Successful surgical separation of conjoined twins: First experience in Rwanda

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Conjoined twins are identical or monozygotic twins whose bodies are joined in utero. Pygopagus or illopagus twins are a type of conjoined twins in which two bodies joined back to back at the buttocks. Surgical Separation of conjoined twins is extremely risk of death and life threatening. Female pygopagus twins of three months were been operated and separated at Kigali Teaching University Hospital. For both babies a posterior sagittal anorectoplasty was performed with derivated ileostomy without problem. No complications occurred during the operation, oral feedings was done at third postoperative day. Ileostomy closure was done three weeks after and babies were discharged from neonatology unit at 35th postoperative day. Adequate preoperative investigation with a well organized and trained team contributed a lot to the success of conjoined twins separation.

Introduction

Conjoined twins are one of the rarest and most challenging congenital malformations arise as an unfortunate complication of monozygous twinning with an estimated incidence ranging from 1 in 50,000 to 100,000 pregnancies, but around 60% of them are stillborns, giving an overall true incidence of about 1 in 200,000 live births with a male-female ratio of 1:3. Conjoined twinning arises when the twinning event occurs at about the primitive streak stage of development, at about 13-14 days after fertilization and is exclusively associated with the monoamniotic monochorionic type of placentation. Two contradicting theories exist to explain the origins of CT. The older and the most generally accepted is fission, in which there is an incomplete splitting of the embryonic axis and, with the exception of parasitic conjoined twins, all are symmetrical and the same parts are always united to the same parts. The second theory is fusion, in which a zygote completely separates, but stem cells find like-stem cells on the other twin and fuse the twins together.

The incidence of the various types of CT is discussed and they are classified according to the most prominent site of attachment. Information from the largest study to date indicates that the most common encountered were thoraco-omphalopagus (28%), thoracopagus (18%), omphalopagus (10%), parasitic twins (10%) and craniopagus (6%). Other less common types of CT include: Pygopagus, Cephalopagus, Xiphopagus, Ischiopagus and Parapagus.

Pygopagus twins are joined back to back facing away from each other, commonly share the gluteal region, terminal spine, and lower gastrointestinal, urological, and reproductive tracts and a recent reported study found that they represented 17% of all conjoined twins. The first recorded pygopagus conjoined twins dates back to 1100. They lived 34 years joined together at the hips and buttocks and shared a vagina and an anus, but when one of them died,
the other refused to be separated and died 6 hours later. The prognosis of pygopagus twins is generally good with an overall survival rate of 87%, but they are also known to have associated vertebral, spinal cord, as well as anorectal anomalies that form a spectrum that differs between males and females 5,9,10.

We report the first successful surgical separation of conjoined twins “pygopagus” type in Rwanda.

Case report

Female pygopagus twins (Figure 1) were delivered by cesarean section at 37 weeks gestation at a district hospital. They were transferred to our neonatal unit immediately after birth. They had a combined weight of 4 kg. They were joined back to back at the buttocks, and perineal areas. They share one anus and two fused genitalia (Figure 2). The physical examination showed hypotrophic newbones but the assessment of the central nervous system, heart, and lungs revealed no abnormalities. The examination of the perineum showed two separated urogenital tracts with a thin membrane which constitute the posterior wall of the vaginal canals.

**Figure 1.** Clinical photograph showing female pygopagus twins joined back to back facing away from each other with attachments at the buttocks, and perineum.

**Figure 2.** This radiograph shows two fused genitalia with one anus

**Figure 3.** Plain radiograph showing two separate spinal columns.

**Figure 4.** Lower gastrointestinal tract studies by barium enema showed two separate rectums with a distal fusion in Y configuration
Plain radiograph of the spine showed the sacra to be separate (Figure 3). There was no evidence of any other vertebral anomalies. Lower gastrointestinal tract studies by barium enema showed two separate rectums with a distal fusion in Y configuration (Figure 4). They tolerated oral feeds and started to gain weight, so it was decided to delay separation until their combined weight is greater than 5 kg. After two months the babies weighted 7.5 kg and the separation was decided. Before surgery, two teams of surgeons with the team of anesthesiologists and operating room nursing staff met to discuss the operation procedure, the management of the team and the theatre room.

The operation started by separating the soft tissues on the posterior gluteal region and we found that the vertebral column was fused distally at the level of the coccyx by a cartilaginous and fibrous tissues which were easily divided. The two slightly fused rectums were separated and vaginas were opened and repaired individually. For both babies a posterior sagittal anorectoplasty was performed with derivated ileostomy. After perineal reconstruction, the skin was closed primarily using the already raised V-shaped skin flaps. No skin grafting was needed. The operation lasted 3 hours, and postoperatively, they were briefly ventilated in the recovery room.

