

INTRODUCTION

Intraductal papillary mucinous neoplasm (IPMN) is a mucin-producing papillary epithelial neoplasm arising from the pancreaticobiliary system. IPMN of the pancreas (P-IPMN) are common and widely recognized. Biliary tract IPMNs (BT-IPMN) are much rarer and have higher malignant transformation potential. We present a case of BT-IPMN with biliary-gastric and biliary-duodenal fistulas and malignant degeneration.

CASE PRESENTATION

A 56-year-old Asian female with history of cholecystectomy presented with abdominal pain and fever to an outside hospital, where she was diagnosed with cholangitis, and a large hepatic cystic lesion. A percutaneous biliary drain was placed and drained 500 cc of bilious fluid.

- **Labs:** Total Bilirubin 7.4, Alkaline phosphatase 1195
- **CT:** Intrahepatic ductal dilatation, a large fistula from the common bile duct to the duodenum, and a large (15.3 x 4.1 x 5 cm) intrahepatic cystic lesion with wide mouthed fistula tracts between the left hepatic biliary system and the stomach.
- **ERCP/ENDOSCOPY:** copious viscous mucin in the stomach originating from the large hepato-gastric fistula. A second fistula extending from the duodenal bulb to the mid-proximal bile duct with thick mucin secretion. Stenting of the biliary tree was not performed due to the drainage of copious amount of highly viscous mucin. A regular upper endoscope was introduced through the duodenal and gastric fistulas to the extrahepatic bile duct and the left intrahepatic biliary ducts respectively. The biliary epithelium was abnormal with villous projections. Biopsies taken.
- **EUS** showed heterogenous hypoechoic appearance and FNA was done.
- **Pathology:** papillary projections lined by intestinal-type epithelium with low grade dysplasia, compatible with an intraductal papillary neoplasm and adenocarcinoma of pancreaticobiliary origin.
- **Diagnosis:** BT-IPMN with malignant degeneration into cholangiocarcinoma.
- Not a surgical candidate and chemotherapy was started.

