Case Report

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Combined type IV A and type III choledochal cysts associated with autosomal dominant polycystic kidney disease in an adult

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ABSTRACT

Combination of different types of choledochal cysts are very rare, seldom reported in literature. Their association with polycystic kidney disease has been never reported before. Caroli's disease is the most commonly associated fibrocystic disease with polycystic kidney disease. Here, we present an a middle aged woman, in whom, evaluation for chronic dyspepsia led to diagnosis of Type IVa and Type III (Choledochocele) associated with polycystic kidney disease, a combination that has never been reported before.

Keywords: Choledochal cyst, Biliary tract, Polycystic kidney disease, Cholangitis, Choledochocele

INTRODUCTION

Choledochal cysts (CC) are rare diseases affecting the biliary tree. Komi et al. classified CCs based on anomalous unions of the biliary tree, while the first anatomical classification was provided in 1959 by Alanso-Lej and co-workers. 1,2 Their classification divided CCs into 3 types, which was further modified by Todani and colleagues into 5 types with subtypes, which is shown in (Table 1) along with their distribution. Todani, further classified Type IVA CCs into cystic-cystic, cystic-fusiform and fusiform-fusiform types for clarifying intrahepatic and extrahepatic morphologies.3 Caroli disease associated Differentiating with extrahepatic CC is difficult. Some advocate that saccular dilatation of intrahepatic bile ducts and diffuse fusiform extrahepatic bile duct dilatation < 3 cm is more likely to be Caroli disease, rather than Type IVA CC. Sarris and co-workers further subdivided Choledochocele (Type III CC) into 5 types based on the cyst's relation with ampulla of water and the pancreatic duct. In their classification, Type A lesions were those in which intramural bile duct opened into cystically dilated segment,

communicated with duodenal lumen through a separate orifice and Type B lesions had a bile duct that opened normally into duodenal lumen with the Choledochocele arising as a diverticulum of the intra-ampullary common channel. Kagiyama et al. described additional Choledochocele variants (Type C to E) based on varying pancreaticobiliary anatomy. 5,6 Another form of CC, proposed by Lilly and co-workers called as 'form fruste choledochal cyst presents with classical features of CCpain abdomen and obstructive jaundice without biliary tree dilatation, but with abnormal pancreaticobiliary duct junction. These patients have similar histological evidence of inflammation and malignancy potential as that of CC. The most common symptom in adults with CC is abdominal pain (90%) with jaundice and nausea (40% to 50%) being the next common presentation. Autosomal polycystic kidney disease (ADPKD) is the most common inherited nephropathy characterized by slowly growing multiple renal cysts, leading to progressive renal dysfunction. It is associated with extrarenal conditions like polycystic liver disease, cerebral aneurysms, abdominal hernias, valvular heart disease or colonic diverticula and rarely, Caroli disease. It is caused by mutations in two genes, PKD-1 (polycystin-1, 85%) and PKD-2 (polycystin-2, 15%) of cases. It is diagnosed in a patient with family history of ADPKD, in whom, ≥ 3 cysts (unilateral or bilateral, age 15-39 yrs), ≥ 2 cysts (in each kidney, 40-59 yrs), ≥ 4 cysts (in each kidney, > 60yrs) or < 2 cysts (for diagnosis of exclusion, > 40 yrs) is seen on screening ultrasonography. Here we present the case of a middle aged lady, who underwent routine evaluation for dyspeptic symptoms and was diagnosed to have rare combinations of CC associated with polycystic kidney disease, a consortium never described in literature before. ^{7,8}

CASE REPORT

A 53 year old housewife visited our outpatient department in the month of February 2013, with complaints of dyspeptic symptoms associated with bloating and abdominal discomfort, ongoing since 1 year, worsening since 2 months. She denied history of jaundice, abdominal pain, progressively increasing abdominal masses or fever with chills and rigors. She has never had any hospital visits before and was consuming over the counter medications in the form of proton pump inhibitors for her symptoms. She did not have any known comorbidities or family history of chronic diseases, except for her mother, who had diabetes mellitus, hypertension and had died of a cerebral event many years ago. On examination the patient was oriented times three and without icterus or palpable masses on abdominal examination. Initial evaluation on ultrasonography, done elsewhere revealed polycystic kidneys with dilated common bile duct and centrally dilated intrahepatic biliary radicles. The blood investigations, including complete haemogram, liver function test, tumor markers, CA-19.9, carcinoembryonic antigen and alpha fetoprotein remained within normal limits, except for kidney function test that revealed a serum creatinine of 1.22 mg/dl (normal 0.2-1). We performed magnetic resonance imaging (MRI) of the abdomen along with magnetic resonance cholangiopancreatography (MRCP), findings of which are described in (Figure 1). A diagnosis of choledochal cyst type IVA in combination with type III, associated with polycystic kidney disease was made. The patient did not have grave consequences that are associated with CC. She was offered regular follow up as a modality of management with medical management and early surgical intervention on a need be basis More than a year after follow up, the patient remains asymptomatic with repeat imaging showing no interval changes as compared to prior imaging.

