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Reports from tropics

Choledochal Cysts in African Infants: A report of 3 cases and a review of the literature

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ABSTRACT

Choledochal cysts are relatively rare and are an uncommon cause of cholestasis. Early diagnosis and prompt treatment can prevent complications such as cholangitis, cirrhosis and portal hypertension. This article reviews a rarely reported disease in Africans in whom only 3 cases were documented over the 18 year period in Nigeria. The 3 cases were all females with ages between less than a month and 13 months at presentation. All presented with abdominal swelling with or without jaundice or acholic stools. The use of real-time ultrasonography antenatally and postnatally aided the diagnosis in our patients. Two of the patients presented and were operated and both made full recovery confirming the importance of early surgical intervention. The third patient died, and exemplified the consequences of delayed diagnosis and treatment which occur not uncommonly in developing countries mostly because of sparse and or expensive tertiary health care facilities.

INTRODUCTION

Choledochal cysts are congenital cystic malformations of the biliary tract, with the highest incidences in the Japan.[1,2] Though cases have been reported in African Americans,[2] there is a paucity of data in indigenous Africans. Choledochal cysts are classified into 5 types based on the location of the cyst.[3] Type I lesion is the most common accounting for 60-90% of cases.[4] It is also the most common in children while adults more often have Type IV choledochal cysts.[3,4] The clinical presentation is variable and may be nonspecific but is often related to the age at presentation.[5] In infancy, when the biliary obstruction is total or near total, the patient may present with cholestasis. Presentation in later part of the childhood suggests that biliary obstruction is incomplete.[6] The patient may then present with the classical triad of jaundice, abdominal pain , and abdominal swelling or mass.[4,5] High resolution real-time abdominal ultrasonography accurately defines the size of the cyst, and is important in its classification and evaluation.[6] The relative rarity, and the absence of any previous report of choledochal cyst(s) from Africa has prompted this documentation.

PATIENTS AND METHODS

Between 1985 and 2003, two cases of choledochal cyst were seen at the Paediatric Department of the University College Hospital, Ibadan, Nigeria and a third case was admitted and managed at the Chevron Hospital, Lagos, Nigeria. The 3 patients were all females with a median duration of symptoms of 30 days at first presentation in the hospital.

Case 1: A 40-day old female infant presented at the University College Hospital, Ibadan, Nigeria with progressive right flank abdominal swelling since birth. Clinical examination was essentially normal except for the disproportionate right-sided abdominal bulge. Bowel movements, stool colour, and liver function tests were normal. However, plain radiograph of the abdomen as well as upper gastrointestinal barium study showed displacement of the bowel loops inferiorly and to the left, as well as an extrinsic impression of a mass on the lesser curvature of the stomach. Abdominal ultrasonography showed a large cystic mass in the right hypochondrium. Exploratory laparotomy revealed a huge Type I choledochal cyst. Two hundred ml of bile was aspirated from the cyst which was then excised and a Roux-en Y hepatico-jejunostomy was performed. The postoperative period was uneventful, and the patient was discharged in apparent good health. However, long term follow up was not possible as the patient failed to keep clinic appointment.

Case 2: A female infant was first seen at the University College Hospital, Ibadan at the age of 13 months after visits to various peripheral hospitals. She initially presented with a history of progressive jaundice since the age of one month. Progressive abdominal swelling was observed from the age of five months. Pale stools as well as difficulty with breathing became apparent to the parents at 1 year of age and this necessitated the ultimate presentation at the University College Hospital, Ibadan at 13 months of age. Clinical examination at this time revealed a deeply jaundiced, chronically ill child with failure to thrive. Examination of the abdomen showed gross irregular distention (Figure). The liver and spleen were 9 cm and 4 cm below the right and left costal margins respectively. Total serum bilirubin ranged from 130 mmol/L (direct reacting fraction 100 mmol/L) on admission to 92 mmol/L (direct reacting of 48 mmol/L). Serum alkaline phosphatase ranged between 1782 and 1568 IU/L (normal < 40 IU/L). Coagulation status was deranged at admission with INR of 3.5 which improved to 1.1 after parenteral vitamin K administration. Abdominal ultrasonography and computed

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Figure: Case number 2, showing abdominal distension due to choledochal cyst.

tomography showed a Type IVA choledochal cyst with multiple intrahepatic and extrahepatic cysts. The child developed pneumonia soon after admission and was treated with broad spectrum antibiotics parenterally. However, the child remained extremely ill and was not fit to withstand surgery. Palliative percutaneous ultrasound-guided aspiration and drainage of cysts yielded about 250 ml of bilious effluent per day for two weeks when the child succumbed to the illness. Postmortem examination revealed huge intra and extra hepatic choledochal cysts. No cysts were present in either the spleen or the kidneys.

