Jaundice with markedly elevated carbohydrate antigen 19-9 (CA 19-9): Biliary stones or malignancy?

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A 59-year-old male with past medical history of untreated type 2 diabetes mellitus, hypertension, and hyperlipidemia presents to the UMC family medicine clinic for a follow up for chronic conditions after being lost to follow up for about 2.5 years. On initial observation, the patient appeared jaundiced. The jaundice is not associated with nausea, vomiting, diarrhea, or constipation. The patient does have generalized pruritus. He denied having pain in spite of questioning during the first two family medicine visits and the first visit with gastroenterology. However, on the 3rd Family Medicine visit, the patient admitted to the medical student that he had epigastric and RUQ pain with episodes over the past 6 months. He noted to the team that he did not admit to his pain because he did not want to take off work due to financial concerns. The patient noted the jaundice worsened when the pain episodes occurred and faded between episodes.

On physical exam, scleral icterus was prominent and palmar icterus was also present. The patient had a dark complexion making it difficult to appreciate any icterus elsewhere on the skin. The patient's liver span was about 10 cm, nontender with negative Murphy's sign. The liver edge was smooth. No telangiectasias or distended surface collateral veins noted. The patient had normal bowel sounds and no tenderness to palpation of the abdomen. Laboratory values included elevated ALT (90), AST (58), and alkaline phosphatase (460) in addition to a markedly elevated bilirubin of 20.5. Low hemoglobin (10.9), hematocrit (33.2), and MCV (73.7) were also found. Hemoglobin A1C was 7.1%. Lipase was within normal limits and hepatitis panel was negative. Carbohydrate antigen 19-9 (CA 19-9) was elevated to 16,947 U/ml. Abdominal ultrasound revealed cholelithiasis and dilated common bile duct. MRCP revealed cholelithiasis and choledocholithiasis and two foci of increased signal near the pancreatic duct. Endoscopic ultrasound revealed a 10 x 9 mm mass in the pancreatic neck. Initial pathology report from a fine needle aspiration of the pancreatic mass suggested no evidence of dysplasia or malignancy; however, the pathology report was addended following immunohistochemical staining and found synaptophysin and neuron-specific enclase (NSE) positive cells in suspended blood raising suspicion for a neuroendocrine tumor; however, more material is required for a diagnosis. Following stent placement in the common bile duct, the patient's CA 19-9 went down to 2,629. Weeks later, the patient had a cholecystectomy revealing chronic and mild acute cholecystitis with cholelithiasis and a cholangiogram. A large stone was unable to be cleared from the common bile duct; thus, he is scheduled for a follow up ERCP and lithotripsy.

This patient presented with jaundice, initially thought to be painless, and episodic epigastric pain that could be explained by the stones in his cholelithiasis and choledocholithiasis. The patient fear of missing work discouraged him from being upfront about his pain. The initial presentation of painless jaundice caused malignancy to be much higher on the differential; therefore, a CA 19-9 lab was ordered and found to be remarkably elevated. This patient's elevated CA 19-9 and findings of a mass at the pancreatic neck suggest he may have an undiagnosed malignancy. Studies suggest limited sensitivity (72%) and specificity (86%) of CA 19-9 as a tumor marker; however, the cutoff used is typically 37 U/ml. One study found that the median CA 19-9 in benign pancreatic disease was 15 (interquartile range 6-39). However, a case report found that a patient with cholelithiasis and choledocholithiasis had markedly high CA 19-9 (9586 U/ml) levels that rapidly declined following treatment. The patient presented here did have a decline in CA 19-9 following initial treatment. Further investigation is still warranted on this patient due to the suspicious findings in his pancreas.