CASE REPORT

Intraductal Papillary Neoplasm of the Bile Duct: Radiological Diagnosis of a Rare Entity: Case Series

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Abstract

Background: Intraductal papillary neoplasm of the bile duct (IPNB) is a rare bile duct neoplasm characterized by an intraluminal papillary growth pattern in bile ducts and is considered a biliary counterpart of intraductal papillary mucinous neoplasm of the pancreas.

Case description: We report here two cases. (1) A case of a 34-year-old woman who presented with complaints of pain in the abdomen, jaundice, and pruritus. Further radiological investigations revealed the possibility of an IPNB, which was confirmed on histopathology. (2) A case of a 61-year-old man who was a known case of Barrett's esophagus and presented with complaints of right upper abdomen and jaundice. Radiological investigations and histopathology further confirmed the diagnosis of an IPNB involving the extrahepatic bile ducts. The purpose of this article is to highlight the role of computed tomography (CT) and magnetic resonance imaging in reaching this challenging diagnosis.

Conclusion: Although the diagnosis is not straightforward, imaging plays a great role in raising the possibility of an IPNB. The correct preoperative diagnosis is necessary for accurate surgical planning and resection.

Keywords: Biliary papillomatosis, Cholangiocarcinoma, Intraductal papillary mucinous neoplasm, Intraductal papillary neoplasm of bile duct.

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Introduction

According to the World Health Organization (WHO) classification, papillary or villous neoplasm covering the fragile fibrovascular stalks and occurring in the bile ducts is classified as intraductal papillary neoplasm of the bile duct (IPNB).[1] Papillary neoplasm of the bile duct along with previous entities, such as biliary papilloma and papillomatosis were newly categorized as bile duct tumors by the WHO.[2] In between 40 and 80% of instances, there is an invasive component. It is now considered a cholangiocarcinoma precursor lesion.[3] According to some authors, IPNBs are believed to be the biliary analog of intraductal papillary mucinous neoplasms of pancreas.[4] Papillary neoplasm of the bile ducts are frequently associated with mucin overproduction and bile duct dilatation. Based on histology and immunophenotypical profile, four subtypes have been identified: pancreatobiliary, intestinal, gastric, and oncocytic.[5,6] The most common subtype is pancreatobiliary.

Risk factors for IPNB are multifactorial and most commonly include infection by clonorchiasis and hepatolithiasis. Other important risk factors include Gardner syndrome, familial adenomatous polyposis, and choledochal cysts. Compared to Western countries, Eastern countries have a higher incidence of IPNB, which may be attributed to endemic clonorchiasis infections and a higher prevalence of hepatolithiasis. Commonly encountered symptoms include intermittent abdominal pain, jaundice, and fever.

We report here two histopathologically proven cases of IPNB with different presentations and sites of involvement, their imaging features and differentials to consider when we encounter this entity.

Case Description

Case 1

A 34-year-old woman came to routine outpatient department (OPD) with chief complaints of pain in the abdomen, jaundice, and pruritus. There was no history of recent fever. Her past medical records were insignificant, except for a cholecystectomy done in view of cholelithiasis. Her blood workup revealed obstructive jaundice: total bilirubin – 4.2 mg/dL (normal range: 0.2–1.4 mg/dL), direct bilirubin – 3.1 mg/dL (normal range: 0–0.4 mg/dL), AST – 82 IU/L (normal range: 15–45 IU/L), ALT – 152 IU/L (normal range: 40–122 IU/L). Hepatitis A, B, or C were not detected in the serologic evaluation.

Ultrasound (US) of the upper abdomen revealed severe intrahepatic biliary dilatation involving the right lobe of the liver with multiple hyperechoic masses within the dilated bile ducts, which showed minimal vascularity on color Doppler examination (Fig. 1). Then, triple phase contrast-enhanced computed tomography (CECT) of the abdomen along with magnetic resonance cholangiopancreatography (MRCP) was performed.

