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A Case of Adult Choledochal Cyst with Unusual Symptom

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Abstract: ***Choledochal cyst is a rare but significant disease affecting Asian population with inherent malignant potential. It has been a major burden to our society. A choledochal cyst is defined as a congenital or acquired anomaly affecting the biliary tree. It involves the dilation of extrahepatic and/or the intrahepatic segments of biliary tree. Early diagnosis and management are critical. A clear regional variation exists for choledochal cysts, where two-thirds of the reported cases in Asia occur in Japan. type I and IV occur more commonly in females and have a female to male ratio of 4:1 or 3:1 The reason for the Asian and female predominance remains unknown. The presentation and therapeutic strategies for choledochal cysts in adult may differ from that of childhood. The surgical management of choledochal cysts in adults is complicated by associated hepatobiliarypathology. Clinical diagnosis including classical trial of abdomen pain, jaundice, righthy pochondrant mass seen only 0-17% of population and mostly in children, majority of cases present with very vague clinical presentation and 40% enter adulthood unnoticed. This case highlights the unique presentation of choledochal cyst in 47 year old female with postprandial dizziness and epigastric pain.***

Keywords: Adult choledochal cyst, post prandial giddiness, stasis, hypovolemia

1. Introduction

A choledochal cyst is defined " as a cystic dilation of the bile duct". In 1959 ***Alonso-LEJ et al[1]*** were the first to classify CC into 3 types based on the site of the biliary duct dilation. In 1977 when ***Todani et al[2]*** modified the classification by adding 2 types.

Type I choledochal cyst

1) Account for 80-90% of all bile duct cysts
2) Characterized by fusiform dilation of the extrahepatic bile duct
3) A Sub Classification has been proposed
   - **Ia:** dilatation of extrahepatic bile duct (entire)
   - **Ib:** dilatation of extrahepatic bile duct (focal segment)
   - **Ic:** dilatation of the common bile duct portion of extrahepatic bile duct

Theorized to form as the result of reflux of pancreatic secretions into the bile duct via an **Anomalous pancreaticobiliary junction**. Some believe them to arise from ductal plate anomalies.

Type II

Also known as a bile duct diverticulum. Account for 3% of all bile duct cysts. Saccularoutpouchings, representing a true diverticulum, arising from the supraduodenalextrahepatic bile duct

Type III

Also known as a choledochocele:

- Account for 5% of all bile duct cysts
- Represent protrusion of a focally dilated, intramural segment of the distal common bile duct into the duodenum

Choledochoceles may be successfully managed with endoscopic sphincterotomy, surgical excision, or both, in symptomatic patients.

Type IV

Multiple communicating intra- and extrahepatic duct cysts:

1) Second most common type of bile duct cysts (10% of the total)(most common in adults)
2) subdivided into subtypes:
   - **Type IVa:** fusiform dilation of the entire extrahepatic bile duct with extension of dilation to the intrahepatic bile ducts(most common in adults)
   - **Type IVb:** multiple cystic dilations involving only the extrahepatic bile duct

Type V

Also known as **Caroli disease**, which is a rare form of congenital biliary cystic disease manifested by cystic dilations of intrahepatic bile ducts. Association with benign renal tubular ectasia and other forms of renal cystic disease.

Recent Additions to the classification

Some authors have coined a 'type VI' bile duct cyst, an entity which is considered rare and not part of the original Todani classification. This nomenclature is not widely accepted but is included for completeness.

Type VI

A rare entity describing isolated dilatation of the cystic duct. Some authors consider this description to be type VIa, while

also defining a type VIb that also involves dilatation of the common bile duct.

This five-category classification is the most commonly used by clinicians today; however, it is in dispute by some experts who claim that each type of CC has its natural course, complications, and management. It is proposed that a classification that focuses more on the pathogenesis rather than the simple anatomy of the biliary tree is recommended.

