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Markedly Elevated CA 19-9 in a Case of Mirizzi's Syndrome: An Atypical Presentation

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Introduction

It has been well described that carbohydrate antigen (CA) 19.9 is synthesized by epithelial cells of the gastrointestinal tract, pancreatic duct and biliary tree. The CA 19.9 antigen is commonly used as a tumor marker for malignancies of the pancreas and biliary tract. It has been suggested that high levels of CA 19-9 (> 300 U/ml) antigen have a strong correlation with malignant biliary pathology. In this paper, we present a patient with extremely elevated levels of tumor marker (CA 19-9 >4000) resulting from Mirizzi's Syndrome.

Case Presentation

Our patient was a 51 year old male with a several week history of epigastric pain who presented with new onset jaundice, tea-colored urine and light stools of one week duration. Total bilirubin was elevated at 19.6. Initial abdominal CT scan revealed markedly dilated intrahepatic and common bile ducts, the latter measuring 1.5 cm. No stones or masses were visualized. CA 19-9 levels were elevated at 4258. Given his presentation, clinical suspicion was high for a biliary tract malignancy. Specifically, there was concern for early pancreatic cancer or a lesion at the Ampulla of Vater. The patient underwent Endoscopic Ultrasound Examination which demonstrated a large stone at the junction of the cystic duct and common bile duct. The bile duct was swept without evidence of stones. A wire was then advanced into the cystic duct, however the stone could not be successfully retrieved. A stent was placed in the cystic duct but the magnitude of the stone was such that subsequent cholangiogram did not demonstrate cystic duct filling beyond the level of obstruction. The patient underwent cholecystostomy tube placement and was continued on antibiotics. He improved clinically with gradual decrease of his hyperbilirubinemia and underwent a laparoscopic cholecystectomy four weeks later.



Fig. 1 Marked dilation of the intrahepatic ducts.



Fig. 2 Distended gallbladder.



Fig. 3 Stone visualized in the bile vs cystic duct.



Fig. 4 Dilated common bile duct.

Discussion

- Elevations in CA 19-9 levels have traditionally been associated with malignancies of the biliary tract and pancreas.
- There is growing data, however, demonstrating elevated levels of CA 19-9 in benign biliary pathology, obstructive and infectious.
- Typically, CA 19-9 levels greater than 300 U/ml are used as the upper limits of normal for benign processes.
- To our knowledge, there are only three other reports of CA 19-9 elevations of this magnitude secondary to Mirizzi's Syndrome.
- Helling et. al described a similar case of Mirizzi's Syndrome with CA 19-9 levels above 6000 U/ml.¹ In 2006 and 2007, 2 additional cases were reported with CA 19-9 levels of >5000 U/ml and >16,000 U/ml.
- ERCP was helpful when the common bile duct could be cannulated, but still did not provide definitive diagnoses.
- EUS was not mentioned in any of these case reports and the culprit gallstones were discovered only intraoperatively during cholecystectomy or, in one case, laparotomy for open liver biopsy.
- EUS can be a useful tool to evaluate for gallstones impinging on the common bile duct to further distinguish between malignant etiology and Mirizzi syndrome which can be difficult to visualize on transabdominal ultrasound.

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