Choledochal cyst associated with HIV (Human Immunodeficincy Virus) disease

MUGUTI G I, NDUKU K, MUTANDWA C, RUSHESHA W

Introduction

Choledochal cysts are believed to be congenital dilatations of the intrahepatic or extrahepatic bile ducts, or both, of unkown aetiology.[1] They were first described by Vital and Ezler in 1723.[2] However, the aetiology of choledochal cyts remains speculative despite a considerable research effort over the years.

Cent Afr J Med 2001;47(9/10):229-32

Correspondence to:
Professor GI Muguti
Department of Surgery
University of Zimbabwe
Faculty of Medicine
P O Box A178
Avondale
Harare, Zimbabwe

Choledochal cyst is a rare abnormality with an incidence in the Western population of 1:30 000 to 50 000[3]. It is far more common in the East with an estimated incidence of 1:1 000 described in Japan.[4] Although anectdotal reports of choledochal cyst in African patients have appeared in the literature[5] the incidence in the African population is not known. It appears, however, that this condition is rare in the African population. The disease occurs more commonly in females with the female to male ratio ranging from 2.4:1 to 4:1.[1,4,6]

The complete classical triad of right upper quadrant pain, mass and jaundice seen more commonly in children is a rare presentation in adults.[7] Patients usually present with one or two symptoms.[8] The authors have treated three female patients with choledochal cyts and HIV disease over the last five years. Only one of these patients who is reported in detail in this paper presented with the classical triad. The appearance of this third patient prompted this report. One of the patients aged 29 years presented with recurrent episodes of upper abdominal pain and the other aged 26 years presented with jaundice and upper abdominal pain. In both these patients fusiform dilatation of the common bile duct was diagnosed by abdominal ultrasound scan and CT scan. The jaundiced patient recovered well following a Roux-en-Y choledochojejunostomy while the other patient was managed conservatively with antibiotics. This other patient had full blown AIDS and was not considered a candidate for major surgery.

The association between HIV disease and choledochal cyst described in this paper has not been reported before. While this might be a chance finding, it is possible that HIV infection may be involved in the aetiology of choledochal cyst in some patients suffering from HIV/AIDS (Acquired Immunodeficiency Syndrome). It is important to note that Zimbabwe has a high incidence of HIV infection. In the year 2000 there were 1.5 million people estimated to be living with HIV/AIDS in Zimbabwe.

Case Report

Clinical Presentation.

A 24 year old woman was referred to our hospital with a three months history of general body itchiness, yellow eyes, passing yellow urine and a painful upper abdominal swelling. She had lost about 10kg in weight since the onset of this illness. Of note in her past medical history is that she was treated at a local hospital for an episode of jaundice two years prior to the present illness. At that time the jaundice resolved spontaneously.

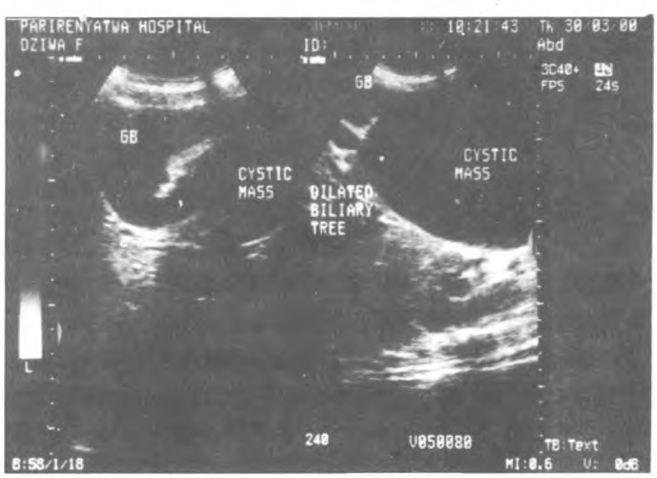
Physical examination revealed a moderately jaundiced young woman. She was noted to have a small mobile lymph node in the left axilla. Abdominal examination revealed a distended abdomen with a mass in the right upper quadrant measuring 10cm x15cm. The mass was slightly tender on palpation. The rest of the physical examination was non-contributory.