Twenty four hours later, they were transferred to the neonatal unit and started oral feeding on the third postoperative day. No postoperative complications were observed and the perineal wound healed without any problem and the ileostomy closure was performed after three weeks and the babies were discharged home on the 35th postoperative day with a weight of 4.250kg for baby A (Figure 5A) and 3.700kg for baby B (Figure 5B). The follow-up 1 year later, they were found to be healthy and thriving normally without any residual problem.

**Discussion**

Conjoined twinning is one of the most fascinating human malformations and has also been reported in other animals especially mammals, fishes, birds, reptiles, and amphibians. The cause of conjoined twins is not exactly known, but it is generally accepted that conjoined twins arise from a single zygote that fails to undergo complete splitting of two inner mass cells during the blastocyst period (5-6 days after fertilization). The conjoined twins arise also from
the incomplete splitting of the inner mass cell or embryoblast while the embryo is undergoing the hatching from the zona pellucida at day 6 after fertilization. In rare cases conjoined twins may result from an incomplete separation of the embryonic disc after 12 days of embryogenesis before the gastrulation begins\(^1,3,4\). This, as well as the classification, management, and prognosis of conjoined twins, is extensively reviewed by Spencer\(^12\).

Treating conjoined twins can be a daunting challenge for the surgeon. Furthermore, these cases often raise religious, moral, ethical and legal issues\(^13,14\). The diagnosis of conjoined twins can be made prenatally in centers where pregnant mothers are subjected to routine ultrasonography as early as 12 weeks’ gestation\(^15\). The diagnosis, however, can be missed on occasions as what happened in our patient. The importance of preoperative diagnosis needs to be emphasized because conjoined twins should be transferred in utero to a center where future management is feasible.

Surgery to separate conjoined twins may range from relatively simple to extremely complex, depending on the point and complexity of attachment and the internal organs which are shared. Most cases of separation are extremely risky and life-threatening. In many cases, the surgery results in the death of one or both of the twins, particularly if they are joined at the head. Of all types of conjoined twins, omphalopagus twins are the most favorable candidates for elective surgery because of good survival rates\(^16\).

Emergent conditions may arise at any time and include intestinal obstruction, rupture of an omphalocele, congestive cardiac failure, severe degree of respiratory compromise, and terminal illness in one of the twins.

The first successful separation of conjoined twins was performed in 1689 by Johannes Fatio. The survival rate of conjoined twins, however, correlates with age at separation. It was less than 50% if surgical separation was attempted in the neonatal period, but increased to 90% if separation was delayed until 6 months of age or later\(^17\). In this case separation was performed when twins were 3 months because we consider that the babies were gaining weight and able to undergo the operation. The success of the separation depends on the case but the overall survival rate of 64% was quoted by Hoyle\(^8\) in 1990. Thoracopagus, craniopagus, and omphalopagus were associated with the highest mortality rate (51%, 48%, and 32%, respectively), whereas lower mortality rates occurred with ischiopagus (19%) and pygopagus (23%) twins\(^8\).

Pygopagus twins are very rare, accounting for about 10% to 18% of all conjoined twins, which gives an incidence of about 1 in 1,000,000 live births of this particular type. They commonly share the gluteal region, terminal spine, and gastrointestinal, urological, and reproductive systems to variable degrees. About 50% of pygopagus twins have anomalies unrelated to the classic fused organs including a high incidence of vertebral anomalies\(^5,9\). Our patients had two separate rectums but fused distally in a “Y” configuration with one anal orifice, two separate urological systems, two sacra fused at the coccyx by a small band of cartilage and no spinal cord fusion or other vertebral anomalies.

The surgical management of pygopagus twins necessitates detailed radiographic examination of all urinary, reproductive, and gastrointestinal systems. Magnetic resonance imaging should form part of the preoperative investigations of all pygopagus twins with particular attention to the anatomy of the spinal cord. Because of limited investigation facilities in our setting we
performed on our patients just radiography to see the aspect of the rectums and the distal column and ultrasound to look for other associated malformations; IMR was not on our reach to see the anatomy of spinal cord but fortunately no spinal fusion was found during operation. The overall survival rate of pygopagus twins was 87%, and for males it was 100%, whereas for females it was 85% 5. Half of the twins had nonfused rectums, and half had fused rectums. The nonfused had 2 rectums (80%) or one rectum and one rectovaginal fistula (20%). The fused had high (46%) or low (54%) anorectal “Y” junction. All reported living male pygopagus twins have had nonfused rectums. All can be managed applying the principles of posterior sagittal anorectoplasty 5,3.

In this case the twins were female and they had two rectums with a lower junction with one anal orifice and the anorectal malformation was managed by posterior sagittal anorectoplasty protected by an ileostomy. The separation of our patients was successful and the babies were discharged 1 month after in good condition without any sphincter troubles or motor deficits.

Conclusion

In conclusion, adequate preoperative investigations, a team approach, accurate operative techniques and good postoperative care contribute to the success of conjoined twins separation in general and pygopagus twins separation in particular.

