Diverticular Choledochal Cyst from Cystic Duct (TYPE VI VARIANT)

Dr. Seema Rohilla[1], Dr. Deepak Singh[2], Dr Rohtas Kanwar Yadav[3], Dr Anand Rai Bansal[4], Dr Manju Bala[5]

ABSTRACT

Choledochal cysts are the cystic dilatation of bile ducts. The widely used Todani classification of choledochal cysts does not mention the choledochal cysts at cystic duct location. Only a few cases of choledochal cyst at the location of cystic duct have been reported and termed as type VI choledochal cysts. We report a case of diverticular form of choledochal cyst attached to cystic duct diagnosed preoperatively by Magnetic resonance cholangiopancreatography (MRCP) and Hepatobiliary iminodiacetic acid (HIDA) scan and confirmed by surgery and histopathology in a young female. This is a variant of type VI choledochal cyst.

Key words: Choledochal cyst, Cystic duct, Todani classification, Diverticular choledochal cyst.

[1]Professor, [2]Junior Resident, [3]Senior Professor & Head
Department of Radio Diagnosis, Pt.B.D.Sharma PGIMS Rohtak India
[4]Professor, Professor, Department of Surgery, Pt.B.D.Sharma PGIMS Rohtak India
[5]Assistant Professor, Department of Anaesthesiology, Pt.B.D.Sharma PGIMS Rohtak India
Corresponding author mail: – deepakphogat266@gmail.com

Conflict of interest: None

INTRODUCTION

Choledochal cyst is a congenital dilatation of the extrahepatic biliary tract with or without dilatation of the intrahepatic biliary tract. Choledochal cyst in the region of cystic duct is a rare entity and is not included in the widely accepted Todani classification of choledochal cysts. Only a few cases of cystic duct dilatation are reported. Diverticular type of choledochal cyst attached to the cystic duct is probably a variant of type VI choledochal cyst; one such case is being reported here.

CASE REPORT

Twenty two years old female had pain in right hypochondrium off and on for six months without fever or jaundice. On examination there was fullness in right hypochondrium but no tenderness. On investigation hematological tests and liver function tests were normal.

On ultrasound (US) a diagnosis of double gall bladder (GB) without calculus was made, as a cystic structure at gall bladder fossa region adjacent to the GB was seen. Thick slab coronal oblique magnetic resonance cholagiopancreatography (MRCP) showed a cystic structure with a filling defect attached through a diverticulum with a narrow neck to the cystic duct (Figure 1).

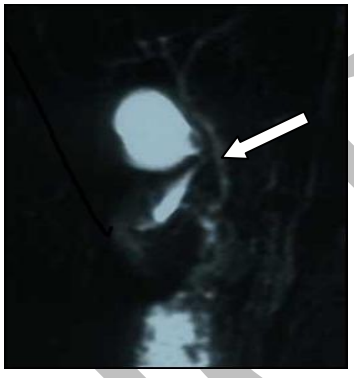

Figure (1): Thick slab coronal oblique magnetic resonance cholagiopancreatography (MRCP) image showing a cystic structure with a filling defect attached through a diverticulum with a narrow neck to the cystic duct

Hepatobiliary Iminodiacetic acid (HIDA) scan showed two structures in GB fossa taking up the tracer after two hours of tracer administration (Figure 2).

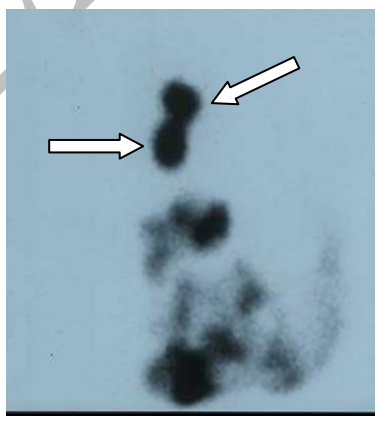

Figure (2): Hepatobiliary Iminodiacetic acid (HIDA) scan image showing two structures in GB fossa taking up the tracer after two hours of tracer administration.

