



Type IV A Choledochal Cyst in A Young Female: A Rare Case Report

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Abstract

Choledochal cysts are rare congenital anomalies of biliary tract. There are 5 subtypes in which type IV constitutes 15-35% of all choledochal cysts. Here we report a case of 25 year old female who came for evaluation of primary infertility with complaints of mass and pain abdomen. Imaging techniques showed a Type IVA choledochal cyst which was subsequently operated and send for Histopathological examination. The microscopy of gall bladder showed choledochal cyst with gangrenous changes of gall bladder and bile duct.

Keyword: Choledochal cyst, Cholangiocarcinoma, Todani classification

Introduction

Congenital bile duct malformations known as choledochal cysts are abnormal, disproportionate cystic dilations of the biliary channel [1].It is prevalent among Asian and female polulations and the incidence is

1:100000-150000. Among the different types only 15-35% of all choledochal cysts are Type IV cyst [3]]. The classic traid includes pain, jaundice and an abdominal mass [1]]. The exact tiology remains unknown but it is believed to be congenital in origin [4].We present a case of 25 year old female who came for evaluation of primary infertility with mild abdominal pain and mass in the upper right side of abdomen. MRCP with MRI showed gross dilatation of bilateral hepatic ducts, common hepatic duct and fusiform diltation of common bile duct. The excision of common bile duct with hepaticojejunostomy was done. The gross dimensions of the specimen were same as mentioned in scan. Microscopy of gall bladder showed areas of ulceration with gangrenous changes, the cyst was composed of fibrocollagenous stroma with lining denuded and the bile duct showed fibromuscular stroma with lining denuded

at most places and focal columnar cells. No features of malignancy was noted.

Case report

A 25 year old female came for the evaluation of primary infertility with mass and pain in the upper right side of abdomen. Physical examination showed a firm mass in the upper abdomen sized 2 cms. On percussion the mass was continuous with liver dullness. Blood investigation revealed increased Total White blood cells count with normal liver function test. MRCP with MRI revealed gross dilatation of bilateral intrahepatic biliary radicals ,maximum diameter of right hepatic duct 22mm, left hepatic duct 20mm, common hepatic duct 24 mm and gross fusiform dilatation of common bile duct maximum diameter 35mm with narrowing of distal end .Excision of Common bile duct with hepaticojejunostomy was done and the specimen was send for for histopathological examination. The gross dimensions of the specimen were the same as mentioned in the radiological finding. Microscopy of gall bladder showed ulcerated mucosal lining with gangrenous changes, the cyst wall was composed of fibrocollagenous stroma with the lining denuded as most places and focal columnar cells . The fibrotic wall of the cyst had extensive areas of hemorrhage with foci of lymphocytes, adjacent pancreatic parenchyma of normal morphology was noted. The bile duct showed fibromuscular wall lined at places by columnar cells with focal areas of gangrene. No atypia or malignancy were noted.



Figure 1 a: (White arrow)Gall bladder with fusiform dilated common bile duct (Purple arrow)

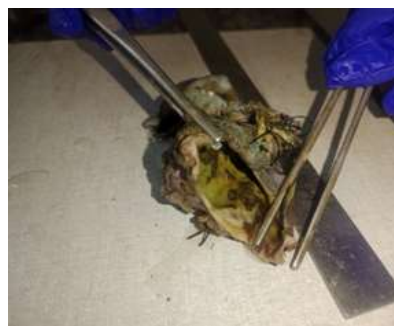


Figure 1b: The ulcerated mucosal lining of common bile duct

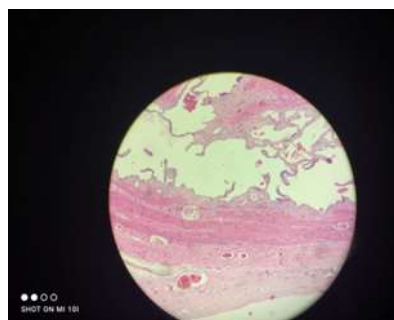


Figure 2: Microcopy of gall bladder shows ulcerated mucosal lining with focal gangrenous changes