References

HOW I DO IT: Technique of Hepatectomy for Primary Liver Cell Carcinoma in a Developing Country.

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The experience in managing a 36 year-old Nigerian male with a histopathologically-confirmed hepatocellular carcinoma confined to the left lobe by surgical resection of the liver, using the finger-fracture technique is presented. The literature review on the aetiopathogenesis of this disease and the role and technique of surgery in hepatocellular carcinoma is also discussed.

Case Report

A 36 year-old lecturer from the University of Maiduguri in the Bornu State of Nigeria was referred to the surgical outpatients department of the University College Hospital Ibadan with a 5-week history of a painful but progressively increasing epigastric swelling. There was no associated nausea and vomiting, no weight loss, no early satiety and no haematemesis or melaena.

There was no previous history of jaundice or of a blood transfusion. He stopped smoking cigarettes 10 years before presentation after having smoked 2-3 cigarettes daily for about 9 years. He however claimed to have a 16-year history of alcohol ingestion of about 3 bottles of beer per day. There was no previous hospitalization and he was not on treatment for any intercurrent disease.

Physical examination revealed a well-built adult male who was not pale, not jaundiced and not chronically ill-looking. He had no stigmata of chronic liver disease. Significant findings were on abdominal examination where an epigastric mass was found; measuring 10 by 8 cm. The overlying skin was normal; the mass was firm and non-tender with a smooth surface. One could get below but not above it, it was not pulsatile, dull to percussion, no bruit or venous hum heard over it on auscultation and there was no demonstrable ascites.

A diagnosis of primary liver cell carcinoma was made and confirmed with fine-needle aspiration cytology. Abdominal ultrasound scan showed that he had a 6.1cm solid mass in the left lobe of the liver. The right lobe was said to be free of masses. His packed cell volume was 36%, white blood cell count was 3000/cmm, and total bilirubin was 3.8mg% with a conjugated fraction of 1.5mg%. Alkaline phosphatase was 544 iu/ml, SGOT 254 iu/ml, SGPT 156 iu/ml and a serum albumin of 3.9 g%. His clotting profile showed a derangement in the PTTK only which revealed a 37-second difference from the control. Urinalysis did not reveal any glucose, ketones or bilirubin, however the ph was 6.0, urobilinogen was increased and there was significant proteinuria (+).

Preoperative treatment with fresh whole blood and fresh frozen plasma improved his PTTK to a 16-second differential with the control. He was scheduled for exploratory laparotomy as a Grade A Childs classification patient. Four units of fresh whole blood and 10 units of fresh frozen plasma were arranged for surgery. At surgery, under light general anesthesia and epidural analgesia, the peritoneal cavity was entered through a bilateral sub-costal (bucket-handle) incision and a quick exploration was carried out. There was a single, bilobed tumor occupying almost the whole of the left hepatic lobe; the larger part measuring about 10 by 10cm
while the smaller part was 5 by 5cm. the right lobe looked and felt grossly normal. There was no ascites or peritoneal involvement attributable to the tumor. Other intra-abdominal organs were grossly normal.

Using the Falciform ligament as the line of demarcation, fracturing of the liver parenchyma between finger and thumb was commenced. This effectively and gently exposed arterial branches, venous radicles and bile ductules which were clamped individually with number 1 or number 2 hemostats, severed and ligated with number 2-0 chromic catgut suture. This progressed slowly but definitely until the entire lobe containing the tumor had been excised. This part was then freed totally by incising the triangular ligament attaching the lobe to the left hemi-diaphragm. The estimated blood loss was 500 mls. Two drains were inserted; one sub-diaphragmatic and the other sub-hepatic to exit through separate stab wounds. The weight of the resected hepatic lobe containing the tumor was 453 grams.

His post-operative period was uneventful. Histopathology of the resected specimen showed a well-differentiated hepatocellular carcinoma with background active macro-nodular cirrhosis. He was discharged for outpatient adjuvant cytotoxic chemotherapy in the radio-oncology unit where he had 4 courses of Cyclophosphamide, Adriamycin and 5-Fluorouracil (CAF) chemotherapy. A routine follow-up liver ultrasound scan performed 3 months after discharge showed a new solid mass in the right lobe of the liver. The patient looked well and claimed he felt well at that time. However he told that further surgery would be futile but we will continue to follow up at the outpatients’ clinic for supportive treatment. He did not come back. Later, we received a letter from his elder brother who regretted to inform us that he passed away suddenly on a date approximately 9 months after his operation.