Three hours after meal, one of these structures still showed the tracer and was diagnosed as choledochal cyst while the other showed total wash out of tracer diagnosed as GB (Figure 3).

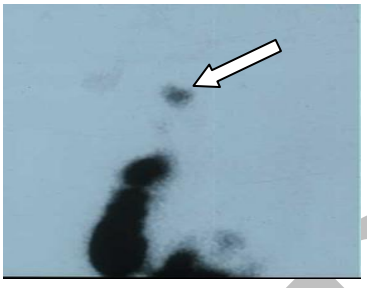

Figure (3): HIDA Scan image three hours after meal, one of these structures still showed the tracer and was diagnosed as choledochal cyst while the other showed total wash out of tracer diagnosed as GB

Hence radiologically, it was suggested to be a diverticular choledochal cyst from cystic duct (? a variant of type VI choledochal cyst), a very rare occurrence. Patient was operated and the radiological findings were confirmed. Histopathology confirmed one structure to be GB and other choledochal cyst. The patient is well one year after surgery. Patient had first episode of pain in September 2012 and presented in March 2013 and was operated in April 2013.

DISCUSSION

Choledochal cyst is a congenital dilatation of the extrahepatic biliary tract with or without dilatation of the intrahepatic biliary tract. Estimated prevalence of choledochal cysts varies from one in 13 000 to one in two million. They are rare in the west and far more common in Japan and Asia. Highest incidence is reported from Japan. The male female ratio is around 1:4.[1]

The commonly used classification was given by Todani. This was a modification of the primary classification given by Alonoso – Lej.[2] As per Toadni classification: type I is a cyst of the common bile duct; type II is a cystic diverticulum coming off the common bile duct; type III is a cyst in the intraduodenal portion of the common bile duct; type IV refers to multiple cysts in the intra, and extrahepatic biliary tract; and type V are single or multiple cysts in the intrahepatic ducts alone.[3, 4]

The exact etiopathology of choledochal cysts is not clear. Two theories are most prevalent. Babbitt et al has proposed the

theory of anomalous pancreaticobiliary junction (APBJ) with a long common channel, resulting in reflux of pancreatic juice into the biliary tract, with resultant inflammation, scarring and biliary dilatation. This can explain type I, III and IV cysts but does not explain type II and V cysts where the common bile duct (CBD) is normal. Some believe that these cysts are congenital due to distal aganglionosis and proximal dilatation explaining the type II and V cysts as well.[1, 5]

Todani classification does not mention the choledochal cysts of cystic duct .First mention of such a cyst was done by Serradel et al. They called it type VI choledochal cyst.[6] Since then only few cases of type VI choledochal cyst have been reported.

Although most reported cases of type VI choledochal cysts were diagnosed intraoperatively Yoon et al diagnosed three cases of such cyst preoperatively by CT and MR.[7] De at al also reported a type VI choledochal cyst as an isolated dilatation of cystic duct in an 18 year old female diagnosed preoperatively and confirmed surgically.[5] Maheshwari et al reported 10 cases of type VI choledochal cysts in 2012. Four out of 10 cases were saccular dilatation of cystic duct and six patients had fusiform dilatation of cystic duct[1]. Convay et al reported an isolated case of cystic duct cyst.[8]

Most studies described the type VI choledochal cyst as a dilatation of cystic duct itself. Loke et al described a variant of this type of choledochal cyst in a 29 years old female patient .They reported a case having a diverticular type pouch containing multiple calculi which joined the common bile duct with large communicating orifice and also communicated with the cystic duct through a wide communication.[9] This was a variant of choledochal cyst type VI as described by Serradel et al.[6] The difference in this cyst and type II cyst was the opening of cystic duct in the pouch.

