A Rare Case of Congenital Choledochal Cyst (CCC) Resulting in Recurrent Acute Pancreatitis in an Otherwise Healthy Young Woman

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Introduction

Congenital choledochal cyst (CCC) is a rare cystic dilatation of intrahepatic or extrahepatic biliary ducts. Clinical presentation is often vague with the most common symptoms being abdominal pain, fever and jaundice making diagnosis difficult. Appropriate treatment is crucial since CCC carries a high risk of malignant transformation. We present a case of a type IVb choledochal cyst presenting as recurrent acute pancreatitis in a young healthy female with initial negative screenings.

Case Presentation

An 18 year-old-female with a history of Covid-19 presented to the emergency department with one month of persistent abdominal pain, nausea, and vomiting. She was hospitalized once prior for similar symptoms and was diagnosed with acute pancreatitis. This admission, blood work showed elevated lipase, elevated liver enzymes, mild bilirubinemia with a normal lipid panel and urine was significant for infection. She received fluids, antiemetics and was started on prophylactic antibiotics for ascending cholangitis. A right upper quadrant ultrasound ruled out cholelithiasis or acute cholecystitis but showed dilation of the common bile duct. MRCP confirmed dilation with bulbous termination in the periampullary region diagnosed as Type IVb choledochal cyst.

References

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Results

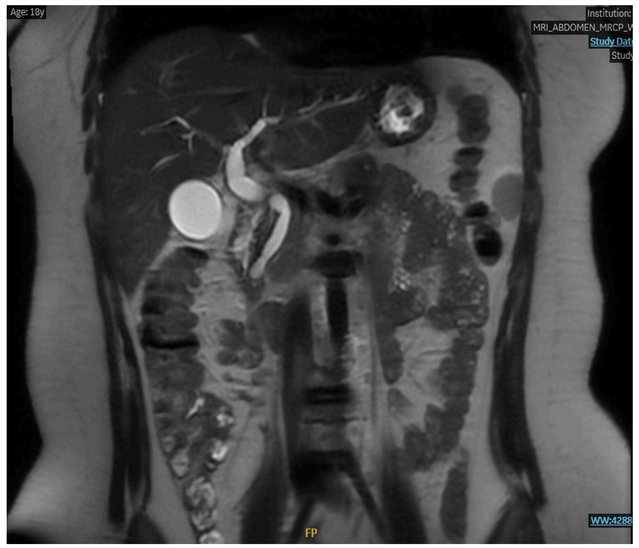

Figure 1: MRCP without contrast demonstrates a dilatation of the common hepatic duct and the common bile duct measuring up to 1.2cm. There is a short segmental narrowing in the mid duct terminating in a bulbous, dilated appearance of the periampullary duct, compatible with Type IVb choledochal cyst.

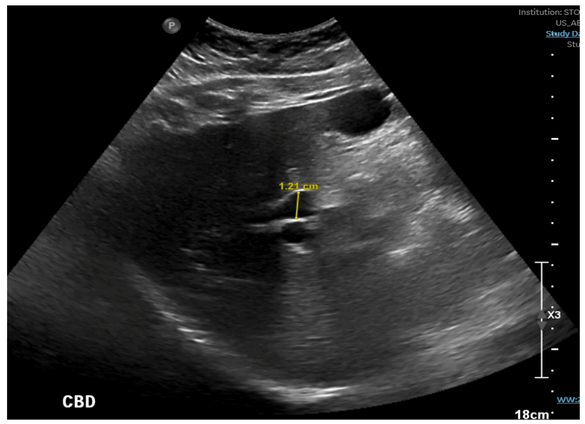

Figure 2: US of the abdomen that shows a distal dilated common bile duct approximately measuring 1.2cm.

Discussion

CCCs are rare in Western countries with an incidence between 1 in 100,000 to 150,000. 80% of these cysts are diagnosed in patients under the age of 10. They are difficult to diagnose due to variable clinical presentations. A study of 214 CCC patients demonstrated the most common symptom was abdominal pain, followed by jaundice and fever. When cysts are found in adults, symptoms resemble atypical acute biliary tract disease. Surgical cyst removal may be needed for patients with significant risk factors such as older age and age of symptom onset, due to increased risk of malignant transformation. Longer periods of observation have been documented to be associated with an increased chance of developing late complications, such as anastomotic stricture, biliary calculi and recurrent cholangitis. Type IVb CCCs, as seen in this case, consist of multiple extrahepatic cysts and hepaticojejunostomy is the treatment.

Conclusions

This patient's young age and recurrent acute pancreatitis combined with her lab and imaging findings strongly suggest the diagnosis of CCC. The anatomical location of the CCC impeded flow of pancreatic enzymes through the ampulla of Vater, leading to recurrent pancreatitis in an otherwise healthy young female. CCC, although very rare, should be considered in the differential of acute pancreatitis when other causes such as gallstones and heavy alcohol consumption cannot be identified, as prompt diagnosis and surgical removal is imperative.

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