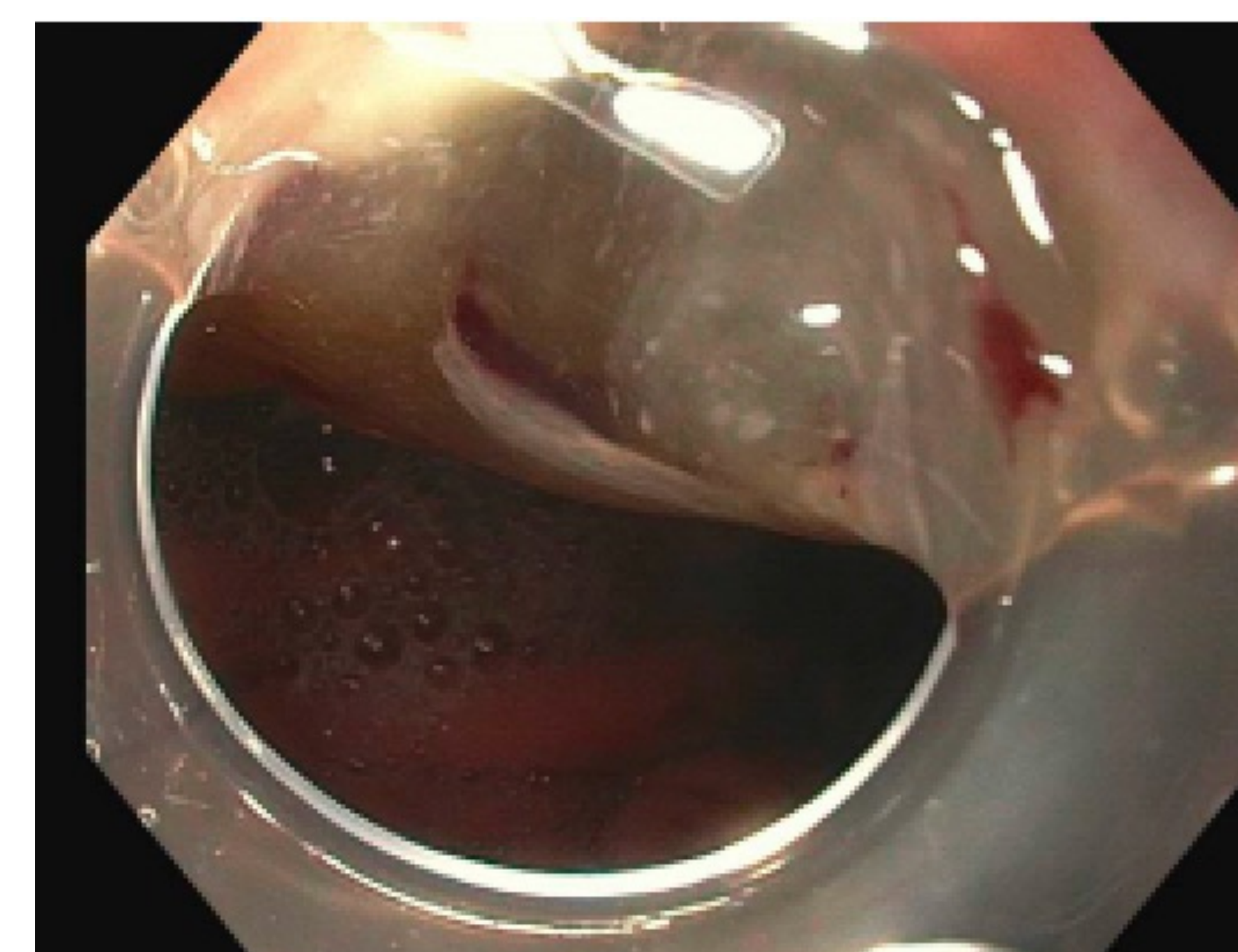


CT showed hepato-gastric fistula and biloma

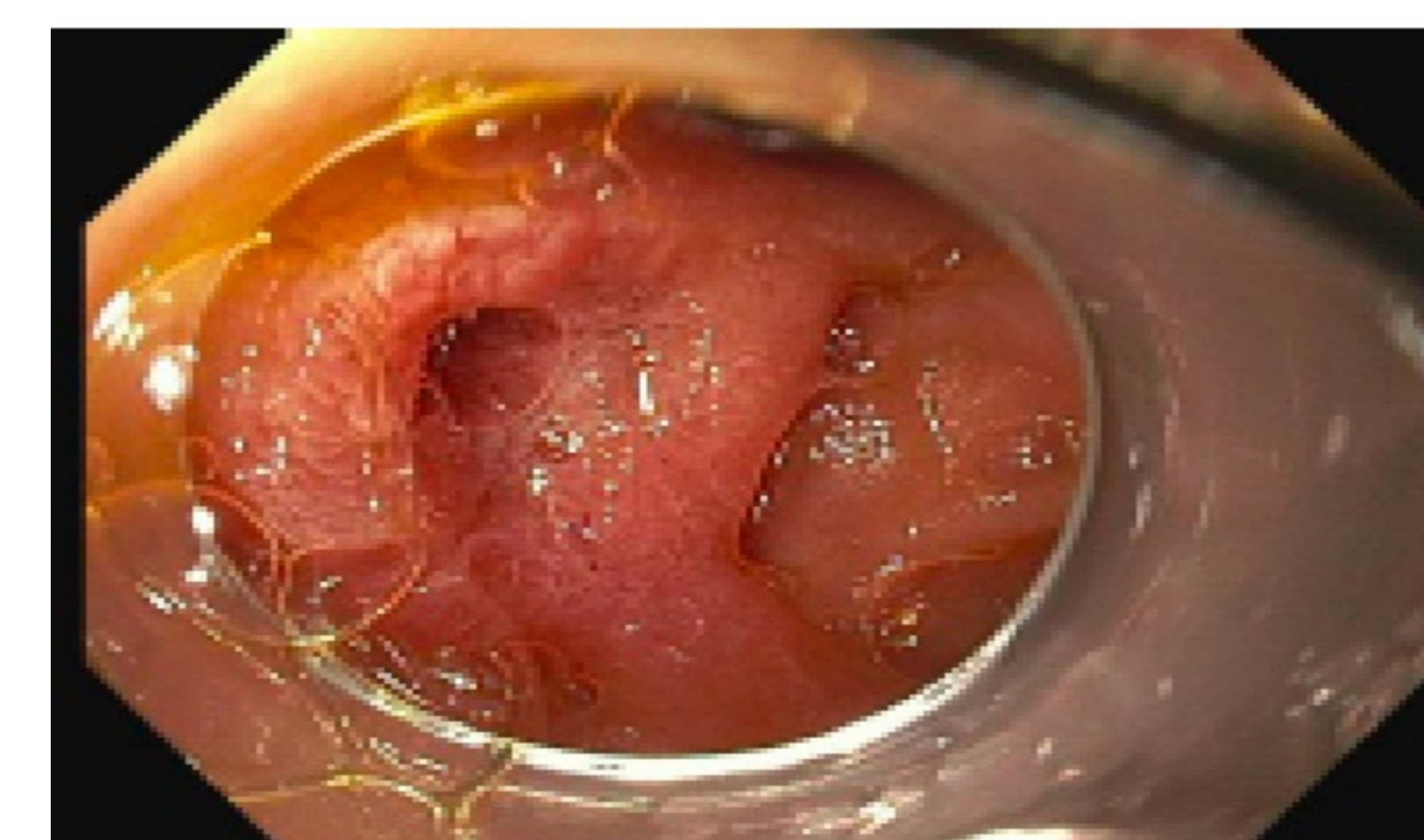