Discussion

Primary liver cell carcinoma (PLCC) affects young middle-aged males commonly in Africa and Asia\(^1\,^2\,^3\). This is attributed to the association with hepatitis B, of which the presence of its surface antigen is common in these populations in up to 80% of cases\(^1\,^2\,^3\). Maiduguri is a city in the Sahel savannah near Lake Chad in Nigeria where a previous study showed that 84.4% of controls had evidence of hepatitis B virus infection. PLCC typically arises in a liver that has been subjected to chronic stimulation, usually by environmental or biologic toxins that result in hepatocellular death, chronic regeneration and cirrhosis\(^4\). The most common causes are hepatitis B, hepatitis C and exposure to hepatotoxins notably aflatoxin B1 and ethanol\(^1\,^2\,^3\,^4\). It is also known that 60-80% of PLCC are in livers with pre-existing cirrhosis\(^4\).

Surgical resection is the mainstay of treatment of PLCC and, besides transplantation, the only definitive treatment\(^4\,^5\,^6\,^7\,^8\). Untreated the outlook is poor as the overall survival of such untreated cases is about 3 to 4 months after symptoms appear\(^5\,^8\). The main hazard after resection of hepatocellular carcinoma is intrahepatic recurrence, with reported rates between 50-100% within 1 to 2 years of operation\(^6\,^7\,^8\). This recurrence is said to be higher in tumors greater than 5cm, unencapsulated and of multicentric origin (e.g. PLCC in cirrhotic patients)\(^6\,^8\). It would seem that our patient had adverse factors for recurrence coupled with the fact that he was domiciled in an area with a high prevalence of hepatitis B infection and high aflatoxin load of the common foodstuff (grains are staple food in Maiduguri)\(^1\,^2\). Anicteric transmission of hepatitis B has been shown to occur frequently and is more important for hepatocarcinogenesis\(^1\,^3\), thus failure to elicit a history of previous jaundice in a patient should not rule out hepatitis B infection as a precursor of PLCC. The patients who may benefit from hepatic resection are those whose hepatic biochemical test results do not show significant derangement\(^6\,^8\). The Childs-Pugh classification is popularly used in liver disease to select...
patients who may be fit for different types of operations with A being ideal and C poor\textsuperscript{5}. Some authors have reserved liver transplantation for the B and C grades\textsuperscript{6,8}.

A satisfactory access for liver operations is usually gained with a bilateral sub-costal incision which was used in this patient\textsuperscript{9}. Generally four types of major hepatic resection are commonly employed, based on the lobar and segmental anatomy of the liver; right hepatic lobectomy, left hepatic lobectomy, right trisegmentectomy and left lateral segmentectomy\textsuperscript{5,9}. A left hepatic lobectomy was performed in this patient because at surgery the involvement was grossly of the left hepatic lobe only. Parenchymal dissection may be done with finger-fracture or the handle of the scalpel, and the biliary-vascular bundles are ligated after being clamped and divided\textsuperscript{5}. Larger centers may use other sophisticated equipment like the ultrasonic aspirator scalpel, the harmonic scalpel or even laser to dissect the hepatic parenchyma\textsuperscript{5}. Non-resective treatment may be achieved with the aid of selective transhepatic arterial or portal venous embolization\textsuperscript{10}. In a low socio-economic tropical West African center, these gadgets are not available, thus finger-fracture dissection has to be the option if hepatic resection is to be performed. The tactile feedback with this procedure ensures sensitivity and gentle tissue handling. It is no doubt slower than the afore-mentioned innovations, but is effective since about a third of the patient’s liver (453 grams) was resected with minimum blood loss. It is possible that this procedure bought the patient an extra 9 months in which to tidy up his affairs before his demise.

References

Pleuropulmonary Blastoma: Case Report

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Pleuropulmonary blastoma (PPB) is a rare and aggressive tumor that is emerging as a distinct entity of early childhood disease. It is characterized by mesenchymal elements (including undifferentiated blastoma and often cartilaginous, rhabdomyoblastic, or fibroblastic differentiation) and epithelium-lined spaces. The tumor arises in the lung and pleura and is regarded as a pulmonary dysontogenetic or embryonic neoplasm. It is the pulmonary analog of other tumors of childhood including Wilms’ tumor, Neuroblastoma, Hepatoblastoma, Pancreatoblastoma and Retinoblastoma. Due to their protean presentation it is often difficult to make a preoperative diagnosis. A high index of suspicion therefore is needed. As a result these are diagnosed late, and these, along with other factors, affect the eventual outcome. We report a case of Pleuropulmonary blastoma diagnosed after the child was operated as a case of massive left hemothorax following blunt trauma.

Introduction

Pleuropulmonary blastoma (PPB) is a rare and highly aggressive intrathoracic malignancy in childhood and less than 100 cases have been reported in the literature. In 1961, Spencer first used the term and suggested that PPB arose from mesodermal blastoma because of its similarities to nephroblastoma. In the year 1988, Manivel et al. described PPB in children as an entity that was distinct from the biphasic epithelial stromal morphology of the classic adult type. Unlike pulmonary blastoma, PPB lacks the malignant epithelial component and entirely consists of primitive blastoma showing varying levels of sarcomatous differentiation (1,2). We present here a case of this rare tumor.

