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A SURPRISE INTRUDER IN THE WALL OF THE CHOLEDOCHAL CYST - A NOSTALGIC BOND, A CASE REPORT AND REVIEW OF LITERATURE.

Gastroenterology

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ABSTRACT

Choledochal cyst is a rare congenital malformation of the biliary tract which can be associated with calculi within the cyst – cystolithiasis and gallstones. It is unusual to find heterotopic pancreatic tissue within the wall of choledochal cyst. We hereby report a case of a 43 year old female, who presented with complaints of recurrent abdominal pain with fever for the past 2 months. Magnetic resonance cholangiopancreatogram (Plain) showed features of acute/subacute cholecystitis. Focal cystic dilatation of proximal common bile duct with choledocholithiasis, type Ib choledochal cyst, Hepatolithiasis with mild intrahepatic biliary radicals dilatation were noted. The patient was planned for cholecystectomy with excision of choledochal cyst. Microscopic examination of the cystwall showed a fibrocollagenous cyst wall lined by single layer of columnar epithelium. The cystwall showed lobules of pancreatic acini consistent with heterotopic pancreatic tissue. Gall bladder showed features of chronic cholecystitis. We report this case of heterotopic pancreatic tissue in choledochal cyst with cystolithiasis and cholelithiasis occurring simultaneously and due to its rarity.

KEYWORDS

pancreatic, heterotopia, choledochal, cyst, acini.

INTRODUCTION:

Choledochal cyst is a rare congenital malformation which can be associated with calculi within the cyst – cystolithiasis and gallstones[1]. It is unusual to find heterotopic pancreatic tissue within the cyst wall. A 43 year old female presented with complaints of recurrent abdominal pain with fever for the past 2 months. Magnetic resonance cholangiopancreatogram (Plain) showed features of acute/subacute cholecystitis. Focal cystic dilatation of proximal common bile duct with choledocholithiasis, type Ib choledochal cyst and hepatolithiasis with mild intrahepatic biliary radicals dilatation were noted. The patient was planned for cholecystectomy with excision of choledochal cyst. Microscopic examination of the cystwall showed fibrocollagenous wall lined by single layer of columnar epithelium. The cystwall showed lobules of pancreatic acini in the cystwall consistent with heterotopic pancreatic tissue. We report this case of heterotopic pancreatic tissue in choledochal cyst with cystolithiasis and cholelithiasis occurring simultaneously due to its rarity.

Case History:

A 43 year old female presented with complaints of recurrent abdominal pain with fever for the past 2 months. Magnetic resonance cholangiopancreatogram (Plain) showed features of acute/subacute cholecystitis. Focal cystic dilatation of proximal common bile duct with choledocholithiasis, type Ib choledochal cyst and hepatolithiasis with mild intrahepatic biliary radicals dilatation were noted (Fig 1&2). The patient was planned for cholecystectomy with excision of choledochal cyst. Microscopic examination of the excised cyst showed a fibrocollagenous cystwall composed of lined by single layer of columnar epithelium. The cystwall showed lobules of pancreatic acini in the cystwall consistent with heterotopic pancreatic tissue (Fig 3-5). Focal pyloric gland metaplasia was noted. Gallbladder showed features of chronic cholecystitis with pyloric gland metaplasia. There was no evidence of dysplasia or malignancy. Common bile duct and common hepatic duct resection margins was unremarkable with no evidence of dysplasia or malignancy. Post operative period was uneventful. On follow up, suture removal was done on POD (Postoperative Day 14) and liver function tests were normal.

DISCUSSION:

Choledochal cyst (CC) is a rare congenital malformation characterised by cystic dilatation of the biliary tract and was reported by Vater and Ezler in 1723[1]. Heterotopic tissue is presence of normal tissue that is located in an abnormal location and not directly continuous with the main organ with no vascular continuity between the heterotopic tissue and the main tissue. Heterotopic pancreas is defined as presence of ectopic pancreatic rests - acini, islets and ducts or any of those which is located elsewhere other than the usual anatomical location of pancreas which lacks anatomic and vascular continuity with the main pancreas[2]. Heterotopic pancreas can be seen at any site of the gastrointestinal tract, mostly in the stomach, duodenum, jejunum, oesophagus, terminal ileum, omentum, spleen, mesentery, liver, colon, Meckel's diverticulum, gallbladder, urinary bladder, ampulla of Vater, lung and rarely in lymph nodes[3]. The incidence of heterotopic pancreas is 0.5% to 14.0% in autopsy studies. Presence of heterotopic pancreatic tissue in the wall of choledochal cyst is quite rare and has been detected incidentally during autopsies (0.11%- 0.21%)[4]. The incidence of Choledochal cyst is highest in South East Asia. It has a marked female preponderance with M: F (Male: Female) ratio of 1: 5. Presence of heterotopic pancreatic tissue is usually asymptomatic but sometimes they can cause bleeding, intussusception, ileus and malignancy in adults. Heterotopic pancreatic tissue can be subtyped according to the Heinrich classification, Type 1 is characterised by the presence of ducts, acini and islets of Langerhans cells, Type 2 comprises of acini and ducts, but islets are not seen and Type 3 is composed only of pancreatic ducts. Peribiliary glands can be identified adjacent to the heterotopic pancreatic tissue in some cases which might indicate a common embryological origin. Different postulates have been suggested as to the origin of heterotopic pancreatic tissue in the wall of choledochal cyst. It could be developmental or could arise from totipotent endodermal stem cells. During development, dorsal and ventral pancreatic buds arise as outgrowths from primitive foregut. The ventral bud rotates and passes behind the duodenum from right to left and fuses with the dorsal bud in the 7th gestational week. The posterior head and uncinate process is derived from the ventral bud. The anterior head, body and tail are derived from the dorsal bud. During this process, ectopic rests may sequester from the dorsal bud and get implanted to other sites and develop as heterotopic pancreatic tissue. It could be an atavistic phenomenon[4]. The other hypothesis is that the gut totipotent endodermal stem cells may differentiate into pancreatic tissue in different parts of intestine[5]. Nakanuma et al. noted increased expression of endodermal stem/progenitor cell markers along the peribiliary glands in extra-hepatic biliary channels. Nakanuma et al. proposed that embryologically and anatomically biliary tract may represent an incomplete pancreas[6].

