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Synchronous Pyloric Gland Adenoma Of The Gall Bladder And Choledochal Cyst In A Child: A Case Report

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Abstract:
Background*: Choledochal cyst is not uncommonly seen in the pediatric population. Long-standing choledochal cysts can lead to malignancy. However, concurrent association with a neoplasm in the gall bladder is seldom seen. The pyloric gland adenoma of the gall bladder is a rare entity. Synchronous association of both poses an embryologic, diagnostic, and therapeutic challenge.*
Case Presentation*: We describe a 7-year-old female child with a history of intermittent pain abdomen for two years. On evaluation was found to have a fusiform choledochal cyst along with cystic dilation of the gall bladder. The patient underwent excision of the gallbladder and choledochal cyst alone with biliary reconstruction. The histopathology was suggestive of a choledochal cyst and a synchronous pyloric gland adenoma of the gall bladder. The patient was kept on follow-up and had a favorable outcome with surgery alone.*
Conclusion: *Pyloric gland adenoma of the gall bladder in children can be safely managed with surgery alone. An association with choledochal cyst should be kept in mind and if present, both can be surgically excised in the same sitting with biliary reconstruction with good outcome.*
Key Word*: Gall bladder neoplasm; pyloric gland adenoma; choledochal cyst; gall bladder adenoma; pediatric gall bladder neoplasm.*

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I. Introduction

Gall bladder (GB) neoplasms are an uncommon subcategory of biliary tract tumors. They are rarely seen in the pediatric age group. No definite pre-existing risk factors that lead to the development of GB neoplasms have been described. A choledochal cyst (CDC) is a common entity in the pediatric age group, which can undergo malignant transformation in the common bile duct. However, most of the cases of tumors in long-standing CDC have been described in adults, and that too in the region of the common bile duct. Embryologically, abnormal pancreaticobiliary maljunction could explain the etiopathogenesis of both CDC and GB neoplasm[1,2]. The association of a gall bladder neoplasm in the setting of a choledochal cyst has been rarely described in the literature. A child with a synchronous neoplasm in the GB with an associated CDC has not yet been reported in the literature and is described.

II. Case Presentation

History and physical examination: A 7-year-old female patient presented with a complaint of intermittent pain in the abdomen for two years. There was associated nausea and occasional non-bilious vomiting. There was no associated pyrexia, weight loss, jaundice, diarrhea, or constipation. There were no other previous co-morbidities. On examination, the abdomen was soft and there was no distension, tenderness, guarding, or rigidity.

Investigations: An ultrasonogram of the abdomen revealed a dilated common bile duct, distended gall bladder, and mildly dilated intrahepatic biliary radicles. The liver function test was ordered. Serum bilirubin was normal, but there was mild elevation of serum transaminases (SGOT: 79.5u/L, SGPT: 70u/L). Magnetic resonance cholangiopancreatography revealed a fusiform dilatation of common hepatic and common bile ducts. The dilatation was just proximal to the junction with the main pancreatic duct and anomalous pancreaticobiliary junction. There was evidence of sludge with microlithiasis within the dilated CBD. A mild prominence of central IHBR was also seen. The findings were suggestive of type 1 CDC (Figure 1).