Case Report

A three and a half years old female child, the 3rd child and a twin, was presented to our hospital with the complaint of left sided chest pain and shortness of breath of 3 weeks duration following a fall accident while playing and was treated in one of the rural hospitals as a case of community acquired pneumonia and later on chest tube insertion revealed hemorrhagic effusion and was subsequently referred to our hospital. On presentation she was in respiratory distress with RR of 55/min, PR of 144/min, Spo₂ of 68% with Atm O₂ and standard weight and height for her age. Chest examination revealed respiratory distress with subcostal and intercostal retractions, tracheal shift to the right side, and absent air entry with dullness on the left side. Initial labs revealed WBC of 14,100 (71.7% neut), HCT of 22.8% and Hgb. of 8.8 gm/dl, ESR of 85 mm/hr, RBS of 108 gm/dl and normal RFT. Pleural tap revealed frank blood and chest x-ray (fig.1) revealed massive left pleural effusion with shift of the mediastinum to the right and with the assessment of left massive haemothorax and possibly empyema she was admitted, put on IV antibiotics and left chest tube inserted and clotted blood came out, transfused with whole blood and HCT rose to 36%. The patient was operated through left thoracotomy and severely lacerated left lung with a solid tumor between the upper and lower lobes with massive clotted haemothorax found and hematoma evacuated, left lung removed en block with the tumor, chest tube placed and chest cavity closed. The patient was subsequently discharged improved on her 18th postoperative day. Unfortunately she returned back two weeks after discharge with a
fungating and infected mass on the previous surgical incision site with empyema and sepsis treated in the hospital with IV antibiotics and chest drain producing thick organized pus and succumbed after one week of stay in the hospital.

**Figure 1.** Chest x-ray of the child after recurrence of the tumor on the chest wall (arrow)

**Pathologic findings**

**Gross Appearance**

Lung tissue (2 lobes) with a 5 cm mass attached to the lung by a stalk. Cut surface: gray white solid, surrounded by a small rim of lung parenchyma, invasive border. (fig 2).

**Figure 2.** Gross appearance of the lesion: cut gray-white solid, invasive border.

**Microscopic Appearance**

Three distinct patterns:

1) Dominant is a loose network of spindle cells, moderate nuclear pleomorphism and hyperchromasia.
2) Sheets and nests of undifferentiated (blastomatous) cells, hyperchromatic nuclei, some with nucleoli.
3) Scattered glandular structures, cuboidal to columnar epithelium. (fig 3 A-D)
Discussion

Pleuropulmonary blastoma is a primary intrathoracic malignancy that occurs mainly in early childhood. This dysontogenetic neoplasm, an analog to the other unique childhood tumors like Wilms’ tumor and neuroblastoma, is classified with the “mesenchymal neoplasms” in the WHO Classification of Lung Tumors. There it is composed of immature mesenchyme, often differentiating toward skeletal muscle, cartilage, fibrous tissue, and sometimes fat, and most often includes epithelium. The mesenchymal elements are regarded as malignant.

Since PPB was recognized as clinicopathologic entity distinct from adult pulmonary blastoma, which is characterized by malignant glands and malignant stroma, the epithelial elements in PPB have been described as benign. In the past, they have been termed pulmonary sarcoma arising in mesenchymal cystic hamartoma, embryonal sarcoma, or rhabdomyosarcoma arising in congenital cystic adenomatoid malformation or bronchogenic cysts. The age onset of presentation was between two weeks to 96 months. Although respiratory difficulty with or without fever is the most common clinical symptom, PPB can present with spontaneous pneumothorax, or empyema. There are few reports of bilateral PPB.

A significant feature of patients with PPB is the extraordinary high prevalence of other tumors in close relatives, which has been reported to be as high as 25%. In general, there are no characteristic findings on imaging studies. Dehner proposed a classification scheme for PPB that divides these lesions into predominantly cystic (type I), cystic and solid (type II), and...
predominantly solid (type III) types. Each type is characterized by increasing histologic evidence of malignancy\textsuperscript{10}. There is a report in which the progression of PPB from type I to type III has been documented over the time\textsuperscript{11}. Grossly, cystic tumors are single or multiloculated and may show nodular, thickened walls or pedunculated nodules. Solid tumors are multi lobulated white-gray with focal hemorrhage\textsuperscript{3}. Microscopically cystic lesions consist of one or more spaces lined by benign alveolar or ciliated columnar epithelial cells, beneath them, there is a layer of primitive oval and spindled rhabdomyoblasts in a loose or dense fibrovascular stroma. Solid tumors consist of blastomal stromal cells, arranged in alternating bands of compact and loose cells in myxoid matrix. The three pathologic types are correlated with both age at diagnosis and clinical outcome. Type I occurs in infants (median diagnosis age, 10 months) in contrast to types II and III (median diagnosis ages, 34 and 44 months, respectively)\textsuperscript{4} Type I PPB has been identified in utero\textsuperscript{18}. If PPB recurs in an individual patient, the type has often progressed to more advanced disease\textsuperscript{4,10}. The diagnosis is made only on histologic evaluation of the excised mass; however, fine-needle aspiration cytology has been used to diagnose it\textsuperscript{13,14,15,16}. Immunohistochemical staining mirrors a range of differentiation, with vimentin, histiocytic markers, or myoid antigens being common\textsuperscript{3,12}.