Some authors attribute type VI choledochal cyst to APBJ. Initial case reports of choledochal cysts of the cystic duct do not discuss APBJ.[1, 5] Weiler et al and Noun et al described an association of choledochal cysts of the cystic duct with APBJ.[1] The etiology of choledochal cyst of cystic duct due to APBJ was also described by De et al as they reported acute angulation of the common hepatic duct (CHD) and cystic duct junction, together with a wide opening of the cystic duct and APBJ, triggers the reflux of

pancreatic juice. The mixing of pancreatic juice and bile as well as stasis results in cystic duct ectasia. The reason for isolated involvement of the cystic duct excluding the CBD was unclear. Possibly the junction of the cystic duct with the CBD being the weakest part due to the least vascularity, causes an ectatic change that continues as a vicious cycle, resulting in further dilatation. This in turn increases the angulation of the already abnormal CHD and cystic duct junction, thereby increasing reflux into the already dilated sac and causing mass effect on the CHD and CBD.[5] Focal aganglionosis akin to Hirschsprung disease could also result in isolated dilatation of the cystic duct. No conclusive etiology has been established. The exact etiology remains debatable, although some authors favor congenital etiology over APBJ as far as choledochal cysts of the cystic duct are concerned[1]

Our case is different from all the reported cases discussed, as in our case the pouch like cystic structure was having a narrow diverticulum connected to the normal cystic duct. The cystic duct CHD junction as well as common pancreatico biliary segment was also normal. In our case focal aganglionosis could be the more appropriate etiology as the cystic duct itself was spared and the pouch like structure could be due to focal weakness because of aganglionosis. The cystic duct and gall bladder were normal. Microlothiasis have been reported in choledochal cysts in 8% cases.[5] In our case choledochal cyst had a filling defect seen on MRCP due to calculi. Nuclear studies showed delayed washout of tracer. Surgical exploration confirmed the radiological findings and the choledochal cyst was excised. On histopathology the cystic structure was confirmed as choledochal cyst. Our case is probably a variant of type VI choledochal cyst which itself is very rare entity.

We suggest that double GB, which is seen more frequently, should be kept as a differential of diverticular choledochal cyst. The double GB if asymptomatic, does not need surgical correction. US of the fasting patient followed by US after fatty meal may help to differentiate the two conditions as a double GB will contract whereas diverticular choledochal cyst from cystic duct will maintain its size and shape. Tracer studies could then be done for confirmation.

On imaging, GB shows a well defined pear shape while choledochal cysts are slightly

globular and maintain their shape on scans done at different times.

REFERENCES

1. Maheshwari P. Cystic malformation of cystic duct: 10 cases and review of literature. World J Radiol. 2012 Sep 28;4(9):413–7.
2. Bode WE, Aust JB. Isolated cystic dilatation of the cystic duct. Am J Surg. 1983 Jun;145(6):828–9.
3. Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K. Congenital bile duct cysts: Classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. Am J Surg. 1977 Aug;134(2):263–9.
4. Chan ES, Auyang ED, Hungness ES. Laparoscopic Management of a Cystic Duct Cyst. JSLS. 2009;13(3):436–40.
5. De U, Das S, Sarkar S. Type VI choledochal cyst revisited. Singapore Med J. 2011 May;52(5):e91–93.
6. Serena Serradel AF, Santamaría Linares E, Herrera Goepfert R. Cystic dilatation of the cystic duct: a new type of biliary cyst. Surgery. 1991 Mar;109(3 Pt 1):320–2.
7. Yoon J-H. Magnetic resonance cholangiopancreatography diagnosis of choledochal cyst involving the cystic duct: report of three cases. Br J Radiol. 2011 Jan; 84(997):e018–e022.
8. Conway WC, Telian SH, Wasif N, Gagandeep S. Type VI biliary cyst: report of a case. Surg Today. 2009;39(1):77–9.
9. Loke TK, Lam SH, Chan CS. Choledochal cyst: an unusual type of cystic dilatation of the cystic duct. AJR Am J Roentgenol. 1999 Sep;173(3):619–20.

Acknowledgement: We would like to thank Mr Jai Singh, Mr Harminder and Mr Arvind all three working as MRI Technicians in Department of Radio diagnosis Pt.B.D.Sharma PGIMS Rohtak India for their help in conducting this study.

Financial Disclosure: None