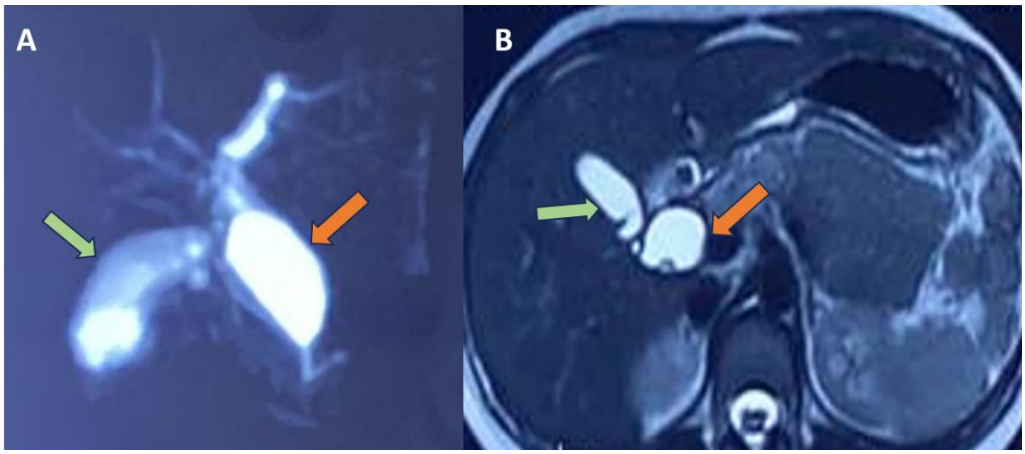

Figure 1: MCRP depicting cystic dilation of the gall bladder as well as fusiform dilatation of common bile duct with distal tapering (A) and (B).

Operative treatment: The patient was planned for surgical excision and biliary reconstruction. A subcostal incision was made and the anatomy was delineated. There was a gross dilation of the gall bladder, cystic duct, and common bile duct. There was no associated vascular variation. Moderate adhesions were encountered suggestive of previous episodes of possible cholangitis. Cyst excision with hepatico-duodenostomy was done. Histopathology revealed papillary changes throughout the gallbladder suggestive of Pyloric gland adenoma (PGA) of the gall bladder and CDC (Figure 2).

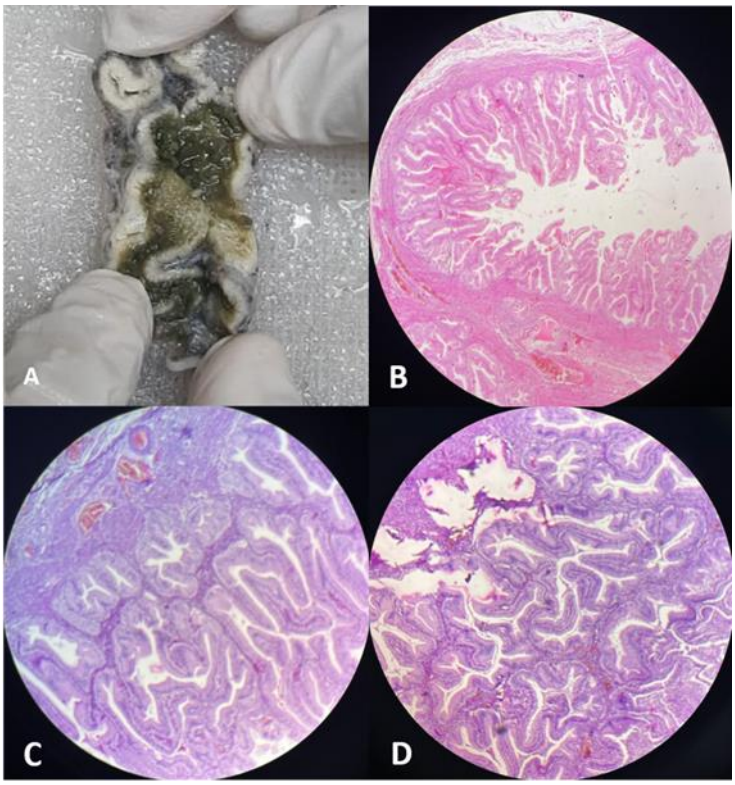

Figure 2: Gross specimen (A) with gall bladder wall infolding and irregularity. Low power microscopy (Hematoxylin and Eosin staining, under 10x magnification) suggestive of tightly packed pyloric glands (B). Histopathology depicting pyloric gland adenoma with tightly packed mucinous glands showing copious apical mucin (C) and (D) (Hematoxylin and Eosin stain, under 100x magnification).