Investigations.

Full blood count (FBC) and urea and electrolytes were within normal limits. The hepatitis B screen was negative. The results of other investigations were as well follows: International Normalised Ratio (INR) = 2.2; Prothrombin time (PT) 24 seconds and abnormal liver functions tests with total bilirubin 238 uMol/L; alkaline phosphatase 778IU/L; alanine transferase (ALT) 108IU/L; total protein 82g/L; albumin 24g/L and conjugated bilirubin 40umol/L.

An abdominal ultrasound scan was requested and this showed "a massive cystic dilatation of the common bile duct approximately 10cm x 15cm with sludge but no stones. The intrahepatic biliary tree and the gall bladder were dilated but the liver appeared normal" Figures IA, IB). At this stage a diagnosis of choledochal cyst type I was made. The chest X-ray was normal.

Figures IA and IB.



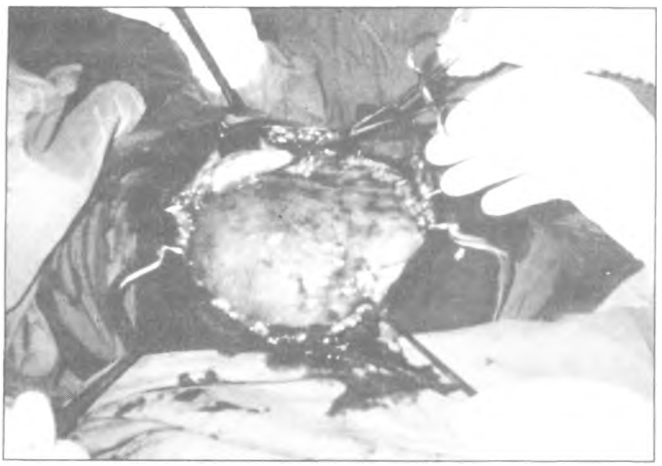
As major surgery was contemplated the patient was precounselled and tested for HIV. The HIV test was positive on both ELISA (enzyme linked immunosorbent assay) and Western blot. Following this result the patient was post counselled in the usual way. It was not possible at the time to do CD4/CD8 counts at our hospital.

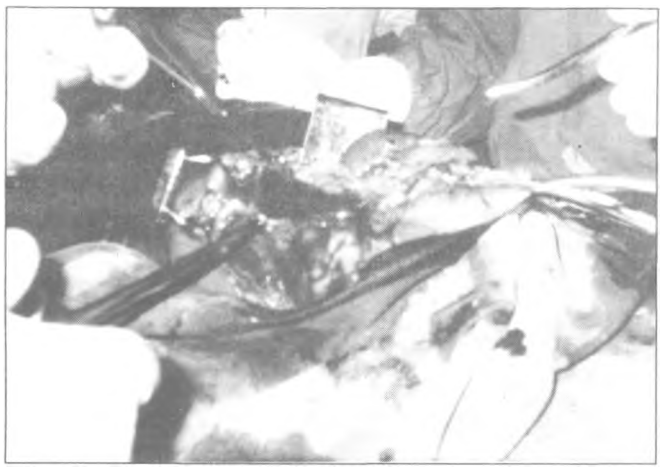
Operation.

After adequate preoperative preparation the patient was taken to theatre. At laparotomy through a right subcostal incision a massive choledochal cyst was found (Figures

IIA, IIB). In view of the immunocompromised state of the patient and the massive size of the cyst it was felt that a cholecystectomy and a Roux-en-Y choledochojejunostomy would be a safer procedure than total cyst excision and hepaticojejunostomy. The patient was commenced on antibiotics and had an uneventful post operative course. She was discharged from hospital within three weeks of her operation. Histology of the cyst wall showed "a choledochal cyst with features of chronic inflammation".

Figures IIA and IIB





Follow Up.

The patient is on our books for long term follow up. She was quite well at her last outpatient visit three months after the operation.

