

Choledochal cyst with adenoma

J. Gupta

Department of General Surgery, I.G.M.C, Shimla, Himachal Pradesh

Keywords: Choledochal cyst; roux-en-y; hepatojejunostomy

Abstract

Choledochal cyst usually presents during first decade. Unresected choledochal cyst is associated with increased risk of cholangiocarcinoma. We report a 46 year female who was incidentally diagnosed to be having choledochal cyst with adenoma.

Introduction

A choledochal cyst is defined as isolated focal or combined congenital dilatation of extrahepatic or intrahepatic biliary tree. Around 50% of the patients usually presents during the first decade of life. Females are more commonly affected than males (4:1). Choledochal cyst with adenoma is a rare entity.

Case report

A forty six year old female who was investigated for menorrhagia at the gynaecology outpatient department underwent an ultrasound scan USS of the abdomen and pelvis. A grossly dilated common bile duct (CBD) with a possible intra luminal polyp was detected and the patient was referred to the surgical unit. She had no history of jaundice, abdominal pain or fever. On examination she was not icteric, pale or cachectic. There were no stigmata of chronic liver disease. Abdominal examination was unremarkable. Complete blood count, renal function test, serum electrolytes and liver function tests including serum bilirubin and alkaline phosphatase were normal. In the ultrasonograph the dilated segment was 6.0 x 2.4 cm in size and the lower part of the CBD was normal. Inside the dilated segment there was a soft tissue pedunculated mass of 1.3 x 0.8 cm in size (Figure1). Intrahepatic biliary radicle and gall bladder were normal. Magnetic Resonance

cholangiopancreatography (MRCP) showed choledochal cyst with small pedunculated polyp (Fig.2 and 3) with non visualized distal end. Rest of the biliary system was normal. Patient was diagnosed to be having type Ib choledochal cyst with polyp and was scheduled for excision of the CBD with reconstruction. Intraoperative findings confirmed the observations made on imaging. Excision of cyst with Roux –en y hepatojejunostomy was done. Post operative period was uneventful. Inside the choledochal cyst was a pedunculated polyp. On histological evaluation the cyst was lined by a benign columnar epithelium, with



Figure 1. USG showing polyp in CBD



Figure 2. MRCP showing pedunculated polyp in choledochal

Correspondence: J. Gupta

E-mail: jagdishdrblp@gmail.com

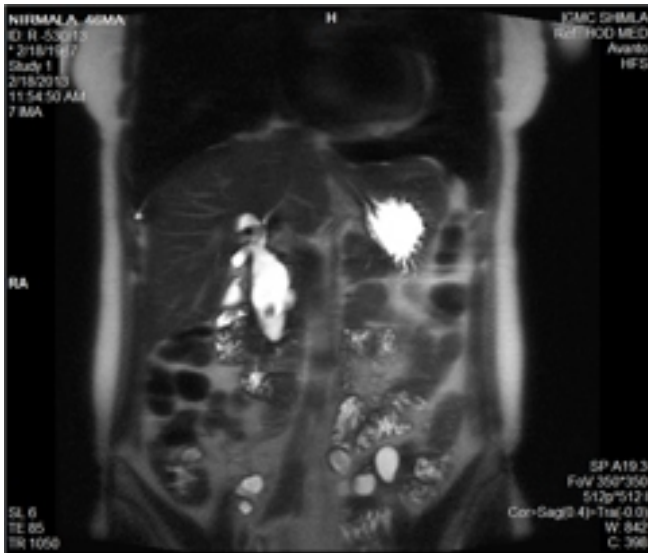


Figure 3. MRCP showing filling defect in non dependent part

thickening and fibrosis of the wall. Polyp showed features of adenomatous polyp with no evidence of dysplasia.

Discussion

Choledochal cyst (CDC) was initially described by Douglas in 1852. Anatomical classification proposed by Alenso et al was revised by Todani et al in 1977 [1]. Type I is the commonest type occurring in 75% of cases. Classic symptom complex of abdominal pain, mass and jaundice is uncommon and is observed in only 20-30% patients. There may be recurrent abdominal pain with minimal jaundice, which may not be readily apparent as in our case. Imaging techniques help in the diagnosis of choledochal cyst. USS helps in diagnosis of 93% of paediatric and 72% of the adult population. Magnetic resonance cholangiopancreatography is the non invasive procedure of choice at present. CDC is associated with abnormal pancreatic and bile duct union (anomalous biliopancreatic junction) in 90% of cases. Differential diagnosis is biliary stricture. In contrast to type I cyst obstructing lesion will often cause elevated alkaline phosphate and bilirubin. Various complication of CDC are cholangitis, gall stones, jaundice, pancreatitis, portal hypertension and cholangiocarcinoma. Various malignancies are cholangiocarcinoma, gall bladder

carcinoma, adenoma and bile duct sarcoma. Benign tumours of extrahepatic biliary tree rare, they account for only 6% [3]. Bile duct adenoma, is a rare neoplasm of the extra-hepatic biliary tree and very few cases have been reported in a choledochal cyst[4]. Although this tumor is benign, it is considered to have a malignant potential[5]. In MRCP a stone as well as tumour will appear as dense shadow. A stone will be detected in a dependent part while a polyp can be in the non dependent area. Lifetime risk of cholangiocarcinoma in CDC paediatric population is 2.3%. In older untreated patients the reported incidence is as high as 75%. Speculated etiological mechanism of carcinogenesis is bile stasis, reflux of pancreatic juices mixed with bile, changes in bile acids, superinfection or inflammation. There is pathological evidence of hyperplasia-dysplasia and cancer in patients with pancreaticobiliary malignancy. Hyperplastic cells have increased cellular proliferation markers cyclooxygenase-2, vascular endothelium growth factor and k-ras mutation. Excision of gall bladder, CDC with Roux-en-Y hepaticojejunostomy is the most commonly used surgical procedure.

References

1. Todani T, Watanabe Y, Narusue M, et al. Congenital bile duct cysts : classification, operative procedures and review of 37 cases including cancer arising from choledochal cyst. *Am J Surg* 1977;134: 263-69.
2. Parikh PY, Lilemoe KD. Choledochal cysts and benign biliary strictures. In: Maingot's Abdominal operations. 12th ed 1032
3. Dowdy GS, Olin WG, Shelton EL et al. Benign tumors of the extra-hepatic biliary ducts: report of 3 cases and review of literature. *Arch. Surg.* 1962; 85 : 503-13.
4. Aggarwal S, Kumar S, Kumar A, et al. Extrahepatic bile duct adenoma in a patient with choledochal cyst. *J Gastroenterol and Hepatol* 2003; 18 : 351-55
5. Davies W, Chow M, Nagorney D. Extra-hepatic biliary cystadenomas and cystadenocarcinomas. Report of seven cases and review of the literature. *Ann. Surg.* 1995; 222

Key points:

- Lifetime risk of cholangiocarcinoma in a paediatric choledochal cyst population is 2.3%
- In older untreated patients the reported incidence is as high as 75 %.
- Ultrasound is used in the diagnosis, but magnetic resonance cholangiopancreatography is the non invasive procedure of choice.