The rarity of PPB has allowed only slow elucidation of its clinical features according to prognosis and its response to therapy\textsuperscript{20}. The treatment is primarily complete excision of the tumor\textsuperscript{21,22}, followed by intense chemotherapy\textsuperscript{23}. Although there is disagreement in the literature, local radiotherapy also has been applied to PPB\textsuperscript{1}. Metastatic spread can also affect the ipsilateral lobes of the lung, the central nervous system including the spinal cord, and skeletal system. The prognosis depends largely on the staging at the time of diagnosis and the grading of the sarcomatous elements, but in general these are aggressive neoplasms with a 5-year survival probability of less than 50% of the cases with a solid component\textsuperscript{24}. Type I PPB is characterized by subtle malignant changes and a good prognosis. Recurrences after type I PPB are usually advanced type II or type III neoplasms with a poor prognosis. It has also been suggested that “extrapulmonary” involvement in PPB, defined as involvement of “the pleura, diaphragm or mediastinum,” indicates a less favorable prognosis\textsuperscript{19}.

References

Midline or Retroperitoneal Approach in Operating on a Retroperitoneal Tumour: A Surgical Dilemma.

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Retroperitoneal tumors are rare form of tumors arising from the retroperitoneal space and account for 10-15% of soft tissue sarcoma.\textsuperscript{1,2} Liposarcoma represent 0.1% of all cancers\textsuperscript{3} and arise mostly from the mesenchyme usually located in muscles, fat and connective tissue.\textsuperscript{4} It is a high grade tumor with high propensity for recurrence.\textsuperscript{1} 80% of patients present with asymptomatic abdominal distension in their 5\textsuperscript{th} - 6\textsuperscript{th} decade of life.\textsuperscript{4} Liposarcoma tend to present as a huge mass posing a diagnostic and therapeutic dilemma especially with regards to its position and surgical approach.\textsuperscript{1}

Case Report

We present a 64 years old man with one year history of progressive abdominal distension and rapid increase in size of 3 months prior to presentation. He lost 20kg in 4 months. He had no history of jaundice or altered bowel habit. He smoked for thirty years during his youthful days in the army. General examination was not remarkable. He had an abdominal mass of 10x8x6cm in the left flank, no hepatosplenomegaly. Abdominal ultrasound showed a mass of 18.8cm on the left flank extending into the left iliac fossa with bilateral mild hydronephrosis. CT scan (Figure 1) showed a 32.3x11.6x21.3cm multiseptated retroperitoneal mass extending from the left hemidiaphragm to the pelvis (S\textsubscript{2}-S\textsubscript{3}). CT angiography showed minimal blood supply from the IMA not warranting embolization.

Figure 1. CT scan of our patient showing a left-sided retroperitoneal mass pushing most of the abdominal organs to the right.
FBC, LFT and RFT were within normal limits. Informed consent was taken and excision of the tumor was done using a midline abdominal incision. A 4.32kg retroperitoneal mass was excised along with the spleen to which it was fixed. Post operative period was uneventful and patient was discharged home.

**Discussion**

Retroperitoneal tumor is one of the spectrums of diseases that pose both diagnostic and surgical management dilemma. Most patients are asymptomatic with abdominal distension being the commonest presentation. They account for 10-15% of soft tissue sarcoma and 0.2% of all tumors. Diagnosis is usually obtained using USS and CT scan as it is difficult to arrive at diagnosis clinically.

Surgical resection remains the mainstay of managing these wide range of tumors and may involve resection of adjacent structures just as in our patient who had splenectomy. The approach to surgical resection remains a dilemma particularly in our patient who had incongruent CT scan and angiography findings.

Most literature favors the use of midline abdominal incision which was used in our patient and all the patients who had surgery in our case study. This approach offers the advantage of better haemostasis, resection of adjacent organs infiltrated by the tumor as reported by Strauss et al in their review of surgical management of primary retroperitoneal sarcoma. Although this approach is the most favored, it has the disadvantage of post-operative ileus and long term post operative intestinal adhesions. Retroperitoneal approach is also one of the favored surgical approach as reported by Maurya et al. This approach has the advantage of reducing intraoperative fluid loss, post operative ileus thereby allowing oral feeding as soon as patient is fit enough to feed. It also saves the patient from developing post op adhesion in the future.