Adult choledochal cyst is a rare enitity, incidence in Asian population 1:1000, Female>Male(3:1)Nearly 25% of choledochal cysts are detected in the first year of life and 60% in the first decade, but 20% are diagnosed after the age of 20 years. Type IV A common in adults (mean age <40 years). They are frequently associated with an **Anomalous pancreatobiliary duct junction (APBDJ).** Managing Adults is of significance as they may be associated with malignancy.

Pathophysiology

Babbits Theory:
Postulated by ***Babbit et al[3]***
APBJ is when the biliary and the pancreatic duct join 1 to 2 cm proximal to the sphincter of Oddi. As a result, the long channel formed is not covered by the sphincter and allows for the backflow and mixing of pancreatic and biliary secretions, which leads to the activation of pancreatic enzymes. This results in a rise in pressure, which eventually causes dilation, inflammation, epithelial damage, dysplasia, and malignancy of the biliary tree

Drawbacks:
Babbitt's theory is challenged by authors who state that APBJ was present in only 50% to 80% of cases and that in CCs diagnosed antenatally, there wasn't the presence of reflux. In addition, they suggest that neonatal pancreatic acini are not capable of producing enough pancreatic enzymes to cause this damage

Other theories:
Some theories postulate that the etiology of this condition is purely congenital.

Kusunoki et al[4]. suggest the presence of scantier ganglionic cells in the distal common bile duct (CBD) of patients with CCs when compared to controls. This leads to the dilation of the proximal segment of the CBD, which is comparable to the pathogenesis of achalasia and Hirschsprung's disease.

Babitts and kusunoki theory holds true for Type I and IV

Regarding type II (True CBD diverticulum)is related to biliary duplications cyst and type III (choledochocele) duodenal duplications cysts.

The etiology of Carolidisease, also known as type V, is presumed to be a halt in the remodeling of the ductal plates. Caroli disease is associated with biliary atresia, Autosomal recessive Poly cystic kidney disease and less frequently with autosomal dominant polycystic kidney disease

Histopathology
The histopathology of CC depends on the patient's age. In younger patients, lymphocytic infiltration into the wall of the cyst, which is lined by columnar epithelium, is demonstrated. In addition, there is a presence of dense collagenous tissue along with bundles of smooth muscle within the wall, indicating cyst wall fibrosis. In contrast, adult cysts show evidence of mucosal inflammation and hyperplasia. There is also a variation in the histologic appearance of each type of CC. Type I and type IV CCs may show absence or patchy distribution of the biliary mucosal layer. Type II CC is difficult to distinguish from gallbladder duplication. Type III CC is frequently lined by duodenal mucosa but could uncommonly be lined by biliary mucosa. Type V CC usually demonstrates hepatic fibrosis.

Here I am reporting a single case of adult choledochal cyst with unusual symptom.

2. Case Report

A 47 year old female, labourer by occupation presented with Complaints of epigastric pain, post prandial dizziness and vomiting on &off for more than 3 months, similar episodes in past settled with medications.

History of presenting illness
The pain started at epigastricregion, dull in quality with radiation to back, present mostly 30-40 minutes after food intake, relieved by analgesic medication, it was present throughout for the past 3 months and there is no symptom free period.

Also associated with vomiting, on and off for the past three months, started after epigastricpain, bilious sometimes and not blood stained, not projectile.

Pt also had post prandial 30-40 mins after food intake giddiness and lightheadedness, with tremors and sweating of palms.

No records available of previous medications

No comorbid, No menstrual Irregularities, No childhood symptoms.

