Diagnosis and Management of Intraductal Papillary Neoplasm of the Bile Duct via Cholangioscopic Evaluation

Mili Parikh, MD[1], Sooraj Tejaswi, MD[1]
1. University of California, Davis Medical Center

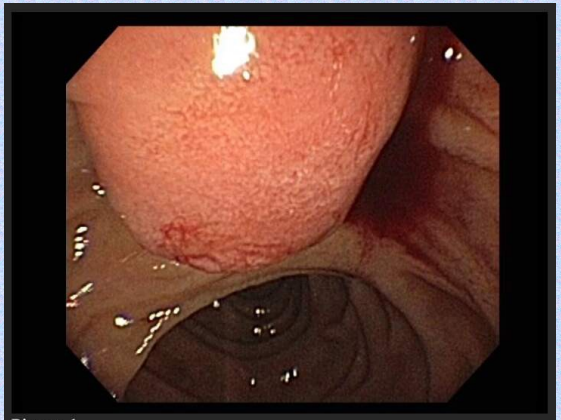
Introduction

Intrampullary papillary neoplasm is a rare tumor, usually seen in females in their 60s, with an incidence of 0.025 per 100,000. It can progress to adenocarcinoma, and the 5-year survival rate is 33%. It is distinct from an ampullary adenoma, which arises from the duodenal mucosa overlying the ampulla. As such, it is unlikely to be cured by ampullectomy as is the case of ampullary adenoma.

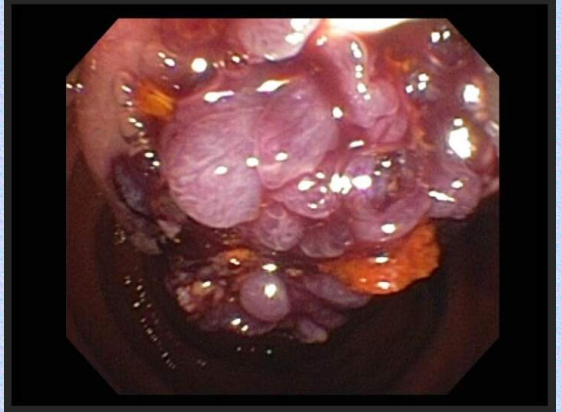
Case Description

63F with prior cholecystectomy was found to have abnormal liver tests (AST 104, AST 233, alkaline phosphatase 290) and mild hyperbilirubinemia (total bilirubin 1.7). On review of symptoms, she described an isolated incident of nausea, vomiting and fever which self-resolved. Patient was otherwise asymptomatic and vital signs were within normal limits. Abdominal ultrasound showed diffuse biliary dilatation without obvious stones or stricture. MRCP showed multiple filling defects in the distal common bile duct (CBD). ERCP revealed a prominent ampulla. Digital cholangioscopy done for further investigation showed an exophytic papillary tumor in the distal CBD (photo 1) along with excess luminal mucous (photo 2). The endoscopic and cholangioscopic findings of a papillary intraluminal mass and mucobilia was suggestive of intraductal papillary neoplasm of the bile duct (IPNB).

69F with a complicated biliary history was seen by her PCP for jaundice and R shoulder pain and was found to have cholelithiasis and choledocholithiasis. She underwent two ERCPs with stone extraction and ultimately laparoscopic cholecystectomy after MRI demonstrated CBD stone. An ampullary prominence of 1 cm was concerning for an invasive carcinoma. ERCP showed multifocal frond like papillary projections at the left hilum and distal CBD and copious thick mucin through the examined extrahepatic duct and hilum; concerning for IPNB with progression to cholangiocarcinoma. Both patients underwent surgical evaluation.



Picture 1



Picture 2

Discussion

Intrampullary papillary neoplasm is a rare tumor with poor prognosis. It is unclear if it is a variant of intraductal papillary neoplasm of the bile duct (IPNB) that also includes the ampulla or if it primarily arises within the ampullary channel and spreads proximally into the bile duct and the pancreatic duct. One of our patients was noted to have involvement of the gall bladder. Diagnosis is established by cholangioscopy with biopsies. Cytology brushing increases the likelihood of definitive diagnosis. Cross-sectional imaging did not reveal involvement of the main pancreatic duct, but this noted intraoperatively in one of our patients. As such, endoscopic ultrasound of the ampulla and pancreas is essential. Cholangioscopy should be undertaken for tumor mapping, and if possible pancreatoscopy can also be undertaken. Pancreaticoduodenectomy should be treatment of choice for IAPN or IAPN with adenocarcinoma without distant metastases in order to achieve tumor free margins, as seen via pathology tissue. A same session cholecystectomy might be prudent, especially if there is cholangioscopic evidence of biliary ductal involvement.

In conclusion, IAPN should be suspected based on endoscopic ampullary features such as a prominent ampulla, prolapse of papillary fronds on sphincterotomy, lack of resolution of cholestasis despite duct clearance. Cholangioscopy should be undertaken to establish an early diagnosis.

References

1. Kawaguchi Y, Kawashima Y, Maruno A, Ito H, Ogawa M, Izumi H, Furukawa D, Yazawa N, Nakagori T, Hirabayashi K, Mine T. An intraductal papillary neoplasm of the bile duct at the duodenal papilla. Case Rep Oncol. 2014 Jul 2;7(2):417-21. doi: 10.1159/000364999. PMID: 25126070; PMCID: PMC4130820.
2. Firkins SA, Chen W, Krishna SG. More Than Skin Deep: Beneath the Surface of a Routine Ampullary Adenoma. Gastroenterology. 2021 Jun;160(7):2255-2257. doi: 10.1053/j.gastro.2021.02.067. Epub 2021 Mar 4. PMID: 33676973.
3. Scomparin RC, Bento LH, Batista CP, de Lima MS, de Paulo GA, Martins BC, Maluf-Filho F. Long-term result of endoscopic treatment of an ampullary adenoma with extension into the common bile duct. Endoscopy. 2018 May;50(5):E109-E110. doi: 10.1055/s-0044-100917. Epub 2018 Feb 16. PMID: 29452434.
4. Small AJ, Baron TH. Successful endoscopic resection of ampullary adenoma with intraductal extension and invasive carcinoma (with video). Gastrointest Endosc. 2006 Jul;64(1):148-51. doi: 10.1016/j.gie.2006.02.034. PMID: 16813830.