Minimal invasive methods using endoscopic surgery is now becoming popular as reported by Johna and co. This is believed to have similar advantage as the retroperitoneal approach with better cosmesis. Liposarcoma is the commonest form of retroperitoneal tumor and has a high propensity for recurrence.

**Conclusion**

Midline approach in the management of retroperitoneal tumors still remains a well acceptable and formidable route of achieving good oncological margin. This, reduces recurrence and improves survival.
References

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Barium enema with reference to rectal biopsy for the diagnosis and exclusion of Hirschsprung disease

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Background: Hirschsprung disease is congenital disease caused by a lack of ganglion cells in the distal bowel wall which results in functional obstruction of the aganglionic segment due to failure of relaxation during peristalsis. Barium enema is the best imaging modality to diagnose Hirschsprung disease but the gold standard to confirm the diagnosis is biopsy.

Methods: A retrospective study to assess barium enema accuracy relative to rectal biopsy in diagnosing Hirschsprung disease was done at Tikur Anbessa Specialized Hospital and data were collected from August 1st to 14th 2010. Seventy one patients who had undergone both rectal biopsy and barium enema examination between January 2002 and December 2009 were studied. In this study patients’ record were reviewed and radiologic findings were compared with histopathology results. The results were analyzed using SPSS16.

Results: In our study the mean age at diagnosis was 19 months which is a late diagnosis compared to other studies. Hirschsprung disease was more common in males and recto sigmoid was the most common area of transition zone. Large number of patients had emergency operation and diagnosis was late. Total percent agreement of barium enema and biopsy to diagnose HD is 79.1% and Kappa agreement of 0.34. In children greater than 1 year old accuracy of barium enema was very high (91.1%).

Conclusion and recommendation: Sensitivity of barium enema is less in neonates. If barium enema has positive result intervention can be done safely, especially in children above 1year old. Biopsy is not always necessary for diagnosing Hirschsprung disease.

Introduction

Hirschsprung disease (HD) is frequently encountered problem in our country. The disease can present in neonatal period with acute intestinal obstruction. Associated complications can be seen especially if it is diagnosed late. The disease more frequently presents in infancy, although some patients present with persistent and severe constipation later in life. Clinical presentation in infants includes vomiting, poor feeding, poor weight gain, and progressive abdominal distension. Early diagnosis is essential to prevent complications such as enterocolitis and colonic rupture. A rectal suction biopsy can detect hypertrophic nerve trunks and the absence of ganglion cells in the colonic submucosa, confirming the diagnosis. The radiological diagnosis of Hirschsprung disease is possible if films of diagnostic quality can be produced. Contrast enema studies are affordable and at the same time associated with minimal risk of complications. The choice of the best diagnostic modality or developing criteria-based diagnoses of Hirschsprung...
disease from a combination of the available diagnostic modalities, may avoid or reduce the complication rates associated with late diagnosis.

**Patients and Methods**

This is a cross-sectional retrospective study conducted in Tuikur Anbessa Specialized Hospital, Addis Ababa. The data were extracted from pediatric surgical operating room registered file of patients with HD from January 2002 to December 2009. The study subjects were all Ethiopian patients in pediatric age group. All study participants had both barium enema examination result and rectal biopsy result. thus 71 participants were recruited. Data were collected using a structured data collection format by principal investigator. Patient card number was retrieved from paediatric operating room registry and cards were retrieved from registration department of the hospital. Individual patient records were reviewed.

Data including demographics and presenting symptoms were extracted. It also included barium enema final radiological diagnosis and site of transition zone of Hirschsprung disease. The result of pathology specimen was also filled from patient chart. Radiological diagnosis of barium enema was reported by final year radiology resident which were always done after consultation of senior radiologists. Biopsy results of patients were also reported by senior pathologists in Tikur Anbessa Specialized Hospital pathology department. Biopsy specimens were taken by senior paediatric surgeons.

The collected data were checked for completeness and cleaned. Statistical Package for Social Sciences for window (SPSS) version 16 was used to enter and analyze the data. The total agreement between barium enema and rectal biopsy in the diagnosis of HD was calculated. Kappa agreement between two investigation modalities was also measured. Permission to conduct the research was given from the department of radiology and IRB of College of Health Sciences. Code number rather than patients name was used for data collection. The data collected were also confidential and only analyzed by the principal investigator.

**Results**

From total 71 patients who were diagnosed to have Hirschsprung disease and have both biopsy result and barium enema study 58 (81.7%) were males and 13 (18.3%) were females. The male to female sex ratio was 4.5 to 1. Their age range distribution was 7(9.9%) less than 30 days, 29(40.8%) between 1 month and 1 year and 35 (49.3%) were greater than 1 year (Table 1). The mean age at diagnosis of Hirschsprung disease was 19 months.

