

Case Report

Papillary neoplasms of biliary tract-a histopathological surprise

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ABSTRACT

Papillary neoplasms of the biliary tract is a relatively new entity which most often comes as a histological surprise. They are often diagnosed as malignancies and are treated like that. But the prognosis is better when compared to malignancy itself. Here we reported two cases of papillary neoplasms of the biliary tract. First case was a 50 year old male who was evaluated for obstructive jaundice and on evaluation found to be type 3b hilar cholangiocarcinoma and underwent left hepatectomy with extra hepatic bile duct resection and portal lymphadenectomy. Histo-pathological report was intra-ductal papillary neoplasm - biliary type (IPN B). Second case was an incidental finding of arterial enhancing lesion in the gallbladder wall on CT scan, which was done for the evaluation of bicytopenia. This also was reported as malignancy in pre-operative imaging and hence underwent anticipated extended cholecystectomy with wedge resection of 2 cm adjacent hepatic parenchyma. Again histopathology revealed it as intra-cholecystic papillary neoplasm with focal dysplasia. Both cases were followed up for more than one and half years and showed no evidence of recurrence, hence pointing towards better prognosis. Papillary neoplasms are difficult to diagnose preoperatively and are often treated with oncological resections but they carry a better prognosis when compared with their malignancy counterparts.

Keywords: Intraductal papillary neoplasm, Biliary neoplasm, IPN B, Intracholecystic papillary neoplasm

INTRODUCTION

Papillary neoplasms of biliary tract and gallbladder are rare forms of neoplasms with varying spectrum of clinical and histopathological characteristics. They are considered as the histological counterpart of Intraductal papillary mucinous neoplasm of pancreas and are considered as forerunner of invasive carcinoma.¹ We report 2 cases of papillary neoplasms with a varied spectrum of clinical presentations and the challenges faced during the diagnosis and treatment.

CASE REPORT

A 50-year-old male, presented with painless jaundice, acholic stools and pruritis of 2 months duration. There was associated anorexia but no weight loss. On evaluation, he had an obstructive jaundice pattern in LFT with a total bilirubin of 4.8 and direct bilirubin of 3.1. All other routine blood investigations were within normal. He

had a waxing and waning pattern of jaundice after admission. His CA 19-9 was 16.8 IU/ml, well within the normal limits. His triple-phase contrast CT scan evaluation showed an ill-defined soft tissue lesion filling the biliary confluence (Figure 1). The lesion extends into the left system, causing distension, with upstream dilatation of the intrahepatic biliary radicals without any vascular involvement. His MRCP also showed an ill-defined T2 hypo intense soft tissue lesion with diffusion restriction in the CHD extending into the left system (Figure 2). With the above investigations, he was diagnosed with TYPE IIIB Hilar cholangiocarcinoma. His viral screen was negative. He underwent intra-op frozen biopsy assisted Left hepatectomy + caudate lobe resection and CBD resection with biliary reconstruction. During surgery, because of the difficulty in assessing the extension of the polypoidal growth into the right hepatic duct, the bile duct was opened and the right hepatic duct was divided under vision (Figure 3, Figure 4). A frozen section from the right hepatic duct margin and growth

was negative. He had an uneventful postoperative course and recuperated well. His post-op histopathological analysis showed IPN B type neoplasm (Figure 5). Currently, he is on follow-up with no recurrence after a follow up of 2 years.



Figure 1: CT scan of lesion filling bile duct confluence with extension to left hepatic duct causing upstream biliary dilatation.

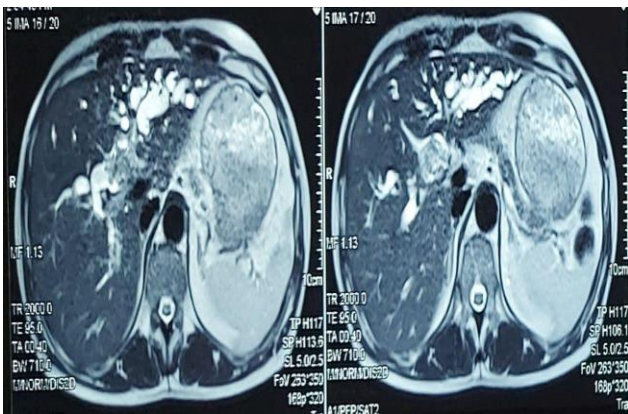


Figure 2: MRI scan showing a lesion filling the bile duct confluence with extension to left hepatic duct causing upstream biliary dilatation.