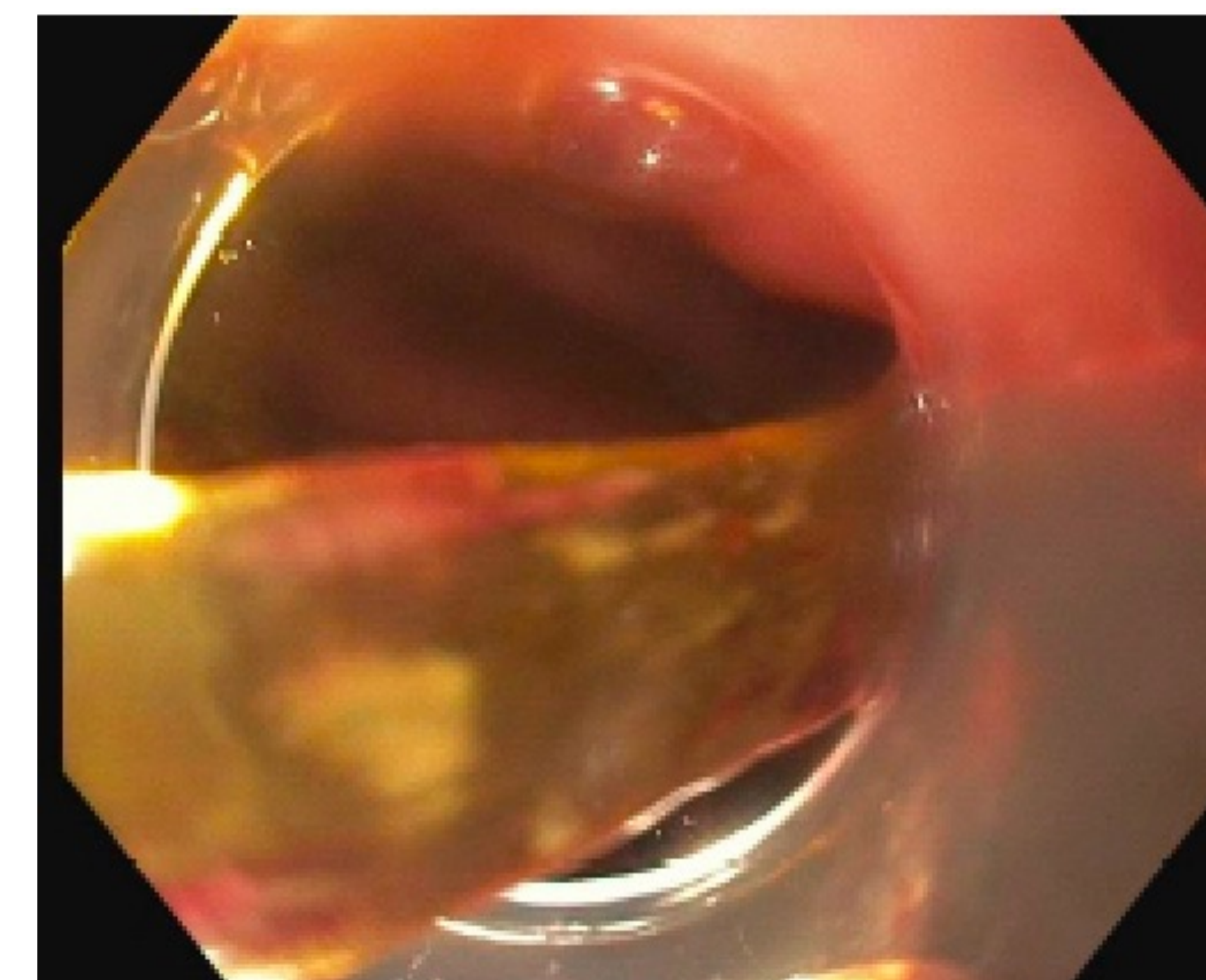


EUS: diffuse abnormal echotexture in left lobe of liver with heterogenous hypoechoic appearance. FNA performed.

