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COMPLICATED MASSIVE CHOLEDOCHAL CYST: A CASE REPORT

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ABSTRACT

Choledochal cysts are rare congenital anomalies resulting from congenital dilatations of the common bile duct (CBD) and usually they present during infancy with cholestatic jaundice. This report is on a massive-sized choledochal cyst associated with massive abdominal distention, respiratory embarrassment, postprandial vomiting and intractable electrolyte imbalance leading to death within 48 hours after emergency surgery in a five-month old infant. This form of presentation is considered unusual and notable. This case report is aimed at highlighting this unique presentation and reviewing literature on choledochal cyst. It is hoped that this report will prompt early recognition and attention to the potential complications of choledochal cyst, in order to forestall morbidity and mortality. We recommend evaluation of the in-hospital and long-term postoperative outcomes of cases of choledochal cyst.

KEY WORDS: ***Choledochal cyst, Massive dilatation, Complications, Death.***

INTRODUCTION

Choledochal cyst is a rarity with an estimated incidence of 1 per 10,000 live births and a female preponderance (F:M 4:1)[1] It is a congenital cystic dilatation of the common bile duct (CBD) due to congenital defect of the duct wall, a mucosal valve or an abnormal course of the duct through the duodenal wall. Japanese are said to be more prone to this condition[1] Its pathogenesis is uncertain but it is thought to be due to a reflux of pancreatic enzymes into the CBD leading to inflammation, localized weakness, and dilatation of the duct. It may represent malformation of the CBD or occur as part of a disease spectrum that includes neonatal hepatitis and biliary atresia. Fusiform dilatation of the common bile duct (CBD) is the most common type[2-4] Other variants include a diverticulum of the CBD and dilatation of the intraduodenal portion of the CBD, which is called a choledochocoele. Symptoms and signs due to choledochal cyst seldom manifest before the age of six months. Cholestatic jaundice is the hallmark of the disorder. Characteristically, the infant presents with features of obstructive cholangiopathy, mainly jaundice with conjugated hyper-bilirubinaemia, acholic stools and bile in urine. An abdominal mass though classic, is rarely palpable and the well-described symptom triad of abdominal pain, jaundice and right upper abdominal mass, occurs in only 33% of older children[5] Complications of choledochal cyst may include severe liver dysfunction associated with ascites and coagulopathy, ascending cholangitis, biliary cirrhosis, diffuse peritonitis following rupture of the cyst and carcinoma[2-5] Delayed surgical intervention is associated with progressive biliary obstruction, irreversible liver damage and cirrhosis. The treatment of choice is a Roux-en-Y hepaticojejunostomy, as this procedure best forestalls the risk of postoperative cholangitis, stone formation and malignancy. The present report is meant to describe and draw attention to this case of life-threatening, complicated massive choledochal cyst, and to review the literature on this rare condition.

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CASE REPORT

S.D., was a five-month old female infant who presented to the Children's Emergency room of the Lagos University Teaching Hospital (LUTH), with a one month history of jaundice, passage of clay-colored stools and dark yellow urine, associated two weeks prior to presentation with fever, poor appetite, progressive abdominal distension, postprandial vomiting, respiratory distress and weakness. There was no report of diarrhoea or abnormal bleeding tendency. Although patient's illness was associated with some weight loss, her developmental history had been within normal limits before the onset of her illness. She was taken to a public secondary health institution where she was admitted for two weeks and managed as a case of septicaemia and malaria. Laboratory investigations carried out included complete blood count, blood culture, liver function tests and urine analysis. An abdominal ultrasound and plain abdominal x-ray were not done. Treatments given included antibiotics, antimalarials, blood transfusion, analgesics and multivites. While on admission at the referral hospital, the fever subsided, yellowness of the eyes and urine reduced substantially but not completely. However, the other symptoms including abdominal distension, vomiting and other features of cholestasis persisted. She was then referred with a diagnosis of pyloric stenosis, to the Lagos University Teaching Hospital (LUTH), for further management. At presentation in LUTH, she was seen at the Children's Emergency unit and subsequently admitted into the children's ward. Physical examination showed an acutely ill-looking undernourished infant who was tachypnoiec (RR=72 breaths/min),

moderately dehydrated, mildly pale and jaundiced. The pulse rate was 136/min, regular but of moderate volume and body temperature was normal (37.4°C). The abdomen was massively distended and tense, (abdominal girth of 50.4cm) with positive fluid thrill but no shifting dullness. There was no area of tenderness and it was difficult to ascertain any intra-abdominal organ enlargement such as hepatomegaly and splenomegaly because of the tense abdominal distension.