Case 3: A female infant delivered at term with a birthweight of 3.8 kg presented in the immediate postnatal period with progressive abdominal swelling since birth. A prenatal abdominal scan at 33 weeks gestation had revealed a cystic mass associated with the liver and this was confirmed on day 22 of life by an abdominal ultrasound scan. A contrast-enhanced computed tomography of the abdomen showed a huge cystic mass closely related to the extrahepatic biliary tree with marked displacement of the bowel loops to the left. There were no cysts in the kidneys, spleen, or within the liver. Exploratory laparotomy revealed a 12 cm diameter Type II choledochal cyst arising as a diverticulum from the common bile duct. The cyst was excised and a Roux-en-Y hepatico duodenostomy was performed. Immediate post operative period was satisfactory and the patient remained healthy over a 4-year follow up period.

DISCUSSION

Previously considered rare, choledochal cysts have been increasingly reported from various parts of the world [2,4,6] except from the African continent. The incidence is highest in the Far East particularly in Japan where rates as high as 1 per 1000 have been reported compared to 1 in 13,000 to 15,000 in Western countries.[1] Although reported in African Americans[2], a literature search yielded no previous report from the African continent. In a previous report of 102 Nigerian infants with conjugated hyperbilirubinemia, two children had diverticulum of the common bile duct but no further details were given regarding the exact anatomy and nature of the diverticulae.[7] A later review of infantile cholestasis in the same hospital did not reveal any case of choledochal cyst.[8] In this report also from Nigeria, only 3 cases were documented over a 18 year period and all three were females. This is in consonance with the documented female preponderance of this malformation. [1, 2, 4, 6, 7]

Choledochal cysts are classified into 5 types according to the location of the cysts.[3] Type I, exemplified by our Case 1, is a fusiform cystic dilatation of the common bile duct, and it is the most common in published series. Type II, as in our Case 3, is a true diverticular malformation of the common bile duct, and comprises less than 2 % of reported cases.[9] Type III cysts or choledochocoele account for up to 5 % of cases in the literature, while Type IVA cysts (Case 2) occur in about 10 % of cases and are usually multiple intra and extrahepatic cysts. Less commonly, there are extrahepatic cysts only (IVB). Type V consists of single or multiple hepatic cysts.

The pathogenesis of choledochal cyst is uncertain. It has been suggested that a defect at the junction of the common bile duct and the pancreatic duct allows pancreatic fluid to reflux into the common duct thus weakening it.[10, 11] While the finding of amylase in some choledochal cysts supports this theory, the presence of prenatal choledochal cysts on ultrasound examination in the first trimester raises doubt about the pancreatic reflux theory. [12] Primary abnormality of epithelial proliferation during embryological ductal development resulting in congenital weakness of the bile duct wall, as well as congenital obstruction are also probable causes. [13]

While jaundice is the most common presenting symptom in infancy, older children are more likely to present with abdominal pain as well as other clinical features such as hepatomegaly, acholic stools, and vomiting. [14] Presentation with the classical triad of jaundice, abdominal pain, and abdominal mass is infrequent and is more likely to occur in older patients.[14, 15] All three of our patients presented with abdominal swelling. In addition, our Case 2 presented also with splenohepatomegaly and deranged liver function secondary to chronic cholestasis due to delay in diagnosis.

Laboratory findings in the patient with choledochal cyst indicate cholestasis in the majority of patients. Significant compromise of hepatic synthetic function may also occur as in our Case 2. The most useful initial imaging study is abdominal ultrasonography. Also, antenatal sonography has been used in the prenatal diagnosis of choledochal cyst[12, 16] as it was in our Case 3. If ultrasonography is not conclusive, an upper gastrointestinal barium examination, hepatobiliary nuclear scan, computed tomography, or magnetic resonance imaging may provide important diagnostic Clues. [2, 17, 18]

The treatment for choledochal cyst is surgical excision of the entire cyst wherever possible. Where extensive intrahepatic and extrahepatic involvement precludes complete resection, or the patient presents late with complications as in our Case 2, decompression and drainage of the cysts may be all that can be

done. Total extirpation of the cysts is essential because the epithelial lining of the cysts can undergo malignant transformation with time.[19] Long term follow-up is important in patients who have drainage procedures because of probable complications such as cholangitis, cholelithiasis, pancreatitis, and malignancy.[20]

This study confirms that ultrasonography, a non invasive and relatively inexpensive procedure that is likely to be available in many referral centers in tropical developing countries, is a good diagnostic tool in choledochal cyst both pre and postnatally. It also shows that early diagnosis and prompt surgery are essential to ensure a successful outcome. Delay in diagnosis and treatment increases the morbidity and mortality that can occur in this treatable condition.

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