Choledochal Cyst in an Adult: Congenital or an Acquired Clinical Entity?

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Abstract

Cystic dilation of biliary ducts, otherwise known as choledochal cyst, is an uncommon condition frequently present in infancy and childhood but increasingly diagnosed in adults. Until now, no unifying etiologic theory exists for choledochal cysts, but most literature would conclude they are congenital bile duct anomalies. This report discusses choledochal cyst in a 50-year-old female post-laparoscopic cholecystectomy who had previous cholangiogram that would prove its non-existence and emphasizes the possibility of acquired causes. On this second admission, a type IVa choledochal cyst was identified by MRCP and patient subsequently underwent left hepatic lobectomy with en-bloc resection of extrahepatic ducts, reconstructed with a roux-en-y hepaticojejunostomy. Histopathologic findings were consistent with choledochal cyst and post-operative course was unremarkable.

Introduction

Cystic disorders of the bile ducts, although rare, are well-defined malformations of the intrahepatic and/or extrahepatic biliary tree. Choledochal cysts are four times more common in females than males.[1] The prevalence of this disease varies in different parts of the world. It is most common among Asians, with an incidence of 1 in 13,000, compared with Western countries, where its incidence is 1 in 2 million. However, Asians in Western countries do not exhibit a high incidence.[2,3]

Although initially thought of as a disease of the young, reports on choledochal cysts in adults have increased.[4,5] The understanding that it is a different clinical entity from those found in children is slowly being realized. The exact etiology is still not fully understood and the huge variations of characteristics among pediatric patients as well as adults make it all elusive. Although a multi-factorial cause seems universally accepted, the possibility that it is an acquired disease had been tenuously considered.

We present a choledochal cyst in an adult with a previously normal biliary tree and a review of possible causes, important clinical profile of choledochal cysts in adults, as well as review of evidence and recent literature.

Case Report

The patient is a 50-year-old female who presented with right upper quadrant pain without jaundice and was diagnosed with chronic calculous cholecystitis, normal common bile duct via ultrasound (Figure 1). She subsequently underwent laparoscopic cholecystectomy with intra-operative cholangiogram (IOC). IOC was requested due to a consideration of stone or obstruction as manifested by an elevated alkaline phosphatase, however, this turned out to be normal - no filling defects, no dilated ducts, with egress of dye into the duodenum and normal pancreatico-biliary duct junction. Patient was well until nine months after cholecystectomy when she started to experience epigastric and right upper quadrant pain later accompanied by slight icterus.

On this second admission, patient was initially treated for an acid peptic disease however no relief of symptoms was noted. A whole abdominal ultrasound was then requested which revealed a dilated common bile duct, likely post cholecystectomy changes, and a septated fluid filled structure adjacent to the common bile duct (CBD), with consideration of choledochal cyst type 2 or post-operative biloma. Liver function tests were normal. Further investigation of the fluid-filled structure led to a request of magnetic resonance cholangiopancreatography (MRCP) that showed findings consistent with a choledochal cyst, type IVa, measuring approximately 3.5 x 5.2 x 6.5 cm (Figure 2). The right hepatic duct and intrahepatic biliary radicles were normal. The patient was then prepared for surgery.

A right subcostal incision with extension to the left was done. Minimal adhesions were encountered likely a result of the previous surgery. Liver was grossly normal. There was fusiform dilation of the proximal common bile duct and common hepatic duct as well as the main left hepatic duct consistent with the MRCP findings (Figure 3). A left hepatic lobectomy with en-bloc resection of the extrahepatic ducts and reconstruction via a roux-en-y right hepaticojejunostomy was performed. The patient’s post-operative course was unremarkable.
Discussion

Choledochal cysts are generally regarded as a disease of infancy and childhood. However, an increasing proportion of patients with this diagnosis are being seen in adults.[6,7] About 20% are diagnosed after 20 years of age.[8] Although it maybe related to the increasing use of diagnostic imaging,[9,10] it raises the assertion that the cause may not be always congenital.

The etiology of choledochal cysts retains obscure. One etiologic factor may be the reflux of pancreatic juice into the common bile duct, possibly due to presence of an anomalous pancreatobiliary junction (APBJ) which is frequently associated with choledochal cysts.[11,12,13] It is hypothesized that this anomaly leads to weakening of the duct wall and causes inflammatory changes within the endothelium.[14,15,16] Other causes which may be attributed to an acquired or adult-onset choledochal cyst are viral infection, most likely reovirus,[17,18,19] which was detected in 78% of patients with choledochal cysts,[16], history of previous surgery and pancreatitis. In this case, a history of previous surgery-laparoscopic cholecystectomy was noted.