ENDOSCOPIC IMAGES:



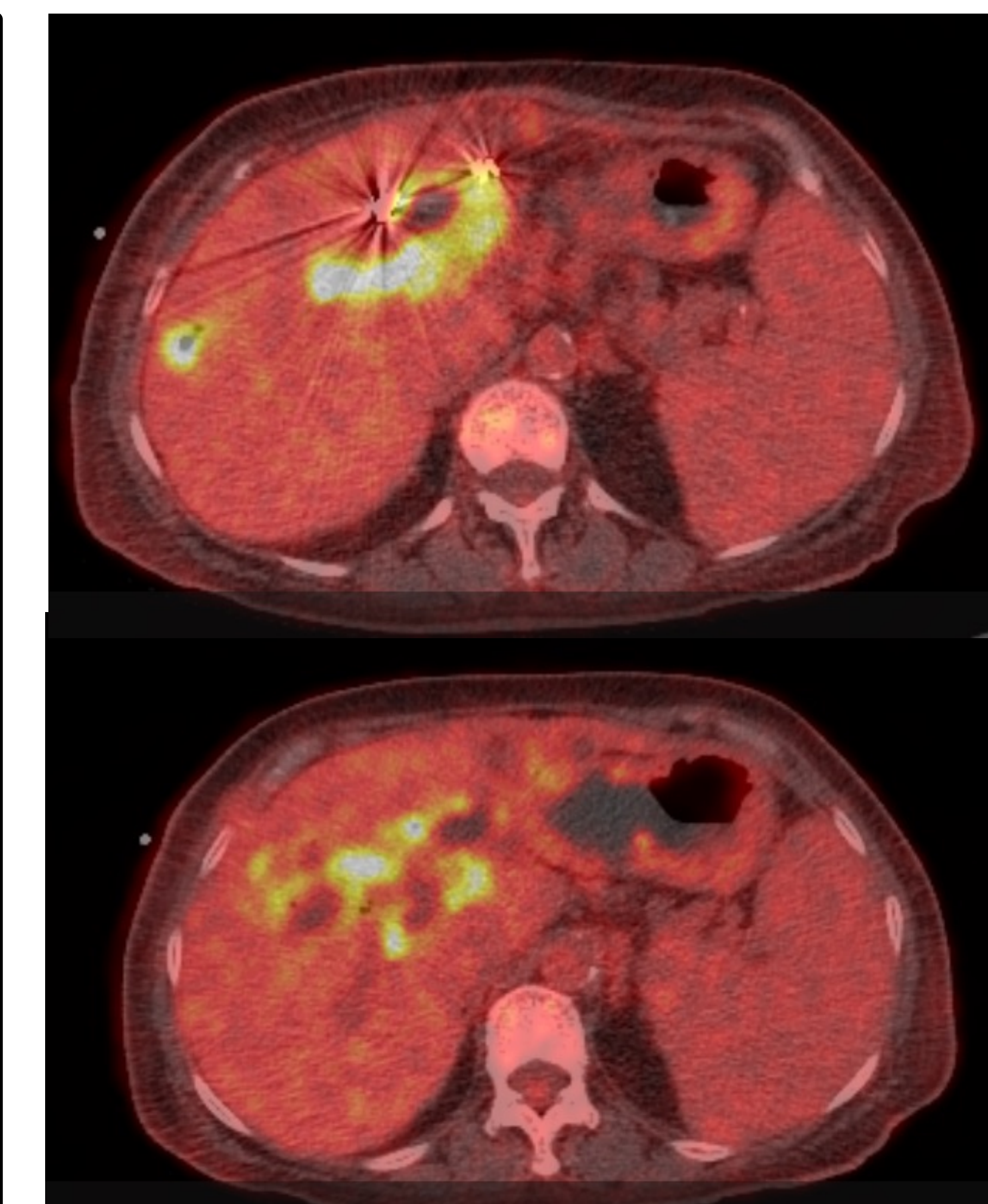
Thick mucinous secretion originating from hepato-gastric fistula



A large duodenal-biliary fistula was noted and traversed. A cavitory lesion within the liver with thick mucinous secretion was observed.



The distal biliary epithelium appeared abnormal with long villous projections



PET/CT: Intense FDG uptake within the biloma suspicious for malignancy vs infection

DISCUSSION

- IPMN-B is rare and not well understood
- More research and understanding of IPMN-B is needed
- IPMN-B has a higher malignant transformation potential than its pancreas counterpart
- IPMN-B is a known precursor for cholangiocarcinoma
- Imaging typically shows upstream biliary dilation with or without visible biliary mass and variable downstream biliary dilation/filling defects based on amount of mucin production
- Early detection/diagnosis can prevent complications such as fistula formation and malignant transformation
- Early surgical resection is curative