Other systems were essentially normal. The lung fields were clinically clear and apart from a tachycardia of 136 beats/min, the cardiovascular system was normal. A diagnosis of extrahepatic obstructive liver disease was entertained. The main diagnostic consideration was an obstructive cholangiopathy with chronic liver disease. The Paediatric surgeons reviewed and entertained a diagnosis of complicated obstructive cholangiopathy. Exploratory laparatomy was planned. Investigations done included an abdominal paracentesis, which yielded 50mls of bilious fluid containing 5.5g/dl of protein on analysis. Liver function tests showed total serum bilirubin of 4mg/dl with direct bilirubin of 2mg/dl and slightly elevated SGOT (66iu/l). Other liver enzymes including alkaline phosphatase were normal. Urinalysis revealed presence of bile (++) in urine. Clotting studies done (bleeding time and prothrombin time) were within normal limits. Three serial daily serum electrolytes showed persistently low sodium (range, 123 – 133meq/l), and low potassium (range, 1.9 – 2.9meq/l). Other serum electrolytes, serum creatinine and urea remained within normal limits. Abdominal ultrasound showed a sub diaphragmatic, posterior hepatic mass measuring 135mm by 110mm extending anteriorly and displacing all contiguous structures. A small fluid collection in the Morison's pouch was noted, and the gall bladder was reported as normal. Initial medical treatment given consisted mainly of intravenous correction of fluid and electrolyte imbalance. However, worsening life-threatening respiratory embarrassment secondary to massive distention of the abdomen necessitated an emergency laparatomy on the 4th day of patient's admission. The electrolyte replacement and monitoring were continued intraoperatively. The findings at surgery were bile-stained peritoneal and serosal linings of bowel with minimal greenish peritoneal fluid (5mls) and a thick-walled cystic dilatation suspected to be a pseudocyst, connected to the common bile duct and containing 500mls of thick greenish fluid. The common hepatic and cystic ducts were all dilated. Liver and gall bladder appeared normal.

A Roux-en-Y hepatico-jejunostomy was subsequently performed. Postoperatively, serum potassium and sodium remained consistently low, the least values being 2.2mEq/l and 130mEq/l respectively. This was despite intravenous replacement of estimated electrolyte losses based on the patient's weight. Unfortunately, the index patient could not have the added benefit of electrocardiographic monitoring that may have detected any cardiac abnormalities especially arrhythmias, which may have been associated with the electrolyte derangements. Although the immediate cause of death could not be categorically ascertained, the authors believe that complications from electrolyte imbalance may have contributed to the patient's demise within 48 hours after surgery.

A histopathology report of the surgical specimen showed it was a true cyst lined by cuboidal epithelium similar to that of the common bile duct. The histopathological diagnosis was choledocal cyst.

DISCUSSION

This case report was aimed at drawing attention to a massive-sized choledochal cyst leading to fatal complications. The initial symptomatology in the case under review was consistent with cholestasis with cholangitis and possibly septicaemia, early in the course of the disease. This is further corroborated by the initial response to antibiotic therapy at the referral hospital. The cholangitis may have contributed to the progressive enlargement of the choledochal cyst, leading to severe abdominal distension and respiratory embarrassment. The splinting pressure effect of the distended abdomen on the diaphragm was responsible for the respiratory distress. The electrolyte derangement in the patient may have been multifactorial in aetiology and the possible causes include increased losses from vomiting, bile ascites, electrolyte sequestration into the cysts and reduced intake due to inadequate feeding during the illness. Furthermore, drainages from the surgical site postoperatively, may have accentuated the electrolyte and fluid deficits and replacement therapy may have been insufficient. Delays in the processing of laboratory investigations and results, as well as constraints in obtaining serial electrolyte analysis at the desired frequency are some of the obstacles to appropriate electrolyte replacement. Bile contains about 134– 156mEq/l of sodium, 3.9 – 6.3mEq/l of potassium, 83-110mEq/l of chloride, 38mEq/l bicarbonate as well as cholesterol, fatty acids /lipids and bile salts[7] Bile therefore serves as a rich reservoir of body electrolytes, and so internal and external losses of this electrolyte-rich fluid may result in deficits in both electrolytes and fluid. As far as the authors are aware, there have been no previous reports on massive-sized choledochal cyst with associated life-threatening complications. We conclude that even though choledochal cysts are rare, they may present with serious complications, which may include massive abdominal distension, fluid and electrolyte derangements. Massive pooling and sequestration of bile into a huge choledochal cyst may be fatal. A similar presentation to this present case report may occur with biliary peritonitis resulting from rupture of a choledochal cyst; but this is extremely rare. Seema and colleagues[2] in India reported one of such cases, which were successfully treated.

The authors of the present case report believe that the case under review may have been successfully treated if it was detected and referred early. An earlier abdominal ultrasound during the course of the illness would have enabled earlier detection and treatment. It is hoped that this report will prompt early recognition and attention to the potential complications of choledochal cyst in order to forestall morbidity and mortality that may be associated with the condition. We recommend evaluation of the in-hospital and long-term postoperative outcomes of cases of choledochal cyst.

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