Examination

Temperature: Normal
Blood pressure: 122/68 mm of hg
Pulse rate: 92/min, regular, volume good
Random blood sugar 108 mg/dl (normal 70-110 mg/dl)

Vague epigastricmass, borders and extent could not be determined, with Tenderness. No organomegaly

Percussion and auscultatory findings were normal

Other systemic examinations were normal

Course at hospital:
Pt was found to have altered blood sugar profile, diabetologist opinion obtained and in view of suspicion of

newly diagnosed diabetes mellitus type2 insulin sliding scale started based on actrapid.pt was particularly feeling dizzy after food consumption especially 30-40 mins after food intake also with vague abdomen pain and fullness.Bp chart was maintained which is shown below along with random blood sugar chart

Investigations
Total bilirubin: 1.2 mg/dl
Direct bilirubin: 0.6 mg/dl
Sgot/sgpt/alp: normal
Amylase /lipase: normal
Mri abdomen +MRCP:

- Fusiform Dilatation of both Intrahepatic and CHD& CBD
- Abrupt narrowing in terminal CBD
- CBD Max Dia 3.7cm

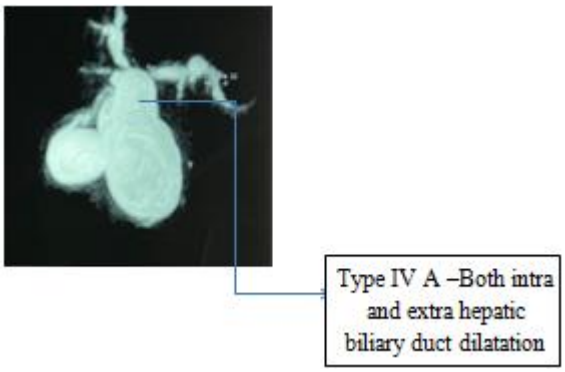

Figure 1

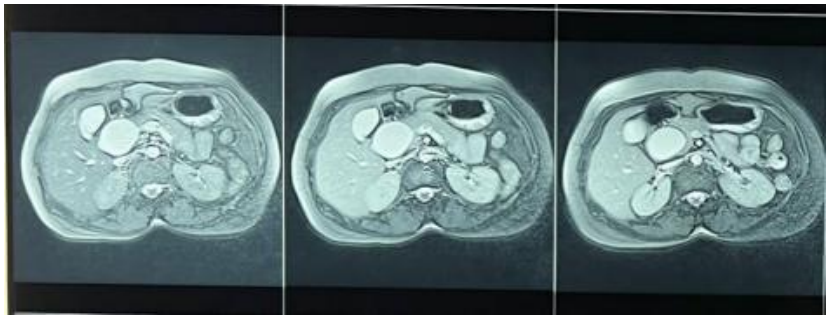

Figure 2

Random blood sugar chart
Insulin intervention (sliding scale used)→continued till pod 2

Pre-op	Day 1	Day 2	Day 3	Day 4	Post op	Day 1	Day 2	Day 3	Day 4
Pre breakfast	78mg/dl	86mg/dl	100mg/dl	96mg/dl		77mg/dl	70mg/dl	88mg/dl	75mg/dl
Post breakfast (1hr)	170mg/dl	166mg/dl	158mg/dl	158mg/dl		110mg/dl	156mg/dl	145mg/dl	150mg/dl
Pre lunch	112mg/dl	132mg/dl	106mg/dl	112mg/dl		110mg/dl	126mg/dl	110mg/dl	122mg/dl
Post lunch(1 hr)	190mg/dl	178mg/dl	204mg/dl	220mg/dl		160mg/dl	145mg/dl	140mg/dl	146mg/dl
Post lunch (2 hr)	188mg/dl	180mg/dl	220mg/dl	210mg/dl		168mg/dl	186mg/dl	162mg/dl	156mg/dl
Pre dinner	160mg/dl	170mg/dl	158mg/dl	166mg/dl		153 mg/dl	166mg/dl	160mg/dl	172mg/dl
Post dinner	200mg/dl	192mg/dl	177mg/dl	189mg/dl		170mg/dl	180mg/dl	188mg/dl	200mg/dl