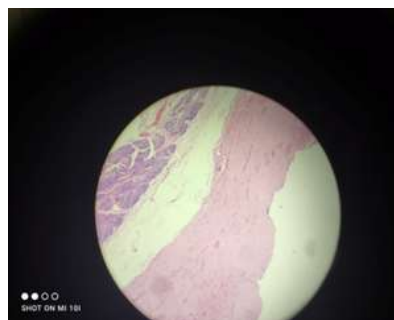


Figure 3: Microscopy of dilated common bile duct shows cyst wall composed of fibrocollagenous stroma devoid of lining cells. The fibrotic wall has areas of

hemorrhage. Adjacent regions shows normal pancreatic parenchyma

Discussion

Choledochal cysts are uncommon dilatation of the biliary tree affecting the extrahepatic and intrahepatic segment or both at single or multiple sites. Patients are usually diagnosed during childhood but in 20-25% cases the diagnosis is delayed until adulthood [9]. In Todani classification Type IV A displays intra and extrahepatic dilatation of the biliary ducts whereas it is less common in Type IVB. In Todani classification Type I A (cystic dilatation of CBD), Type I B (saccular dilatation of the CBD), Type IC (fusiform dilatation extending to the common hepatic duct). Type II bile duct cysts are extrahepatic duct diverticula. Type III is cystic dilatation of the intraduodenal portion of common bile duct. Type IV A as a common bile duct cyst combined with intrahepatic bile duct dilation and Type IVB as multiple extrahepatic bile duct cysts without intrahepatic bile duct dilation. Type V is isolated intrahepatic cyst disease (Caroli disease) [1]. The excision of the extrahepatic component is preferred treatment for Type IV choledochal cyst. The management of the intrahepatic portion of Type IV choledochal cyst remains controversial [7]. However the combined malignancy rate was 10.9 % with cholangiocarcinoma being the most prevalent [10]].

Conclusion

It is important for timely diagnosis of choledochal cysts as these are rare and may not present with classic triad of symptoms. Also a significant proportion of patients who underwent resection for choledochal cyst can harbor malignancy. Cancer risks seem reduced but not eliminated with complete resection which remains the standard treatment.

References

1. Prathik Bhattarai, Bishwdeep Timilsina, Prasun Khanal, Type IV A choledochal cyst with choledocholithiasis in an adult female: A case report, Wiley Clinical case reports, 20 September 2023
2. Vicky S. Budipramana, Putu Ayu Saraswati, Type VI choledochal cyst: A case report, Case report, International Journal of surgery case reports 2020, pg 111-114
3. Wasiq Bin Tariq, Anu Radha Twayana, Neela Sunuwar, Case report: A rare case of choledochal cyst [Version 1: peer review: 2 approved] JFIOO Research 2022, 11:919
4. Utpal Anand, Rajeev N Priyadarshi, Bindev Kumar, A giant type IVA choledochal cyst, Annals of Gastroenterology, (2012) 25, pg 73-75
5. Chantelil Iambaudi Razafindrato, Andry Liliana Rina Rakotozafindrabe, Type IV A choledochal cyst: A rare cause of cirrhosis in adults, The Egyptian journal of Internal medicine, 2022, 34:7
6. Grebennikov, Sarah DO, ET AL, Type 4 Choledochal cyst: Biliary dilation with concomitant stricture presenting a diagnostic and treatment challenge, The American journal of gastroenterology, Oct 2023, s1536
7. Sakthivel Harikrishnan, Servarayan Murugesan Chandramohan, Apsara Chandramohan, Giant choledochal cyst type 4A: A surgical challenge, Pan African medical journal 2020
8. James S Banks, Gaurav Saigal, Joseph M. D'Alonzo, Choledochal malformations: Surgical Implications of Radiologic findings, Gastrointestinal imaging. Review, April 2018

9. Adrian Miron, Liliana Gabriela Popa, Elena Adelina Toma, The curious case of the choledochal cyst- Revisiting the Todani classification : Case report and Review of the literature, *Diagnostics*, March 2023
10. Grace C Bloomfield, Aradhya Nigam, Inochi Gonzalez Calvo, Characteristics and malignancy rated of adult patients diagnosed with choledochal cyst in the West: a systematic review, *J Gastrointestinal Surg*, Jan 2024