<table>
<thead>
<tr>
<th>Age in Months</th>
<th>Frequency</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;30 days</td>
<td>7</td>
<td>9.9</td>
</tr>
<tr>
<td>1month to 1year</td>
<td>29</td>
<td>40.8</td>
</tr>
<tr>
<td>&gt; 1year</td>
<td>35</td>
<td>49.3</td>
</tr>
<tr>
<td>Total</td>
<td>71</td>
<td>100</td>
</tr>
</tbody>
</table>

**Table 1.** Age Distribution of Patients underwent both Rectal Biopsy and Barium Enema.
Table 2. Distribution of Barium Enema findings.

<table>
<thead>
<tr>
<th>Barium Enema Findings</th>
<th>Frequency</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Suggested HD</td>
<td>53</td>
<td>74.6</td>
</tr>
<tr>
<td>Normal</td>
<td>13</td>
<td>18.3</td>
</tr>
<tr>
<td>Other diagnosis</td>
<td>5</td>
<td>7.0</td>
</tr>
<tr>
<td>Total</td>
<td>71</td>
<td>100.0</td>
</tr>
</tbody>
</table>

Table 3. Barium enema results with pathology in patients underwent both rectal biopsy and barium enema at Tikur Anbessa Specialized Hospital from January 2002 to December 2009.

<table>
<thead>
<tr>
<th>Pathology results</th>
<th>Barium enema result</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Suggested HD</td>
<td>Normal</td>
</tr>
<tr>
<td>No ganglion cell</td>
<td>49</td>
<td>8</td>
</tr>
<tr>
<td>Ganglion cells seen</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>Non conclusive</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>53</td>
<td>13</td>
</tr>
</tbody>
</table>

Most patients 43 (60.6%) presented with constipation from birth, 23 (32.4%) had both constipation and abdominal distension and the remaining five presented with acute obstructive symptoms. Barium enema studies in 53 (74.6%) suggested HD, 13 (18.3%) had a normal study and 5 (7%) were reported as showing other diagnosis such as micro colon in 2 and meconium plug syndrome in 2 (Table 2). From the 53 (74.6%) patients whose barium enema study result suggested HD; 25 showed inversion of rectosigmoid ratio as evidence and in 31 patients’ site of transition correctly identified at rectosigmoid area. Transitional zone was reported at proximal colon in one patient and at descending colon in 2 cases.

Rectal biopsy was conclusive and identified HD in 62 (88.7%) patients with no ganglion cells visible, 5 (7%) cases were reported as normal with visible ganglion cells and 4 (5.6%) cases as non-conclusive. The reason for being inconclusive biopsy result was not including the sub mucosal layer and inadequate specimens (Table 3). Total percent agreement of barium enema and biopsy to diagnose HD is 79.1% and barium enema and rectal biopsy have Kappa agreement of 0.34. In children greater than 1 year old accuracy of barium enema is high (91.1%).

Discussion

In this study mean age at diagnosis of HD is 19 months which is a delayed diagnosis compared with other studies. A report from India found the mean age at diagnosis to be only 18 days\(^1,2\). our study excluded neonates and infants who presented with acute intestinal obstruction\(^3\) and failure to pass meconium since rectal biopsy was taken after emergency surgical intervention without barium enema study. This study also showed HD is common in males 81.7% and in majority of cases recto sigmoid disease is seen. This is comparable with other studies\(^4,5,6,7\).
In this study a strong relationship was noted between a positive result of barium enema and age. Only in 2 of 7 neonates was the barium enema consistent with biopsy. Barium enema diagnosis of neonates and infants is difficult and some authors recommended that neonates who do not pass meconium in the first 48 hours of life should undergo rectal suction biopsy to establish the diagnosis of congenital megacolon. Total percentage agreement of barium enema and biopsy to diagnose HD is 79.1% and Kappa agreement of 0.34. Landis and Koch suggested kappa agreement less than 0.4 represents poor agreement. This can be explained by poor agreement seen between barium enema and rectal biopsy in neonates. In children greater than 1 year old accuracy of barium enema is high (91.1%). One study indicated diagnosis of HD with barium enema findings in children over 2 years could reach a diagnostic accuracy of up to 100%.

Most authors took radiological demonstration of a transition zone as the most important diagnostic sign on barium enema but actual detection rate is low. In our study transition zone is only reported in 46% of patients diagnosed to have HD. Other important signs like retention of barium 24 to 48 hours and stool mixed with barium correlated better with the presence or absence of HD than did any of these features alone.

In a country with limited resources barium enema can be used as first line imaging and combined with anorectal manometry or biopsy.

**Conclusion and Recommendation**

Sensitivity of barium enema is less in neonates. If barium enema is diagnostic intervention can be done safely, especially in children above 1 year old. Biopsy is not always necessary for diagnosing Hirschsprung disease. Mean age at diagnosis is late in our study and further study may be needed to know the reason.

**References**

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