DISCUSSION

Babbit's theorem suggest that malformation of pancreaticobiliary system is an important cause of CC mostly, the presence of a long common channel with anomalous insertion. He proposed that reflux of pancreatic enzymes into bile duct resulted in continued inflammation, weakening of ductal wall and cystic dilatation of the biliary tree. This theory, however fails to

explain isolated ductal dilatations seen with Type III and Type V CC, where in sphincter of Oddi dysfunction is proposed to play a role. 9,10 Tung and colleagues reported an association of Type III CC with Type IVA CC. In their case report, a 69 year old female with recurrent episodes of cholangitis underwent total excision of extrahepatic CC with Roux-en-Y hepaticojejunostomy, leaving Choledochocele intact. The patient died of a duodenal ulcer perforation and abdominal sepsis on 35th postoperative day. Gupta and colleagues described the occurrence of Choledochocele (type A3) with Caroli disease in association with diverticula of the CBD, while Kimura and coworkers described 3 cases of multiple extrahepatic diverticulae associated with disease. 11-13 Kamevama et al. described the presence of Type I and Type II CC in combination in their series of patients with an incidence of 1.1%. All these patients had a fusiform Type IC CC with Type II diverticulum arising from middle part of the cyst and cystic duct draining into the right portion of the diverticulum. Similarly, Loke et al described 4 other cases of CC in which diverticular cysts of cystic duct were seen. The authors called for a separate classification for this novel type of CC association. 14,15

Association of ADPKD with Caroli disease even though rare, is a known entity. 16 Hasegawa and colleagues in 1999 and Kim and coworkers in 2011 described the occurrence of ADPKD with choledochal cyst (both Type I). In the former study, resection of choledochal cyst was done even in the absence of biliary complications. Ryu et al. also described the association of ADPKD with choledochal cyst Type I is a 60 year old female I whom, biliary tree involvement was incidentally detected on evaluation for chronic kidney disease. ^{17-19.} In the presence of cholangitis, cholangiolar abscess formation, biliary choledocholithiasis, stricture, spontaneous perforation peritonitis and pancreatitis that recurs, complete excision of cyst and reconstruction of the biliary drainage system has been advocated as the surgical treatment of choice. Carcinomatous transformation within the cyst wall is another dreaded complication. Repetitive medical management has no role in treatment of this entity and development of secondary biliary cirrhosis and portal hypertension could be an ultimatum that needs prevention. Early surgical management has been proposed by experts for ease of surgery and prevents future complications, the main aims of surgery being excision of cyst wall and gall bladder reducing risk of malignancy, halting pancreaticobiliary reflux and reconstruction of biliary enteric channel for adequate bile drainage. Choledochocele are best treated by resection or drainage into the bowel lumen. Other CCs are better managed with resection because of high risk of malignancy in this group; but this decision needs to be tailor according to patient's age, co-morbid illnesses, symptoms and type of choledochocele (type A with local excision, ampullary preservation and type B with excision and sphincteroplasty).²⁰ Regarding Type IVA CC, excision of the extrahepatic part of the cyst and drainage of intrahepatic part by wide hilar or subhilar anastomosis has been shown to provide satisfactory results, but even with finesse, complications related to residual intrahepatic part of disease still occurs on in the long term.

TABLE 1: Choledochal cysts, their characteristics and distribution.

Choledochal cysts and their distribution			
TYPE I Dilatation of entire common hepatic or duct or segments of each	A	Cystic dilatation of common bile duct	50-80%
	В	Focal segmental dilatation of distal common bile duct	
	С	Fusiform dilatation of both common hepatic and bile ducts	
Type II		ticulum from the non bile duct	2%
Type III	Choledochocele, cystic dilatation of intra duodenal portion of the common bile duct		1.4% to 4.5%
Type IV Multiple dilations of the biliary tree	A	Multiple intra hepatic and extra hepatic ductal dilatations	15% to 35%
	В	Multiple dilatations of extra hepatic biliary tree only (string of beads or bunch of grapes appearance)	
Type V	A	Multiple saccular cystic dilatations of intra hepatic bile ducts	20%
	В	In association with congenital hepatic fibrosis Caroli Syndrome	

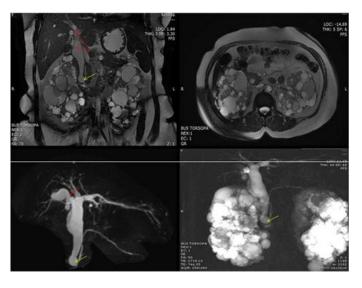


Figure 1: MRI and MRCP imaging showing fusiform dilatation of the common bile duct (2.2 cm in maximum diameter) with associated central intrahepatic biliary radical dilatation with dilated proximal left and right hepatic ducts (Type IVA CC, red arrows) with attendant triple confluence formation by the right, segment II/III and IV ducts. There is also a bulbous dilatation of the distal end of the common bile duct within the pancreas s/o Choledochocele (Type III, yellow arrows); the cystic duct is also mildly tortuous and dilated.

In our patient, the classical features of CC were not present and the diagnosis of CC, that too the rare Type III + Type IVA combination (which is described only once, in world literature) was found to be associated with ADPKD, an association never described in literature before. The management of such patients who present without biliary symptoms and/or complications are largely unknown. In our case, we decided to follow up of the patient and intervened only when there is a need for surgical measures arose. More than a year after diagnosis, the patient remained asymptomatic and repeat imaging revealing no interval changes from the initial diagnosis. The risks of complication, especially malignant transformation is well known in this group of patients, especially the ones with Type IVA CC. But, at a time when the patient has a go quality of life, is there a rationale behind early intervention in the asymptomatic period to prevent a probable future complication and making the patient go through a morbid surgery such as one on the biliary tract. These are management issues that need to be answered in this rare group of patients, who remain asymptomatic.

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Institutional Ethics Committee

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