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A Rare Case Of Anomalous Ampulla Of Vater In Third Segment Of Duodenum Associated With Cholangitis And Gall Bladder Carcinoma

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ABSTRACT

Anomalous opening of ampulla of Vater into the third segment of the duodenum is an extremely rare congenital abnormality, which can be complicated by cholangitis, choledocholithiasis and recurrent duodenal ulcers. Here we, report a case of 50 years old female patient, who presented with complaints of pain in abdomen, fever and symptoms & signs of obstructive jaundice. Though, ERCP (Endoscopic Retrograde Cholangiopancreatography) is essential for the diagnosis of this entity, we diagnosed this case on Ultrasonography, MDCT (Mutidetector Computed Tomography), MRCP (Magnetic Reasonance Cholangiopancreatography) and Endocsopy. Endoscopy clearly showed dilated CBD (Common Bile duct) opening abnormally into the third part of the duodenum along with MPD (Main Pancreatic duct). In our case, there was cholangitis and gall bladder carcinoma associated, as diagnosed very well on CT and MRCP.

Keywords: Anomalous opening, Ampulla of Vater, Cholangitis, Gall bladder carcinoma.

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INTRODUCTION

The major papilla of Vater is normally located on the posteromedial wall of the second part of duodenum, formed by opening of both CBD and MPD surrounded by longitudinal and circular muscles that comprises sphincter of Odii. Sometimes, the ampulla of Vater can be located in aberrant sites along the duodenum, mainly within the, pyloric channel, duodenal bulb or the fourth portion, and it has also been found with a much lower frequency in the third segment. We present a case of ectopic opening of ampulla of Vater into the third segment of duodenum associated with cholangitis and gall bladder carcinoma, which is a very rare association reported till date.

CASE-REPORT:

A 50 year old female patient visited the surgical OPD with complaints of abdominal pain and fever. No significant previous history was present. Physical examination revealed scleral icterus and raised body temperature at the time of admission. The majority of initial lab findings were within normal limits except for the deranged liver function tests. Aspartate transaminase of 161 IU/L, alanine transaminase of 196 IU/L, alkaline phosphatase of 184 IU/L, total bilirubin of 6.5 mg/dL, and direct bilirubin of 4.0 mg/dl. USG revealed an echo complex mass in the gallbladder fossa region and adjacent liver parenchyma, CBD was dilated with few echo dense shadows at terminal part and appeared to open at the third segment of the duodenum, which was not very clear on USG. An abdominal CT scan was advised which showed an abnormally enhancing mass in fundus of GB,

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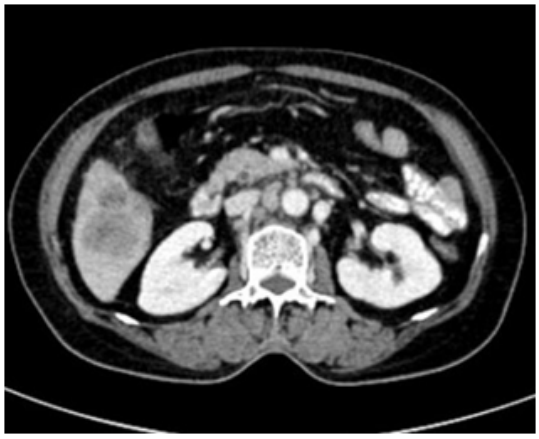

Fig.1-Axial CECT image showing heterogeneously enhancing mass involving fundus of gall bladder and adjacent liver parenchyma with dilated CBD.

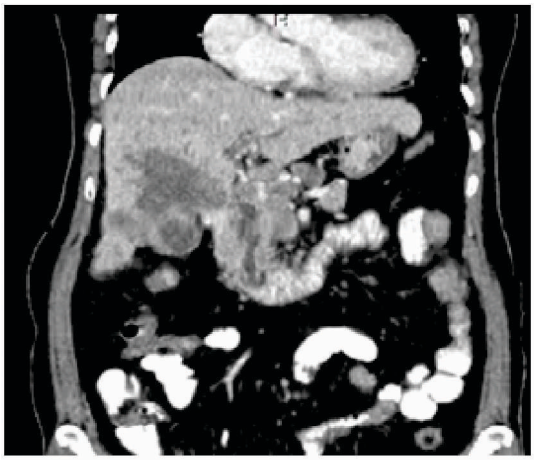

Fig.2 - CECT coronal reformat image shows the dilated CBD opening into third segment of duodenum

infiltrating the adjacent segment of liver (Fig.1). CBD was dilated with thickened enhancing wall and appeared to open in the third segment of duodenum along with main pancreatic duct (Fig.2). MRCP was also done which confirmed the diagnosis made on MDCT (Fig.3). Finally, endoscopy was done which showed that there was no opening in the second segment of duodenum, instead Ampulla of Vater was opening abnormally into the third segment (Fig.4).