Choledochal cysts in pediatric and adult patients are different in clinicopathologic manifestations, prognosis, and the underlying abnormalities of the pancreaticobiliary system, suggesting that patients with choledochal cyst should be managed according to these differences.[20] Abdominal pain is common in adults up to 90%.[2,3,21,22] and also more pronounced as compared to children.

Diagnostic imaging such as an ERCP or an MRCP, as what was done in this patient, is likewise important in planning the management of choledochal cysts since the type of intervention would be patterned to the type of the cyst as based on the Todani Modification of the Alonso-Lej Classification. Choledochal cyst type IVA as what was seen in our patient is the second most common type in adults with an incidence of 42%.[3,23]. Our findings are unique because unlike those reported by Dhupar et al[9], where three patients were diagnosed during cholecystectomy and that the patient who later presented with choledochal cysts after initial surgery was said to have been missed by ultrasound, our patient had documented evidence showing a previously normal biliary tree.

In a retrospective analysis of 32 children and 47 adults with choledochal cysts, Shah et al[24] investigated the differences between these two groups with regard to the presentation, management and histopathology. The following are the points mentioned: 1) A history of biliary surgery (5.1x more frequent in adults than in children), pancreatitis (5.4x), cholangitis (6.4x), early (2.0x) and late (3.3x) post-operative complications; 2) fibrosis of the cyst wall was peculiar to children while signs of inflammation and hyperplasia were primarily seen in adults[25], such as the histologic findings in our patient (Figure 4).

Studies now conclude that for all types of choledochal cyst, a total choledochal cyst excision with Roux-En-Y hepaticojejunostomy is the treatment of choice.[26] The most important rationale for aggressive management of choledochal cyst is the inherent risk of malignancy.[27] Cancer may grow within the cyst or its remnants and may also develop at any area along the biliary tree.[28] The risk of cancer however has not been a consistent finding in all studies. Other reports have found negligible incidence of cancer and that a lesser aggressive treatment maybe as effective but with fewer associated morbidity.[29] Complete resection of choledochal cysts have been associated with high anastomotic leak rates among adults.[30] These differences may point to a more individualized treatment approach to patients especially adults. Although choledochal cysts in adults may be regarded as a different entity, their exact etiology remains diverse. Indeed a multifactorial etiology is most likely. However, inconsistencies in various reports remain unresolved and the exact nature of this disease is far from being realized.

Viral infection and long standing inflammation have been associated with malignancy including liver cancer. This maybe shared by some population of choledochal cyst patients thus opening further our opportunity to improve our understanding of this mysterious disease with the hope of realizing why behavior of choledochal cysts varies. The exact role of inflammation as well as viral infection not only in the development of choledochal cyst but also in the subsequent development of cancer is not fully understood. Hopefully more focus into this matter will aid in fully understanding the intricacies in pathogenesis and will aid in formulating appropriate treatment options as well as monitoring protocols to address the risk of complications and malignancy.

Conclusion
A choledochal cyst, being it congenital or acquired, in itself is a rare disease entity. The increasing number of cases diagnosed in adults is likely due to the increased utilization of diagnostic imaging. However, supporting data that it is a different clinicopathologic entity among adults is increasing. In this case, we have presented another new perspective of adult-onset choledochal cyst that shows it may be an acquired lesion. A normal previous ultrasound and a cholangiogram with non-dilatation of the biliary duct and normal biliary-pancreatic duct union refute the congenital nature of this disorder. The histologic findings of ductal hyperplasia and inflammation further strengthened the theory of an adult-onset choledochal cyst.

References

Illustrations

Illustration 1

Figure 1. Ultrasound showing cholecystolithiasis with thickened gallbladder wall and normal biliary tree.

Illustration 2

Figure 2. MRCP showing 6.5 cm ovoid cystic lesion involving the biliary tree as well as cystic dilatation of the common hepatic duct and proximal common bile duct.
Illustration 3

Figure 3. Diagrammatic representation of intra-operative finding showing extent of the choledochal cyst which involved almost the whole extrahepatic bile duct as well as the left hepatic duct.

Illustration 4

Figure 4. Histology showing dilated duct with hyperplasia and inflammatory infiltrates consistent with choledochal cyst.
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