Blood pressure chart

Pre op	Day 1	Day 2	Day 3	Day 4	Post op	Day 1	Day 2	Day 3	Day 4
Morning 6 am	110/60 mm of hg	112/58 mm of hg	106/56 mm of hg	110/64 mm of hg		110 mm of hg	98/60 mm of hg	112/70 mm of hg	110/70 mm of hg
Pre lunch	122/68 mm of hg	116/60 mm of hg	118/70 mm of hg	114/76 mm of hg		118/56 mm of hg	122/70 mm of hg	116/70 mm of hg	128/80 mm of hg
Post lunch	102/50 mm of hg	96/50 mm of hg	92/45 mm of hg	86/56 mm of hg		118/70 mm of hg	112/62 mm of hg	100/50 mm of hg	116/70 mm of hg

Operative Notes:

Rt sub costal incision
Fusiform Dilatation of CBD of size 3x4 cm
Hepato-duodenal ligament divided and kocherisation done
Gb removed, F/b Delineation of Cbd done
Hepatico-jejunostomy 15cm from DJ flexure done
Jejuno-jejunostomy of efferent limb done 40cm down the HJ site

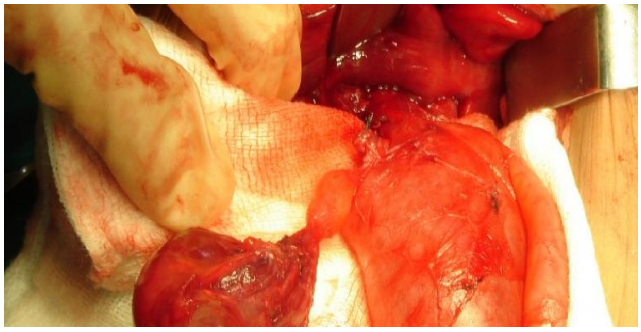

Figure 3: HPE

Specimen of cbd+chd with cystic duct and GB, showed infiltration of lymphocytes and fibroblasts suggesting chronic inflammation and fibrosis. hyperplasia not found.

Concurring diagnosis of choledochal cyst

3. Discussion

Choledochal Cyst is common in paediatric population.

Presenting in adults is a rarity, also it harbours tendency of malignancy. It should be treated with surgery as early as possible.

Excision with Biliary channel reconstruction is commonest surgery performed.

Children and adults with choledochal cyst often have different signs and symptoms. The classical triad of jaundice, right hypochondriac pain, and a palpable mass was found more commonly in children compared to adults (85% versus 25%, resp.)

In adults, the complications include stone formation secondary to bile stasis in the cyst and intrahepatic ducts, recurrent cholangitis, pancreatitis, and spontaneous cyst rupture due to raised intra-abdominal pressure as in pregnancy. In addition, the coexistent congenital hepatic fibrosis in patients with type V predisposes to portal hypertension and oesophageal varices. The reported incidence of cholelithiasis due to bile stasis is around 37.5 to 74%. Hepatolithiasis is most often noted in type IV-A and may be related to the presence of membranous or septal stenosis or segmental bile duct near main biliary convergence. Abnormal pancreaticobiliary junction in the presence of obstruction by stones or protein plug impaction predisposes to the risk of acute pancreatitis, which is reported to be seen in 30–70% of adults.

The major concern is the risk of malignant transformation, which is well documented in the literature. The whole biliary tree is considered at risk of malignant transformation and may arise either in cystic dilatation or remnant tissues after excision or in nondilated parts of the biliary tree including the gall bladder

Choledochal cyst may remain asymptomatic for many years and diagnosis may be made incidentally. A lag period of 6 years is often noted particularly in adults between the development of symptoms and diagnosis and treatment.

