Atypical Presentation of an Advanced Obstructive Biliary Cancer without Jaundice: A Case Report

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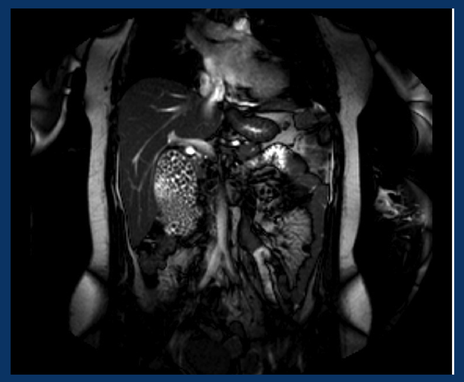
INTRODUCTION

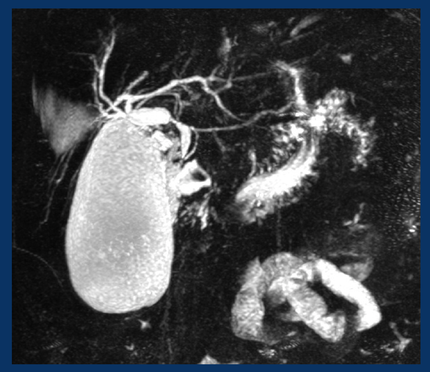
Biliary tract cancer is the second most common primary hepatobiliary cancer, after hepatocellular cancer. Clinical manifestation due to biliary obstruction tend to occur early if the tumor is located in the common hepatic duct, common bile duct or ampulla of Vater.[1] We present a case of an elderly female diagnosed with advanced cholangiocarcinoma presenting with Courvoisier's sign in the absence of cholestatic jaundice.

CASE PRESENTATION

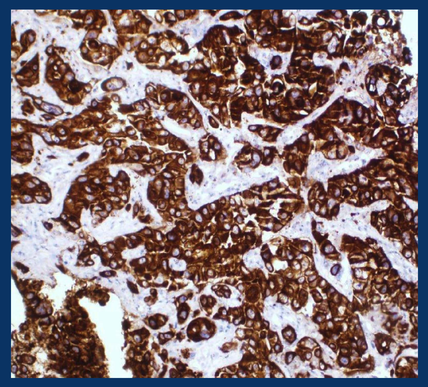
A 60 year old female with hypertension, hypercholesterolemia, and osteoporosis comes in with a 3-day duration of worsening RUQ and lower abdominal pain described as sharp, constant, non-radiating without associated fever, chills, jaundice, weight loss, nausea, vomiting or changes in bowel movement. Physical exam revealed anicteric sclerae, soft abdomen with normal bowel sounds, palpable gallbladder and left lobe of the liver, no abdominal tenderness or guarding, no splenomegaly appreciated. Pelvic exam was remarkable for palpable right-sided pelvic mass. EGD was normal. Colonoscopy showed non-bleeding diverticulosis. MRI with contrast revealed a hypointense left hepatic lobe mass, an enlarged gallbladder with cholelithiasis without pericholecystic fluid. MRCP revealed intrahepatic biliary ductal dilation measuring approximately 1 cm with abrupt short segment narrowing in the extrahepatic common bile duct measuring 0.2 cm in diameter. The distal common bile duct measures normal in diameter. There was no obvious filling defect to suggest choledocholithiasis. Liver function panel was deranged showing mild cholestatic pattern without bilirubinemia. Tumors markers were significant for markedly elevated CA19-9 (12394 U/mL) with moderate elevation in CEA (394 ng/mL) and CA-125 (85 U/mL) while serum AFP was within normal limits. CT-guided liver biopsy was obtained and histopathological exam revealed high-grade carcinoma. Epithelial nature was confirmed by positive stains for cytokeratins. Patient subsequently underwent surgical resection of the liver mass. Intraoperative findings included a large gallbladder and a palpable mass within the mid-common bile duct acting as the source of obstruction for the gallbladder. Histopathological examination revealed invasive poorly differentiated adenocarcinoma involving common bile duct, cystic duct and gallbladder neck. It invaded also into the adjacent left liver lobe. Metastases were identified in the portal lymph nodes.



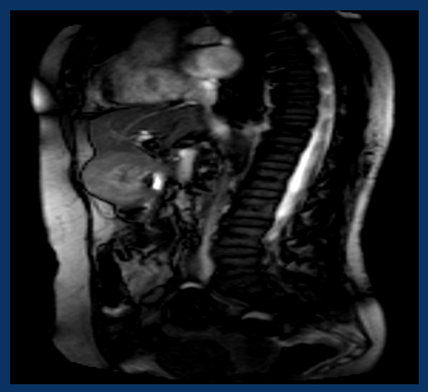
MRI Abdomen: The distended gallbladder with multiple stones



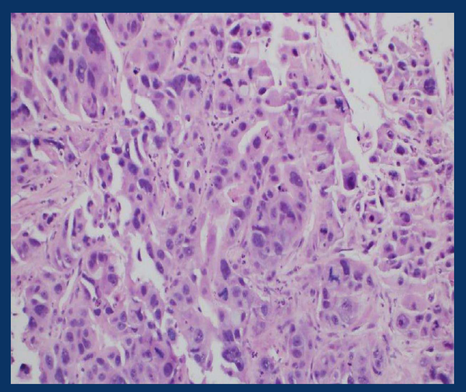
MRCP: The narrowing on the mid common bile duct



Needle biopsy stained with Cytokeratin 7



MRI Abdomen: The left hepatic lobe mass



Needle biopsy: High grade carcinoma (x200)

DISCUSSION

It has been reported that the major symptoms of cholangiocarcinoma were abdominal pain, weight loss, pruritus and jaundice but about one quarter of the patients were not clinically icteric. A palpable gallbladder (Courvoisier's sign) occurs rarely with cholangiocarcinoma, unless it arises from common bile duct distal to cystic duct.[2] Chung reported that chronically increased ductal pressure is the probable cause of dilated gallbladders seen in malignant obstruction of the common duct. Patients with Courvoisier gallbladder usually have longer history of and deeper jaundice in presentation.[3] Our case was atypical in presentation since patient already had distended gallbladder (Courvoisier' sign) suggestive of severe obstruction secondary to advanced biliary cancer, despite the absence of clinical evidence of jaundice and laboratory evidence of bilirubinemia. La Greca et al. reported a similar case involving squamous cell carcinoma of the common bile duct presenting atypically without jaundice despite the proximal bile duct dilatation.[4] It has been shown in retrospective epidemiologic review that adenocarcinoma was the most common histologic type of biliary cancer which has an overall 5-year survival rate of 12.7%.[5]

CONCLUSION

We presented a case of an elderly female with advanced biliary carcinoma with severe obstruction in the biliary tree causing Courvoisier's phenomenon but had remained anicteric and without bilirubinemia on initial presentation. This scenario may suggest that laboratory evidence of cholestasis might lag behind the severity of the biliary obstruction in cholangiocarcinoma.

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[3] Chung R. S., Pathogenesis of the Courvoisier Gallbladder. Dig Dis Sci. 1983 Jan; 28(1):33-8.
[4] La Greca G, Conti P, Urrico GS, Catanuto G, Di Carlo I, Russello D, Latteri F. Biliary Squamous Cell Carcinoma. Chir Ital. 2004 Mar-Apr;56(2):289-95.
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