Though rare pancreatitis, Islet cell tumor, pancreatic cysts and pancreatic adenocarcinoma can occur in heterotopic pancreas with few case reports of such cases. Ogata H et al., had described 12 cases of Heterotopic pancreas in different parts of gastrointestinal system[7]. Pragya sharma et al. identified heterotopic pancreatic tissue in 13 out of 200 cases of surgically excised choledochal cysts (6.5%)[8]. Radiological investigations are not much useful in the detection of Heterotopic pancreas as they are tiny foci and doesnot show any specific diagnostic feature most of the times. However, on CT scan,

they can appear as a small oval intramural mass with microlobulated margins and an endoluminal growth pattern. The attenuation and enhancement characteristics of these lesions depends on the histologic components present in Heterotopic pancreas. After intravenous contrast material administration, acinus-dominant lesions demonstrate avid homogeneous enhancement whereas duct-dominant lesions appears hypovascular and heterogeneous. On MRI, the heterotopic pancreas is isointense to the orthotopic pancreas with T1 hyperintensity and early avid enhancement after intravenous gadolinium-based contrast material administration[9]. Detection of heterotopic pancreatic tissue requires diligent histological examination of the entire excised choledochal cyst. Treatment of type 1choledochal cyst includes Roux-en-Y hepaticojejunostomy with complete excision of the cyst.

CONCLUSION:

Though rare, heterotopic pancreatic tissue can be seen in the wall of choledochal cyst and histopathological examination never fails to fascinate us explaining the nostalgic embryological bond with regards to the location of heterotopic pancreatic tissue. This case is reported due to its rarity and occurrence of choledochal cyst with cystolithiasis simultaneously. Even complete removal of the cyst is not preventive against malignancies because malignancies may arise elsewhere along the biliary tract. The prognosis is usually good but long term follow-up is necessary due to increased risk of development of malignancy.

Figures

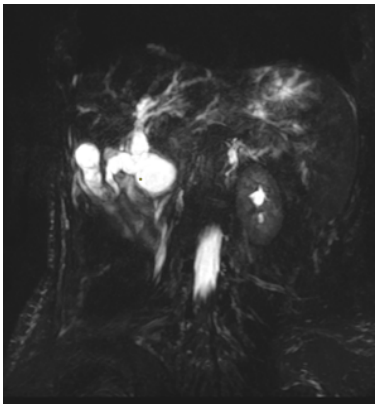

Fig 1: Coronal MIP* MRCP image showing focal cystic dilatation of proximal common bile duct with choledocholithiasis - Type Ib choledochal cyst.
*Maximum intensity projection (MIP)

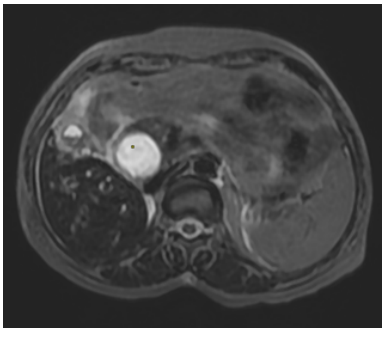

Fig 2: Axial MRI T2 sequence shows focal cystic dilatation of proximal common bile duct with choledocholithiasis - Type Ib choledochal cyst.

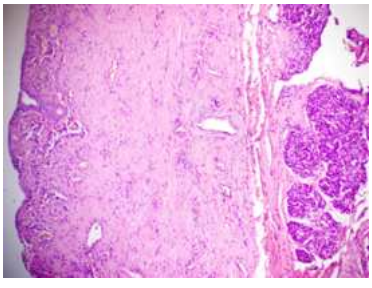

Fig 3: Biopsy showing fibrocollagenous cystwall with lobules of pancreatic acini in the cystwall (H&E at 40x)

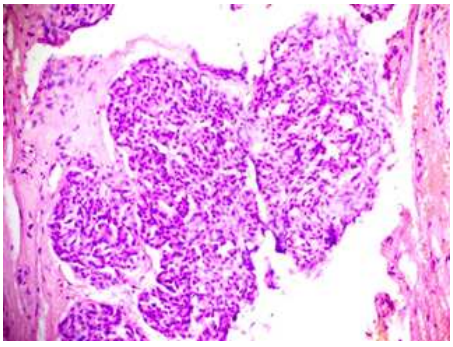

Fig 4: Biopsy showing lobules of pancreatic acini in the cystwall (H&E at 100x)

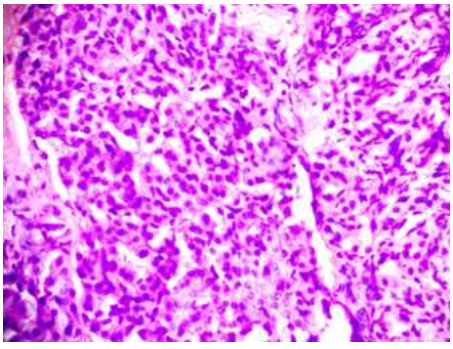

Fig 5: Biopsy showing lobules of pancreatic acini (H&E at 200x)

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