A Young Female With Acute Cholangitis Due to Type IVa Choledochal Cyst With Undifferentiated Spindle and Giant Cell Carcinoma Hiding Inside

Ryan Burkholder MD [1] Nahel Tunio MD [1] Kelly Zhang MD [2]
Kevin El Hayek [2] Marc Landsman MD [1]

1- Division of Gastroenterology and Hepatology; 2- Department of Hepatobiliary and Foregut Surgery
MetroHealth Medical Center at Case Western Reserve University School of Medicine, Cleveland, OH

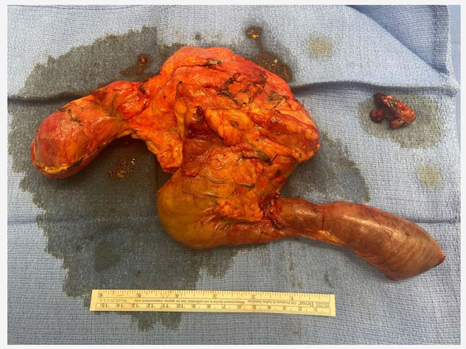
Abstract

We present a young female patient who presented with new-onset abdominal pain and jaundice, who was found to have a large type IVa choledochal cyst.

On pathology, she was found to have an undifferentiated spindle and giant cell carcinoma. This is exceedingly rare in young patients, but has a better prognosis than pancreatic or gallbladder malignancy since it manifests sooner with clinical manifestations.

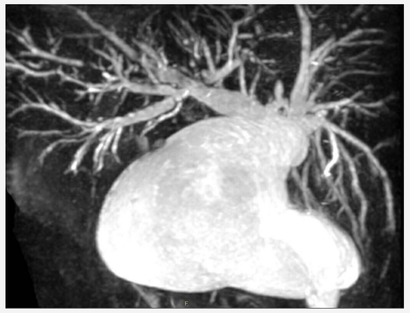
Case Presentation

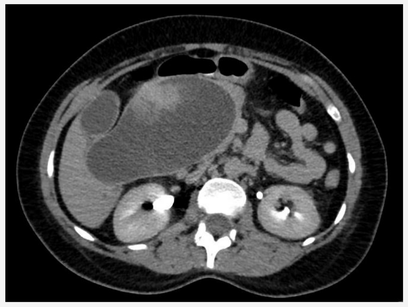
A 29 year old female with no significant medical history presented with 2 days of sharp epigastric pain, radiating to the back associated nausea and non-bilious vomiting. Patient's exam noted scleral icterus and abdominal tenderness to palpation in the right upper quadrant. The patient's labs showed a white blood cell count of 26,300. Liver enzymes aspartate aminotransferase (AST) 114, alanine aminotransferase ALT 289, alkaline phosphatase of 343 and total bilirubin of 9.76.



Case Presentation

Computed tomography (CT) of abdomen showed dilated proximal extrahepatic CBD with a large complex cystic mass in the porta hepatitis measuring 10 cm x 9 cm x 8 cm and contiguous with gallbladder and proximal common bile duct, with mass effect on duodenum and pancreatic head, concerning for choledochal cyst. Given suspicion for ascending cholangitis, an endoscopic retrograde cholangiopancreatography (ERCP) showed a significantly dilated CBD with cystic dilation. Balloon sweep with 9-10-12 mm balloon catheter was performed with passage of stone, pus and bile. Two plastic stents (10 Fr x 7 cm and 10 Fr x 9 cm) were placed into the bile duct with appropriate bile drainage.
The patient failed to improve, with severe sepsis secondary to acute cholangitis, and developed worsening fevers to 101.5 degrees F. Total bilirubin increased to 14.2 and alkaline phosphatase reached 535. Patient was taken for laparoscopy which showed very large choledochal cyst intimately involving head of the pancreas and duodenum, necessitating a Whipple procedure for distal margins. The proximal margin entered both the left and the right hepatic ducts consistent with a type IVa choledochal cyst. Anatomic pathology results of the 7.0 cm x 6.0 cm x 3.0 cm mass in the cyst showed an undifferentiated spindle and giant cell carcinoma , and lymphovascular and perineural invasion present. The tumor did not invade to adjacent organs and 25 lymph nodes were negative for malignancy. There was no gallbladder involvement. The patient improved and was discharged to home after extended recovery.





Discussion

The outcome of patients with this type of carcinoma of the extrahepatic bile duct is still uncertain because of the limited number of reported cases. In a case series of 10 patients with undifferentiated carcinoma of the bile duct, 4 patients were recurrence-free for 7–60 months following surgical treatment (1).

While reported one-year survival rate for patients with undifferentiated carcinoma of the gallbladder was 18 percent, patients with undifferentiated carcinoma of the extrahepatic bile duct have a relatively better prognosis due to early jaundice (2). As a result, such cases may be diagnosed at a resectable stage before tumor progression.

Our patient was placed on adjuvant chemotherapy due to lymphatic and perineural invasion. Data that includes longer follow up for recurrence free survival would help prognosticate these patients.

References

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2. Guo K, Yamaguchi K, Enjoji M: Undifferentiated carcinoma of the gallbladder. A clinicopathologic, histochemical, and immunohistochemical study of 21 patients with a poor prognosis. Cancer 1988;61:1872–1879