

Hyperbilirubinemia due to Klatskin Tumor

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Abstract

- Klatskin tumor is a rare form of cholangiocarcinoma that generally carries poor prognosis despite surgical resectability.
- We present a case of an elderly female presenting with painless jaundice and unintentional weight loss, with laboratories pertinent for hyperbilirubinemia and subsequent MRCP findings of a 1.5 cm Klatskin tumor.

Case

- 74 y/o female** with PMH of NIDDM2, HTN, CHF, RA, and h/o breast cancer in remission s/p chemotherapy and right breast mastectomy, presented with **3 week onset of painless jaundice**
- ROS: **12 lb unintentional weight loss, early satiety, decreased appetite, generalized pruritis, and occasional chills.** Patient denies nausea/vomiting, abdominal/rectal pain hematochezia/hematemesis, or additional complaints.
- Patient denied similar presenting symptoms in the past, sick contacts, travel history, or any exacerbating/alleviating factors
- Vitals: T: 36.9°C; HR: 92bpm; RR: 16/min; O2 sat: 97% RA
- PE: **cachexia, AAOx4, bilateral scleral icterus, dry mucous membranes, diffuse jaundiced skin.** Abdomen was soft, NT, ND w/out organomegaly. No additional pertinent PE findings

Decision-Making

Labs:

AST	ALT	ALP	T. Bili	D. Bili	I. Bili	Serum Protein	Serum Albumin	Lipase
258	262	677	19.4	14.0	5.6	8.2	4.1	76

- Na **124**, Glucose **229**. UA with **elevated bilirubin** and normal urobilinogen.
- Ultrasound and CT abdomen/pelvis w/ IV contrast performed on day 1 of hospital admission (Figures 1 & 2 respectively) showed findings for **intrahepatic biliary ductal dilation** without extrahepatic biliary ductal dilation, gallstones, hepatomegaly. No gallbladder, common bile duct, or pancreatic abnormalities were found. **Suspicion for obstructive pathological cause of hyperbilirubinemia.**
- ANA titer revealed **1:1280 with speckled pattern** while AMA level was **48.9**, suggestive of autoimmune hepatitis/primary biliary cirrhosis
- MRCP (Figure 3), revealed **diffuse intrahepatic biliary ductal dilation** without extrahepatic biliary dilation and a **1.5cm irregular hypointense signal at the liver hilum** was noted, with suspicion for a Klatskin tumor.
- Tumor markers (CA19-9, AFP, CEA) revealed **elevated CA-19-9**
- GI was consulted. On day 5, patient underwent ERCP with sphincterotomy, brushing, and common bile duct (CBD) stent placement to decompress intrahepatic biliary obstruction while evaluating and conducting biopsy of suspected Klatskin tumor