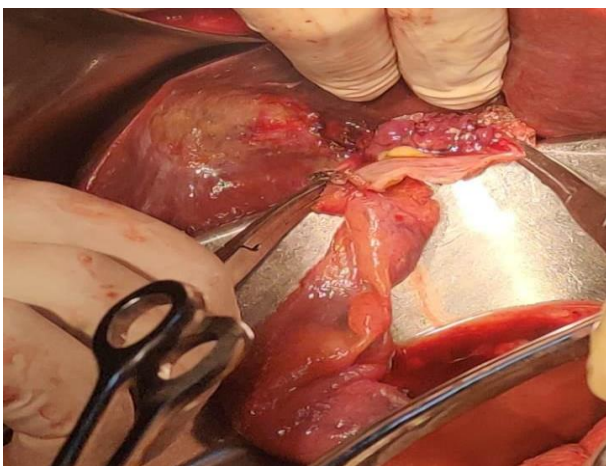


Figure 3: Intra -op photo showing opened bile duct and papillary neoplasm.



Figure 4: Post-operative specimen showing left hepatectomy with caudate resection and extrahepatic bile duct.

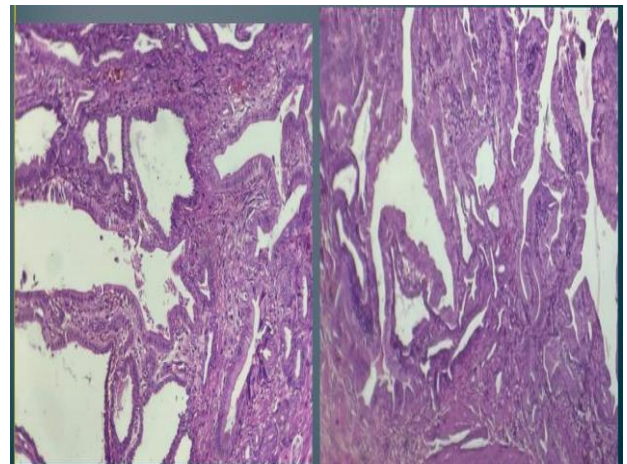


Figure 5: Histopathology showing papillary growth pattern.

A 45-year-old female, during evaluation for thrombocytopenia, showed a soft tissue lesion in the fundus of the gallbladder on USG of the abdomen. She was asymptomatic and her routine blood tests all were within normal limits, except for bicytopenia with TLC of 3400 and platelet count of 50000. Her viral screen was negative and there was no evidence of chronic liver disease or portal hypertension. Her OGD scopy and fibroscan and marrow studies were normal. Her CEA was 1.2 and CA 19-9 was 22 IU/ml. Her triple-phase contrast CT scan showed heterogeneously enhancing mass in the fundal region measuring 13×13×9 mm, with few enlarged peri choledochal nodes (Figure 6). Her MRI scan revealed a focal endophytic lesion seen to arise from the fundus and posterior wall of GB with diffusion restriction, consistent with malignancy (Figure 7). She underwent intra op frozen biopsy guided anticipatory

extended cholecystectomy with lymphadenectomy and splenectomy (Figure 8). She recovered well. Post-op histopathological examination showed intra cholecystic papillary neoplasm with focal high-grade dysplasia (Figure 9). She is currently on follow-up, after 20 months and is asymptomatic.

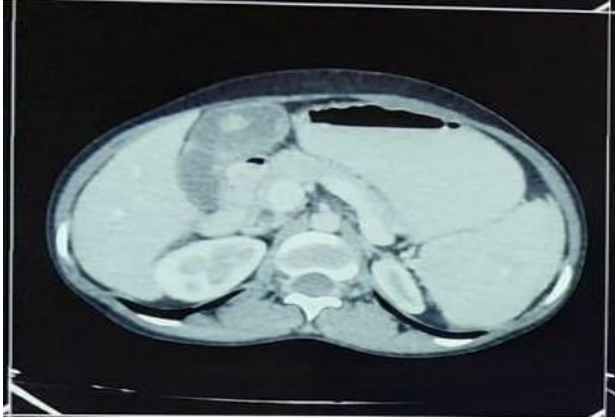


Figure 6: CT scan axial section showing a small enhancing polypoid lesion in fundus.



Figure 7: MRCP showing a small polypoid lesion from the fundus of gallbladder.



Figure 8: Gallbladder showing a small papillary lesion from fundus.