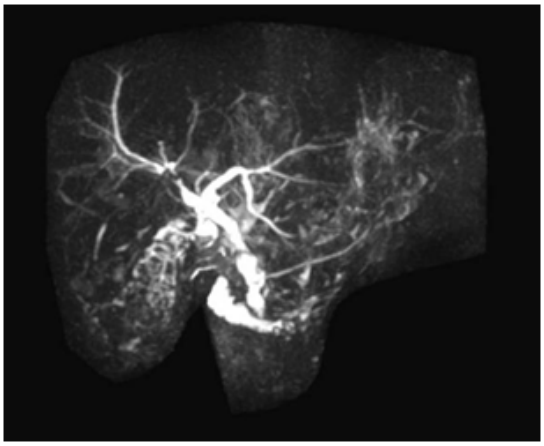

Fig.3 - MRCP 3D reformate image, showing CBD and MPD opening in third segment of duodenum

Fig.4-Upper GI Endoscopy image showing ampulla of Vater in the third segment of duodenum

Though, ERCP is confirmatory for diagnosis of anomalous opening of ampulla of Vater, but it could not be done in our case because of severe symptoms of cholangitis in the patient. Unfortunately, we lost the patient during follow up and hence treatment is not discussed here.

DISCUSSION:

Ectopic location of ampulla of Vater in the stomach or first, third, and fourth part of the duodenum is a very rare congenital anomaly and has a frequency rate of 5.6% to 23%. This wide range is due

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to the limited number of cases, but this condition has been reported more frequently due the extensive availability of ERCP.[1] The exact etiology of this condition remains unclear, however, it has been ascribed to the defect in embryogenesis. The liver originates from the primitive hepatic furrow, which, during embryogenesis, is divided into pars hepatica and pars cystica. The pars hepatica then develops into both liver and hepatic ducts, while the pars cystica develops into the gallbladder and cystic duct. The CBD originates in hepatic antrum, which is the common area of the hepatic furrow. According to the hypothesis of Boyden, anomalous drainage of the CBD occurs due to disproportional elongation and early subdivision of the primitive hepatic furrow.[2,3] Previous reports have shown diverse complications of ectopic papillae, including gallstone, choledocholithiasis, obstructive jaundice, cholangitis, pancreatitis, and peptic ulcer due to anomalous biliary drainage, biliary stasis and poor sphincter control mechanism.[4] In addition, this biliary stasis or reflux may play central role in the process of carcinogenesis, probably as a result of a sequence of pancreatobiliary reflux, inflammation, dysplasia with or without intestinal metaplasia, and invasive carcinoma. Though, there is no definite report on association of ectopic opening of CBD and gall bladder carcinoma till date but biliary malignancy was more common in patients with P-C type of pancreatico-biliary maljunction PBM without dilated CBD, as compared with those with other types of PBM, according to one study;[5] furthermore, it has been hypothesized that persistent stasis of bile and pancreatic juice may cause hyperplasia, metaplasia, and dysplasia of the ductal epithelium. Tanno et al[6] found that stasis of pancreatic juice occurs exclusively in the GB in PBM without a dilated CBD, and Funabiki et al reported that the incidence of GB cancer was higher in PBM without a dilated CBD, as compared with PBM with a dilated CBD. In the present case, no communicating duct was visualized between the common bile and main pancreatic ducts on MDCT or MRCP. The mechanism of carcinogenesis in this case is different from that for anomalous pancreatico-biliary junction. However, the presumption hypothesis of GB cancer with the ectopic opening of CBD may be chronic inflammation of the bile duct. Reflux of intestinal contents and bacteria into the bile duct may cause recurrent cholangitis in such patients because they lack a sphincteric barrier between the bile duct and the intestine, and chronic inflammation can be a predisposing factor for gallbladder cancer. Furthermore, the bile acid fraction contains carcinogenic substances such as lysolecitine and taurodeoxycholic acid.[7] Further studies are needed to reveal the pathogenesis of malignant disease in ectopic biliary drainage. Although plain upper GI barium X-ray, intraoperative cholangiography,[8] MDCT,[9] endoscopic ultrasonography have been used as the diagnostic tools, ERCP is the "Gold standard". Fistula secondary to ulcer or choledocholithiasis, spontaneous or iatrogenic surgical fistula, and surgical choledochoenteric diversion should be included in the differential diagnosis.[10]

An ectopic papilla of Vater in the third segment of duodenum increased the difficulty of performing therapeutic interventions during ERCP. This anomaly has a high risk of perforation or bleeding during endoscopic sphincterotomy because the intramural portion of the duct is not fully developed. Therefore, balloon dilatation may be the technique for removal of the stones. However, if the insertion of a balloon catheter into an ectopic opening is difficult due to the acute

angulation of the distal CBD and deformity of the bulb, stent installation or surgery may be necessary. Surgical treatments included choledocho-enterostomy, surgical stone extraction by choledochotomy and rarely, pancreato-duodenectomy. However, 13%-20% of patients who received these endoscopic procedures experienced recurrent cholangitis.

CONCLUSION:

Ectopic papilla of Vater in the third segment of duodenum increases the difficulty of performing therapeutic interventions during ERCP. This anomaly has a high risk of perforation or bleeding during endoscopic sphincterotomy because the intramural portion of the duct is not fully developed. Thus, customized treatment is needed for each patient, and long-term efficacies of endoscopic and surgical treatments should be compared.

Support: Nil.

Conflicts of interest: None.

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