Relevant Imaging Findings

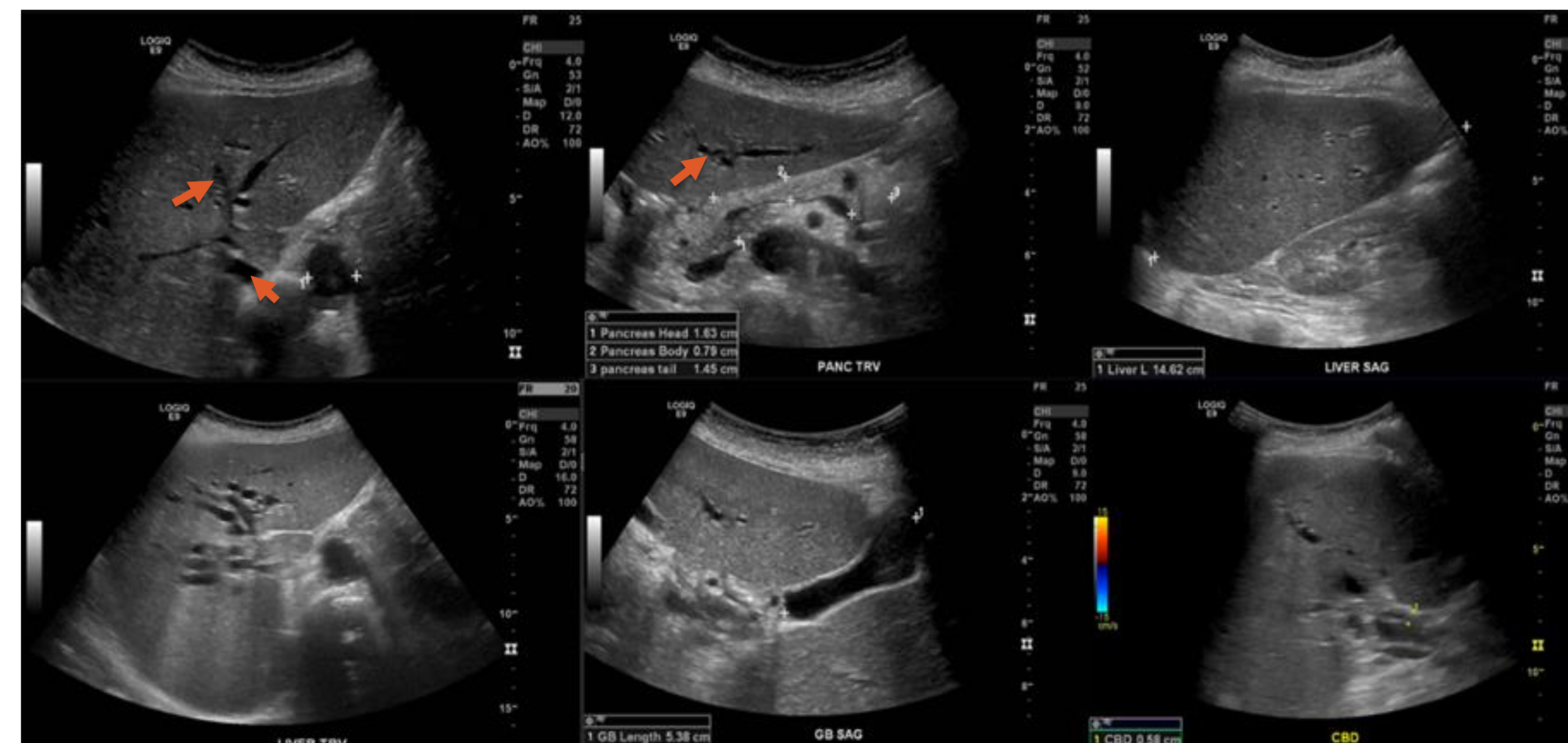


Figure 1: Abdominal ultrasound with findings of intrahepatic biliary ductal dilation (some indicated with arrows) without gallstones, hepatomegaly, or abnormalities in gallbladder, common bile duct, or pancreas.



Figure 2: CT Abdomen/pelvis with IV contrast with findings of intrahepatic biliary ductal dilation (some indicated with arrows) without extrahepatic biliary ductal dilation or distended gallbladder.

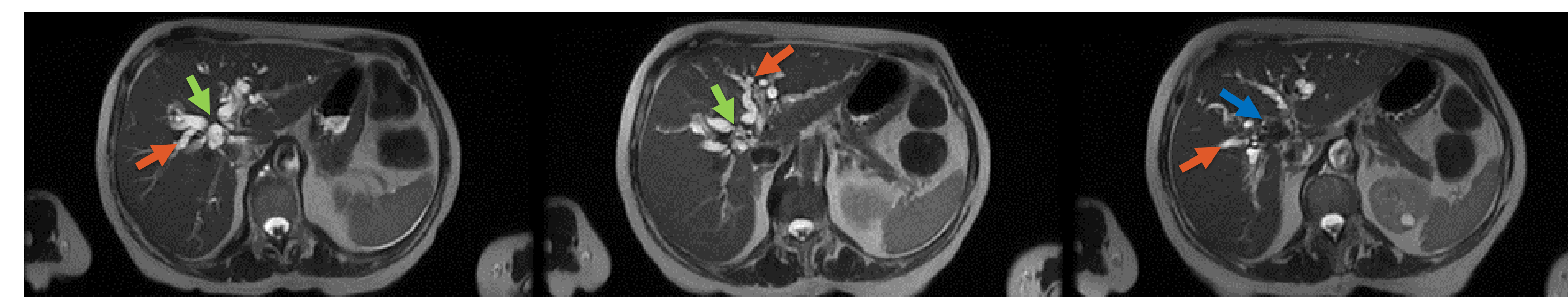


Figure 3: MRCP with diffuse intrahepatic biliary ductal dilation (some indicated with orange arrows) without extrahepatic biliary ductal dilation, with an obvious hypodense obstruction on intrahepatic biliary duct indicated with green arrows as well as findings of 1.5cm hypointense signal (blue arrows) at liver hilum for concerns for Klatskin Tumor.



Figure 4: CT Abdomen/pelvis with PO contrast s/p ERCP and CBD stent placement (arrows) with findings of decreased intrahepatic biliary ductal dilation.

Decision-Making (contd.)

- CT abdomen/pelvis w/ PO contrast (Figure 4) on day 7 revealed **decreased intrahepatic biliary ductal dilation s/p ERCP and appropriate CBD stent placement.**
- Day 5-13: **Significantly reduced T. bili, D. bili, and LFTs.** Pathology report from brushing revealed no malignant cell findings. Patient was transferred to another hospital for further advanced evaluation and management.

Discussion

- Klatskin tumors arise at the hepatic duct bifurcation and account for the most common types of cholangiocarcinoma. Patients with intrahepatic involvement, like in Klatskin tumor, present typically with right upper quadrant abdominal pain, elevated ALP and/or weight loss^{1,2,3,4,5}
- Diagnostic testing: LFTs, bilirubin levels and tumor markers including CA19-9, CEA, and AFP. Tumor markers play a role in monitoring and evaluating for recurrence⁶
- Imaging includes MRCP/contrast enhanced MRI. Tissue biopsies (FNA or CT/MRI guided biopsy) are difficult due to perihilar location and considered if imaging is indeterminate, patient/physician reluctance on surgery or non-operative treatment is considered⁷
- Cholangiocarcinoma spreads locally initially, nodal involvement and then distant spread thereafter. Klatskin tumor specifically spreads via perineural and periductal lymphatic channels after local spread. It is considered distant metastasis when hepatic duodenal ligament is involved⁸
- CT abdomen/pelvis is recommended, followed by PET scan to further look for metastasis. ERCP or percutaneous transhepatic cholangiography can also be obtained for staging distal lesions.
- For patients with symptomatic obstructive jaundice, percutaneous drainage is recommended. Treatment for intrahepatic cholangiocarcinoma often includes hepatic resection to achieve negative resection margins, however this only has a 30% cure rate and requires adjuvant fluorouracil-based chemotherapy.^{9,10}

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