Discussion

Choledochal cysts typically present in infancy and childhood. About 25% of patients are diagnosed within the first year of life and 50% to 60% before the age of 10 years.[3] In 20% to 30% of cases, diagnosis is delayed until adulthood.[3,9] The three patients we treated with choledochal cyst and HIV disease were all adult female patients.

The prevailing view of choledochal cyst as solely a congenital condition has been challenged by some authors.[10,11] Chaudhary *et al* in a study of 49 patients with choledochal cyst describe two adult patients who had normal bile ducts at operative cholangiogram performed during cholecystectomy who later developed choledochal cysts.[10] Schmid *et al.*[11] reported similar findings in some of their patients, suggesting an acquired aetiology in such patients. These observations and the association between choledochal cyst and HIV disease reported in this paper suggest that not all choledochal cysts diagnosed in adulthood represent a delayed diagnosis of a congenital condition. The possibility of an acquired aetiology in such patients should also be considered.

Although our report is the first one to describe an association between HIV infection and choledochal cyst a few reports have described an association between choledochal cyst and some other types of viral infection.[12,13] Shito et al reported a case of asymptomatic intra hepatic choledochal cyst associated with chronic active hepatitis C.[12] In a detailed study of 23 patients with extra hepatic biliary atresia (EHBA) nine patients with choledochal cyst (CDC) and 33 patients with other hepatobiliary diseases, Tyler *et al.* demonstrated that the prevalence of reovirus RNA in tissues from patients with EHBA and CDC was significantly greater than that in the patients with other hepatobiliary diseases.[13]

Landing suggested that an inflammatory process injuring bile duct epithelial cells could lead to either duct obliteration (EHBA) or weakening of the bile duct wall with subsequent dilatation (CDC).[14] Histopathological study of bile duct and liver tissues obtained from children undergoing porto enterostomy (Kasai procedure)[13] has provided support for the concept of a persistent inflammatory process leading to progressive bile duct destruction.[13,15] As a result of their observations Tyler *et al.*[13] and Riepenhoff-Talty *et al.*[16] believe that a causal relationship exists between reoviruses and rotaviruses *(reoviridae)* and the majority of cases of EHBA and CDC.[13]

Due to limited facilities in our institution we were not able to use reverse-transcriptase polymerase chain reaction (RT-PCR) on the tissue biopsy from the cyst wall to look for the presence of the HIV virus. Despite this, we believe our observation of this association is important and should lead to further research in this area. The role of the HIV virus in the aetiology of CDC remains speculative at this stage. It has, however, been suggested that, in humans, EHBA and CDC might result from exposure of an immunologically susceptible or genetically predisposed infant to a specific strain or strains of reovirus with increased virulence.[13]

It is interesting to note that an aetiological association between HIV infection and aneurysms is believed to exist.[17] This raises the possibility of a similar association between HIV and choledochal cyst. However, considering the high incidence of HIV in Zimbabwe it is possible that this association could be a chance finding.

At present most choledochal cysts are believed to be congenital dilatations of the bile duct associated with an anomalous junction of the pancreaticobiliary ductal system

in more than 70% of patients.[18] Reflux of pancreatic juice into the CBD is presumed to be a chronic irritant in such cases.[19]

The main principle of treatment of choledochal cyst is total cyst excision provided it is technically feasible.[1,7] This is particularly important to prevent the most serious complication: malignant degeneration of the cyst.[1] There is an increased incidence of carcinoma in choledochal cyst, ranging from 2.5% to 26%, which is well above the rate of <1% for the general population.[7]

Although Roux-en-Y choledochojejunostomy is not regarded as an optimal procedure for the definitive treatment of choledochal cyst it should be seriously considered as a safer alternative for cases in which technically the cyst is difficult to excise due to inflammation and adhesions caused by repeated cholangitis.[20] Given the overall poor prognosis of our patient with HIV disease we were satisfied that a simple procedure such as the Roux-en-Y choledochojejunostomy was more appropriate than total cyst excision which involves a more extensive dissection.