Outcome: The child had an unremarkable post-operative period and was discharged on full orals on postoperative day 6. The patient was kept on regular follow-up. A repeat ultrasound of the abdomen done after one year was unremarkable. The patient was doing fine on follow-up at two years.

III. Discussion

Dyslipidemia Primary PGA of GB is quite rare with less than a dozen cases reported in literature[3]. The incidence is much higher in middle-aged females. No case has yet been reported in the pediatric age group, nor any case has been reported associated with a CDC in a child. This is the first reported case in the literature of PGA of the GB in a child with a CDC.

As per the World Health Organization Classification of Digestive System Tumours (2018 Edition), it is listed as a benign neoplasm of the gall bladder. It is considered under a blanket terminology of intracholecystic papillary-tubular neoplasm[4]. PGA is an uncommon benign malignancy with most of the cases occurring in the stomach. Rarely it is seen in extra-gastric locations such as the gall bladder, bile duct, cystic duct, pancreatic duct, esophagus, duodenum, gastroesophageal junction, rectum, and cervix of the uterus[5,6]. A similar neoplasm based on the presence of ovarian stroma has been classified as Mucinous cystic neoplasm (MCN) [7]. In the absence of ovarian stroma, intracystic papillary neoplasms of the GB are also the preferred terminology.

There can be a wide range of clinical presentations ranging from asymptomatic patients to those with abdominal pain, vomiting, and jaundice. There can be vague abdominal and constitutional symptoms or rarely it is incidentally detected post-cholecystectomy. Rarely it can also present as obstructive jaundice with bile duct obstruction[3]. As expected, laboratory results are also varied with occasionally raised liver enzymes and serum bilirubin.

An association with choledochal cyst in the present case implores the study of the embryology of the biliary tract. Ectopic remnants of embryonal GB tissue and aberrant hamartomatous bile ducts could be a causative factor in the origin of PGA[8]. The association of cystadenoma with mesenchymal stroma as reported by Wheeler et al. denotes a dysplasia in the genesis of the biliary tract wall, like the embryological aetiological factor in the pathogenesis of choledochal cyst[8]. PGA arising contiguously from GB and cystic duct implores the possibility of involvement of the biliary tract during embryogenesis[9].

Ultrasonography is a good first-line investigation that might delineate the anechoic, thickened irregular walls with papillary infolding. A more definitive diagnosis can be made using either computed tomography or Magnetic resonance tomography. However, the gold standard of diagnosis remains histopathology. A diffuse cytoplasmic positivity of MUC6 (a pyloric marker) with occasional focal staining of MUC5A and MUC1 has been described[4]. However, these markers are non-specific and do not confer any pragmatic diagnostic or prognostic significance.

Preoperative diagnosis is difficult, most cases are diagnosed by histopathology. Surgical removal is advisable, especially considering the risk for malignant transformation in gall bladder polyps of size greater than 10mm or 15mm, though the exact cut-off remains controversial[10]. No such reports or data is available for pediatric cases. Surgical resection in the form of laparoscopic or open cholecystectomy is recommended. Differentiation between benign and malignant cysts is difficult with imaging, however, in PGA complete surgical excision is curative[6]. In case of an associated choledochal cyst, a hepatico-duodenostomy or hepatico-jejunostomy along with the removal of GB is necessary. Close follow-up is necessary as there is a definite risk of recurrence and they also have a risk of malignant transformation to cystadenosarcoma or cystadenocarcinoma.

IV. Conclusion

Benign neoplasms of the GB though rare, can still be found in the pediatric age group. It should be kept in mind that these can be associated with choledochal cyst. Surgical excision is curative with a good outcome without any need for any adjunct therapy. Repeated evaluation on follow-up is paramount as there is a risk for malignant transformation.

Abbreviations

CDC: Choledochal Cyst
GB: Gall bladder
MCN: Mucinous cystic neoplasm
MRCP: Magnetic resonance cholangiopancreatography
PGA: Pyloric gland adenoma

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