic, families with laterality defects have been shown to have an autosomal dominant, autosomal recessive and X-linked recessive inheritance patterns. Thus,

[1]Molecular Genetics Laboratory, Department of Anatomy, University of Ruhuna.
[2]Department of Paediatrics, University of Ruhuna.

multiple genes exist that are responsible for human laterality defects, although mutations of single genes may cause laterality defects that can be seen as Mendelian patterns (2,3). Genes corresponding to the chromosome bands 3p12-3p21 has been described in the literature causing midline and laterality defects. Furthermore, mutations in *ZIC3* gene in human X chromosome have been identified in causing midline and laterality defects (4).

There is clear evidence that model animals with disturbances of LR asymmetry to have midline defects more commonly than expected by chance. Some of these defects include neural tube defects, oral clefts, omphalocele, esophageal atresia /tracheoesophageal fistula, imperforate anus, epispedias conotruncal heart defects, diaphragmatic hernia (5).

Four families with a distinctive malformation syndrome which includes diaphragmatic defects, limb deficiencies and ossification defects of the Skull were also reported (6).

Wolf-Hirschhorn syndrome (WHS) is characterized by typical craniofacial features in infancy consisting of wide bridge of the nose continuing to the forehead, microcephaly, high anterior hairline with prominent glabella, widely spaced eyes, epicanthic folds, high arched eyebrows, short philtrum, downturned corners of the mouth, micrognathia, and poorly formed ears with pits and tags. The diagnosis of Wolf-Hirschhorn syndrome is established by detection of a heterozygous deletion of the Wolf-Hirschhorn syndrome critical region (*WHSCR*) within 4p16.3 at 1.4-1.9 Mb from the terminus (7).

CASE REPORT

Index case was a 7 year old male child with cleft soft palate, Omphalocele, epispedias, right sided inguinal hernia, posterior prominence of the skull, prominent forehead with high anterior hair line, mental retardation, hypotonia, dextraposition of the heart, flexion deformity of both toes and fingers. Furthermore, he has having subtle dysmorphic features such as, broad nasal bridge, hypertelorism and low set ears. He was born at term with a birth weight of 2.4kg. He was presented with global developmental delay. No other respiratory or gastro intestinal defects. He was the second child of the family with no other family history of congenital anomalies. His elder brother was apparently normal.

Echo cardiogram of the child reveled dextraposition of the heart and no other congenital cardiac anomalies were present. Ultra sound scan of the abdomen revealed normal. His karyotype was 46XY.

The following differential diagnoses were made:

1) Mutations in chromosome bands 3p12-21 causing midline and laterality defects.
2) Mutations in ZIC3 gene in human X chromosome causing midline and laterality defects.
3) Wolf-Hirschhorn syndrome involving heterozygous deletion of 4p16.3 region (4p syndrome).

The omphalocele, cleft palate and epispediasis were treated by the Pediatric and the Maxio-Facial surgeons at the Teaching Hospital Karapitiya. The child was followed up in the Rheumatology clinic and the Physiotherapy department for his rehabilitation.

DISCUSSION AND CONCLUSION

According to the reported literature there is clear evidence that midline and laterality defects occur together more commonly than expected by chance (5). The midline defects include neural tube defects, oral clefts, omphalocele, esophageal atresia/ tracheoesophageal fistula, imperforate anus, epispedias, conotruncal heart defects, diaphragmatic hernia whereas the laterality defects include dextraposition of heart, malrotation of gut and other dextraposition of viscera such as right sided spleen (5). It was stated that the mutations in chromosome bands 3p12-21 causing midline and laterality defects associate together more frequently. Furthermore, it was reported that mutations in *ZIC3* gene in human X chromosome causing midline and laterality defects in some families (1).

The 4p syndrome or Wolf-Hirschhorn syndrome is characterized by typical craniofacial features and other abnormalities in infancy (7).

The index case also has shown some clinical features such as, dextraposition of the heart, omphalocele, epispedias, congenital inguinal hernia, cleft palate, facial dysmorphism and cranial anomalies, in favor of the above mentioned differential diagnosis. Furthermore, his karyotyping was 46 XY.

Molecular genetics investigations are important in this case for diagnosing the condition and to identify the etiology of the anomalies.

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CORRESPONDENCE

Dr. L. B. L. Prabodha
Molecular Genetics Laboratory,
Department of Anatomy,
University of Ruhuna, Sri Lanka.

E mail- lahiruprabodha@gmail.com

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