

## **Pleomorphic Rhabdomyosarcoma within a choledochal cyst; a rare cause of obstructive jaundice**

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### **Abstract**

Bile duct cysts are rare and of uncertain origin. Most have been reported in young females of Asian Descent. The long term complication of choledochal cysts includes malignant transformation in the epithelial lining of biliary tree. Histopathologically it is Cholangiocarcinoma in the majority of cases with poor long-term prognosis. However, Rhabdomyosarcoma associated with choledochal cyst in an adult is rarely reported before. The authors report one such case in an adult female patient who presented to us with features of obstructive jaundice.

### **Introduction**

Choledochal cysts can be both intrahepatic and extrahepatic in distribution. For reasons which are not yet clear it is much more prevalent in Asians and in females.<sup>1</sup> These cysts are clinically important because of the complications of recurrent cholangitis, biliary stricture, choledocholithiasis, recurrent acute pancreatitis and rarely malignant transformation into cholangiocarcinoma. We describe a patient with malignant polyp within a choledochal cyst, which was identified as Rhabdomyosarcoma. To the best of authors knowledge, no previous reports of such an association in adult population exist.

### **Case Report**

A 56 years old lady presented with complaints of generalized itching and a significant weight loss of 20 kg in the last 3 months. She also complained of yellowish discoloration of her eyes and clay colored stools for the last 1 month. She had no prior medical history except an open cholecystectomy, which was carried out 20 years back due to symptomatic gallstones. No other pathology was picked up in that operation and postoperatively she remained asymptomatic. On presentation she was afebrile, jaundiced, had a right subcostal scar and enlarged liver along with a firm mass in right hypochondrium with ill-defined borders. She was found to be having a total bilirubin of 6.8 mg/dl with predominantly conjugated hyperbilirubinaemia, her alkaline phosphatase was 597 mg/dl and ESR was 103 mm first hour.

Ultrasound examination showed intrahepatic biliary dilatation along with a massively distended common bile duct (CBD). It also revealed a large hypoechoic mass within the CBD. CT scan showed mixed density mass close to the head of pancreas and duodenum with both cystic and solid component (Figure 1). Based on the above findings an ERCP was performed. Although contrast showed markedly distended CBD, a large filling defect was also noted in it (Figure 2). A biliary stent was passed above the upper limits of this mass in the same setting. Workup did not show any evidence of lymphadenopathy or vascular invasion and



Figure 1.

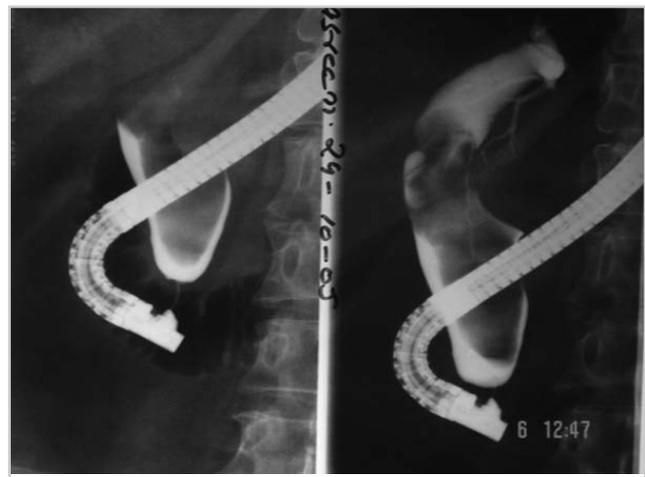


Figure 2.

despite the large size of the filling defect, there was no evidence of any local invasion. These features were quite consistent with a preoperative diagnosis of benign polyp within a Type I choledochal cyst.

Patient was explored and peroperatively large cystic dilatation of the CBD with absent gall bladder was seen. Once this was dissected a polypoid mass of firm consistency having a size of approximately 8 x 3 cm size was found within the CBD. It was arising from the posterolateral wall of CBD close to the confluence and was well confined within it. No evidence of any lymphadenopathy or vascular invasion was identified. We were able to dissect the cyst completely and after obtaining grossly tumour free excision margins Roux-en-Y Hepaticojejunostomy was performed. Gross examination of the polypoid like growth revealed both cystic and solid component.