Choledochal cyst is a disease which is still not well understood. Our report draws attention to the possibility of an aetiological association between choledochal cyst and HIV disease. This association also raises questions as to what the optimum surgical treatment should be in patients suffering from HIV/AIDS.

References

1. Postema R R, Hazebroek W J. Choledochal cysts in children: a review of 28 years of treatment in a Dutch children's hospital. *Eur J Surg* 1999;165:1159-61.
2. Vater A, Ezler CS. Dissertatio de serrichis visceru occasione sectionis viri typanite defunte. Wittenburgae 1723,4 Pamphlers; 881:22.
3. Vereruysse R, Van den Bossche MRP. Choledochal cyst in adults. *Acta Chir Belg* 1998, 98: 220-2.
4. Babbit DP. Congenital cysts: new aetiological concepts based on anomalous relationships of the common bile duct and pancreatic bulb. *Ann Radio* 1969;12: 231-40.
5. Munkonge L. Choledochal cyst: personal experience in the management of choledochal cyst. *Cent Afr J Med* 1993;39:224-7.
6. Lopez RL, Pinson CW, Campbell JR, Harrison M, Katon RM. Variation in management based on type of choledochal cyst. *Am J Surg* 1991; 161:612-5.
7. Weyant MJ, Maluccio MA, Bertagnolli MM, Daly JM. *Am J Gasrtoentrol* 1998; 93: 2580-3.
8. Suita S, Shono K, Kinugasa Y, Kubota M, Matsuo S. Influence of age on the presentation and outcome of choledochl cyst. *J Pediatr Surg* 1999;34:1765-8.
9. Flanigan DP. Biliary cysts. *Ann Surg* 1975; 182: 635-41.
10. Chaudhary A, Dhar P, Sachdev A, Kumar N, Vij JC, Sarin SK *et al.* Choledochal cyst — differences in children and adults. *Br J Surg* 1996; 83:186-8.
11. Schmid C, Meyer HJ, Ringe B, Scheumann GFW, Pichlmayr R Cystic enlargement of extrahepatic bile ducts. *Surgery* 1993;114:65-70.
12. Shito M, Shintoku J, Miyazaki H, Mukai M. Asymptomatic intrahepatic choledochal cyst associated with chronic active hepatitis C. *Hepato-Gastoenterol* 1998;45:2356-8.
13. Tyler KL, Sokol RJ, Oberhaus SM, Mysan LE, Karrer FM, Low R, *et al.* Detection of reovirus RNA in hepatobiliary tissues from patients with extrahepatic biliary atresia and choledochl cysts. *Hepatology* 1998;27:1475-82.
14. Landing BH. Considerations of the pathogenesis of neonatal hepatitis, biliary atresia and choledochl cysts; the concept of infantile obstructive cholangiopathy. *Prog Pediatr Surg* 1974;6:113-39.
15. Desmet VJ, Callea F. Cholestatic syndromes of infancy and childhood. In: Zakim D, Boyer TD, editors. Hepatology. Philadephia, Saunders, 1996;1649-98.
16. Riepenhoff-Talty M, Gouvea V, Evans MJ, Svensson L, Schakel K, Zhaori G, *et al.* Detection of Group C rotavirus in infants with extrahepatic biliary atresia. *J Infect Dis* 1996;174:8-15.
17. Nair R, Abdool-Carri MA, Robbs J. Arterial aneurysms in patients infected with human immunodeficiency virus: a district clinicopathology entity? *J Vasc Surg* 1999;29:600-7.
18. Kelly DM, O'Donnell PJ, Howard ER. Pulmonary lymphangitis carcinomatosa and acute pancreatitis: a rare presentation of choledochal cyst. *HPB Surg* 1999;11:163-9.
19. Behrns KE, Shaheen NJ, Grimm IS. Type I choledochal cyst in association with fimilial adenomatous polyposis. *Am J Gastroenterol* 1998; 93:1377-9.
20. Hasegawa T, Kim M, Kitayama Y, Kitamura K, Hiranaka T. Choledechal cyst associated with polycystic kidney disease: report of a case. *HPB Surg* 1999;11:185-90.