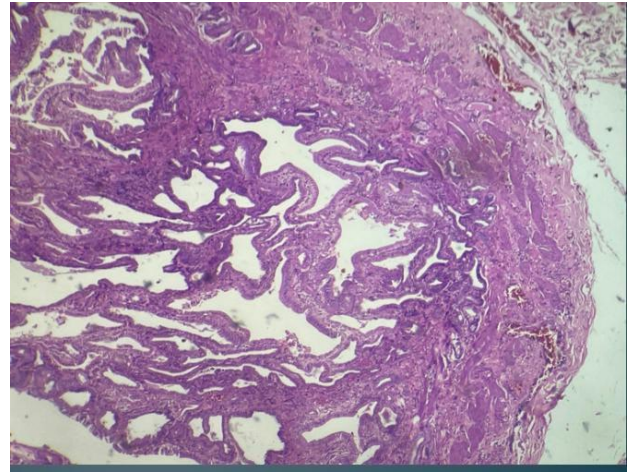


Figure 9: Histopathology showing papillary growth pattern.

DISCUSSION

Papillary neoplasms of the biliary system were reported in the past in various literature. However, it was included in the WHO classification of biliary tumours in 2010.¹ They are tumours showing distinct patterns of evolution and symptomatology.¹ It's a rare group of neoplasms with malignant potential. In fact, it is considered a forerunner of malignancy and the progression to malignancy due to the associated genetic alterations.²

Intraductal papillary neoplasm of the bile duct (IPN-B) is a rare bile duct neoplasm, characterized by endophytic proliferation of biliary epithelium with fibrovascular stalks within the duct lumen, sometimes with mucin hyper secretion and cystic dilatation of affected bile ducts.² They are the histological counterparts of intraductal papillary mucinous neoplasm of the pancreas, but with or without mucin secretion.^{1,2} It's mostly found in East Asian countries where there is a high incidence of hepatolithiasis and clonorchiasis infestations.^{3,4} It has slight male preponderance and an age predilection towards the 5th to 6th decades.^{3,4} They may harbour areas of low-grade and high-grade dysplasia or even foci of carcinoma in situ. It may mimic cholangiocarcinoma clinically by presenting as a mass lesion with jaundice.^{5,6} Imaging most often shows a lesion filling the lumen and causing biliary obstruction with or without lobar atrophy.⁷ Surgical management in the form of extra hepatic bile duct resection and hepatectomy to achieve negative margins. The prognosis is better than hilar cholangiocarcinoma with a 5-year survival up to 80%.⁸ Recurrence depends on the invasiveness and may be up to 20-60%.⁹ Different histological types are identified in papillary neoplasms- gastric, pancreatobiliary intestinal and oncocytic and express MUC1 and CK7 positivity.¹⁰

Intracholecystic papillary neoplasms (ICPN) are rare, non-invasive epithelial neoplasms of the gallbladder counterpart of IPN B. It is found in 0.4% of GB specimens.^{10,11} They are more common in women, mainly

in the 5th and 6th decades.¹¹ It may be an incidental finding or can present with upper abdominal pain. Commonly used modalities include ultrasound sonography (USG), computed tomography (CT) scan, and magnetic resonance imaging (MRI) which often misdiagnose these lesions as malignancy. Treatment will be cholecystectomy alone. The prognosis of ICPN lies between that of benign lesions and malignant lesions. ICPN can show foci of low-grade to high-grade dysplasia. It can evolve finally into an invasive carcinoma, and the histological findings may show areas of focal high-grade dysplasia or even invasiveness. ICPN is distinguished from papillary adenocarcinoma as ICPN rarely infiltrates and metastasizes, and the prognosis for ICPN is typically much better than that for gallbladder adenocarcinoma. The 5-year survival rate for ICPN is 60% if including invasive carcinoma and 78% if non-invasive.^{10,11} In contrast, the 5-year survival rate for gallbladder adenocarcinoma is 30%. The difference in prognosis may be due to their endophytic nature of growth nature causing symptoms earlier in the natural course and thereby earlier diagnosis or an inherent more indolent biology.¹¹ Therefore, it is important to diagnose ICPN correctly.

CONCLUSION

Papillary neoplasms of biliary tract may be suspected in cases where there is a polypoid lesion projecting from the wall of the biliary system, however confirmation and differentiation from malignancy is extremely difficult. It should always be approached like a malignancy and has to undergo oncological resections. Diagnosis and the degree of dysplasia or foci of invasiveness can be confirmed only by final histopathological examination. Chemotherapy can be omitted and patients may be put on long term follow up.

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