Histopathology of the specimen showed singly scattered pleomorphic elongated to polygonal cells containing hyperchromatic bizarre nuclei and eosinophilic cytoplasm. The neoplasm consisted of diffuse sheets and cords separated by fibrovascular cores. Uninucleated and multinucleated tumour giant cells were also present focally. Both the proximal and distal margins of the specimen were found to be tumour free. The histopathological findings and subsequent immunohistochemical staining with Cytokeratin, Desmin and Myoglobin stains confirmed a diagnosis of Pleomorphic Rhabdomyosarcoma. Her postoperative course in the hospital was uneventful.

## Discussion

Bile duct cysts are typically described as a disease of childhood. The incidence of congenital cyst is reported to be between one in 13000 to one in 2,000,000 live births.<sup>2</sup> Although more than 60% of patients with bile duct cysts are diagnosed during the first decade of life, some 20% go undiagnosed into adulthood.<sup>3</sup> The most favored hypothesis about the formation of choledochal cysts is the "long common channel theory" according to which an anomalous pancreatobiliary ductal junction (APBDJ) with a long common channel (more than 15mm) allow reflux of pancreatic juice into the biliary system causing an increase in ductal pressure, inflammation and subsequent ductal dilatation. Also this pancreatic juice reflux into the biliary duct, combined with cholestasis and repeated bacterial infection may play an important role in provoking malignant changes in the biliary epithelium. APBDJ is noted in 60-96% of patients<sup>4</sup>, suggesting that there must be other factors involved as well. These might include abnormal function or dysmotility of the sphincter of Oddi and oligoanglionosis of distal CBD.

Choledochal cysts are classified according to the

Todani Classification System<sup>5</sup> into 5 major and 5 subtypes based on anatomical location. Type I cyst are confined to the extrahepatic bile duct (EHB) and can be further subdivided into I a involving the entire EHB, I b involving only a focal segment of EHB and I c involving only the CBD. Type II cysts are true diverticula of the EHB. Type III cysts are also referred as choledochoceles are confined to EHB within the duodenal wall. Type IV cysts are multiple. They are labeled as IV a involving both the EHB and intrahepatic bile duct (IHB) and type IV b involving multiple segmental dilations of the EHB. Type V cysts (Caroli's disease) are confined to the IHB. Type I cysts are the most common variety (78%).

It is well established that 10-30% of adults with bile duct cyst develops malignancy, which in most of the cases is cholangiocarcinoma.<sup>6</sup> This may arise either in the cyst wall itself, in remnant tissue or undilated parts of the extrahepatic or intrahepatic bile duct. Premalignant alteration in the epithelial wall of the cysts increases with age. Here it is important to note that the classical triad of jaundice, pain and an abdominal mass, which is common in the children, is less frequently seen in adults (85 versus 25% respectively).<sup>7</sup> Adults usually present with vague symptoms, generally in the right upper abdominal, quadrant and like this patient, as many as 10-50% have undergone cholecystectomy or other surgical exploration before diagnosis.<sup>7</sup> The incidence of bile duct cancer has been reported to be more than 120 times greater than that in the general population, and this risk remains high even after surgical treatment.<sup>8</sup> Type I and IV cysts are associated with a higher incidence of malignancy.<sup>5</sup> Long-term survival in patients who have developed malignancy is rare. For these reasons cystenterostomies, which were frequently performed before 1980's is not recommended any more. Consensus now favors complete cyst excision with cholecystectomy and Roux-en-Y Hepaticojejunostomy reconstruction for type I cysts. Operative Cholangiography and cyst endoscopy is recommended by some not only to clarify the anatomy, but also to reveal proximal ductal stenosis, calculi and aberrant ducts. One should realize that the management of choledochal cyst has developed from institutional experience and it's basically a level III evidence (opinion of respected authorities, in the absence of proper trials) as the rarity of this disease is limiting the clinical experience.

Although Rhabdomyosarcoma is rarely described in the hepaticobiliary tract, its association with Choledochal cyst is not an established entity. To the best of our knowledge only one other case of Rhabdomyosarcoma in a choledochal cyst has been reported in literature, which occurred in an 8 years old female.<sup>9</sup> Furthermore in the vast majority of cases the Rhabdomyosarcoma within the

hepaticobiliary tract is of embryonal type. According to the Intergroup Rhabdomyosarcoma Study<sup>10</sup> the recommended treatment is multidisciplinary where a combination of major resection, chemotherapy with or without radiation of the porta hepatis could improve the outcome.

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