It may be diagnosed in asymptomatic patients undergoing health screening test, when liver function test (LFT) is found to be abnormal. Choledochal cysts can now be diagnosed at any age of life including antenatally by ultrasonography. Precise preoperative identification of the type, extent of biliary tree dilatation, and information on the pancreatic and bile duct anatomy and disease are essential to plan surgical strategy. The current “gold standard” for staging CCD is magnetic resonance cholangiopancreaticography (MRCP). MRCP has the distinct advantage of being noninvasive in nature and in its ability to assess cyst anatomy, identify size, site, and shape of bile duct dilatation, and detect APBDJ making it distinctly superior. It also avoids the risk of potential complications of pancreatitis and cholangitis associated with invasive procedures like ERCP and percutaneous cholangiography. However, the age at diagnosis of CCD is related to the development of carcinoma in the gall bladder, the cyst, or the intrahepatic ducts. In patients who have CCD at 10 years of age or younger, the risk of developing cholangiocarcinoma is approximately 1%, whereas the risk increases to 15% for patients older than 20 years of age, 26% in patients above 40 years, and 45.5% in patients above 70 years **[5, 6, 7].** The incidence of synchronous cholangiocarcinoma associated with CCD is estimated to be 2.5 to 30% and was 6% in the largest reported western series **[8, 9, 10, 11].** The histological types of cancer are adenocarcinoma (73%–84%), anaplastic carcinoma (10%), undifferentiated cancer (5%–7%), squamous cell carcinoma (5%), and others (1.5%) [31]. The locations of the cancer are extrahepatic bile ducts (50–62%), gall bladder (38%–46%), intrahepatic bile ducts (2.5%), liver (0.7%), and pancreas (0.7%) **[12].**

Surgery for CCD has evolved, both in the timing of surgery and in the type of surgery carried out **[6-8, 13–16]**. The current approach involves control of biliary sepsis and pancreatitis and defining both the superior and inferior extent of the cyst, before scheduling surgery in semielective setting. Inadequately prepared patient may lead to technically difficult operative field due to adhesions to adjoining structures.

4. Operative Technique

Following the cyst excision the hepaticoenterostomy can be carried out by 2 types of anastomosis: hepaticoduodenostomy or Roux-Y hepaticojejunostomy **[17].** In general, the success of an anastomosis is measured by the ease of performing it and the short- and long-term complications. The reported success of hepaticojejunostomy is 92% with complication rate of 7% compared with complication rate of 42% following hepaticoduodenostomy **[17, 18].** Hepaticoduodenostomy is not recommended by some, because of the reported complications (33.3%), which include bilious gastritis due to duodenogastric bile reflux and adhesive bowel obstruction and cholangitis. In addition, increased risk of gastric cancer (due to bile reflux) and biliary cancer has been reported **[18].** On the other hand, there are others who are proponents of hepaticoduodenostomy because of its simplicity, being quicker to perform, and importantly preservation of normal anatomy and physiology and minimum complications.

Here in our case we performed Hepaticojejunostomy and post operativept recovered well. In relation to here presenting symptoms vague abdomen pain and giddiness settled after surgery.

Proposed pathophysiology:

2 factors were found during this case

1) Patient was not a known diabetic and she was newly diagnosed during her admission to the hospital, as with random blood sugar chart, mostly her post prandial sugar levels were significantly higher probably because

of associated sub clinical pancreatitis with islet cell dysfunction which were not documented and no further investigations pertaining to that was done. So her dizziness could probably due to altered blood sugar levels

2) Her post prandial blood pressure dropped significantly>20mm of hg systolic Probable theories are associated gastroparesis and slow motility of gastro duodenal region due to inherent parasympathetic (vagus nerve) disorder associated with choledochal cyst **[4, 19] oligoganglioma theory with bile duct stenosis causes stasis and gastro paresis which results in poor emptying and motility of gastro duodenal junction and upper thirds of 2nd part of duodenum causing sequesteration of fluid from circulating blood resulting in hypovolemia it is further supplemented with patients constitutional symptoms like tremor, giddiness and excessive sweating of palm getting alleviated by intravenous crystolloids where 0.9% NACL and Ringer lactate were used.**

Post operatively patient was stable and recovered well, was discharged and followed up at outpatient basis for 6 months.she had no cecurrence of her symptoms and she was currently discharged with followup to diabetologist.

5. Result

As of now no direct studies have been conducted to substantiate our findings, in futureit may be possible.

Based on our findings we claim that the probable reason for her giddiness during post prandial period could be due water sequestration and hypovolemia.

Large patient pool and further studies including neuroimaging would be of particular value in this field

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