Triple-phase CECT abdomen revealed multiple ill-defined variable sized lobulated lesions within dilated biliary radicals, showing mild enhancement in the late arterial phase in the right lobe of the liver. The lesions were seen to extend along and expand

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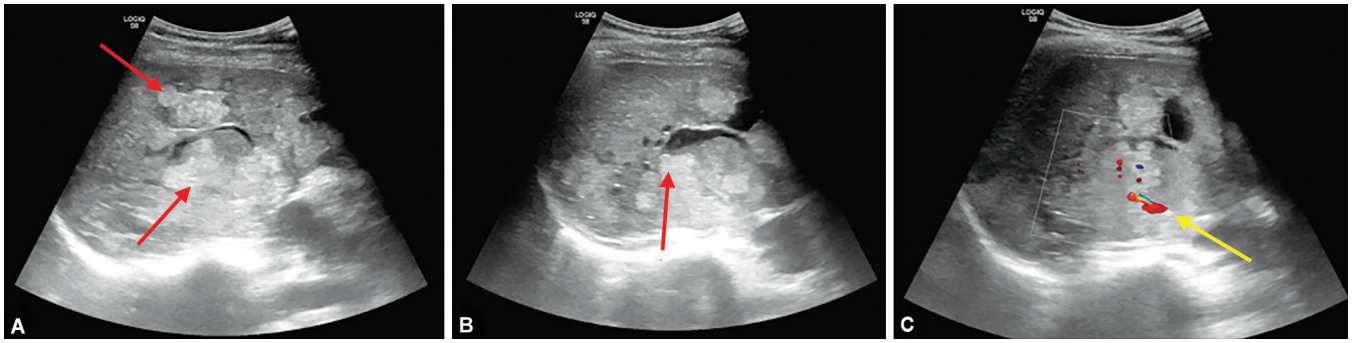
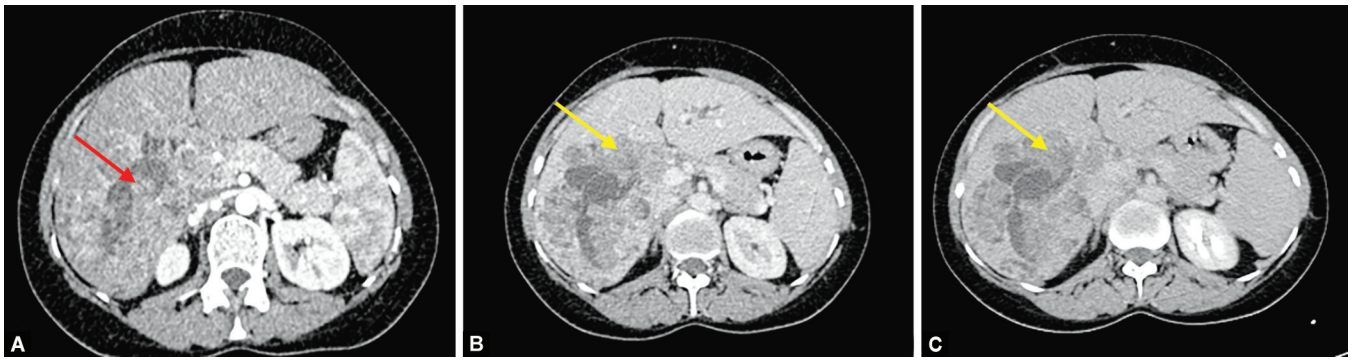


Figs 1A to C: Ultrasound of the upper abdomen. (A and B) Multiple hyperechoic masses within the dilated biliary radicals in right lobe of liver (red arrows); (C) On color Doppler, the lesions showed minimal vascularity (yellow arrow)



Figs 2A to C: Triple phase CECT abdomen in (A) late arterial, (B) portal venous, and (C) delayed phases show multiple ill-defined variable sized lobulated lesions within dilated biliary radicals showing mild enhancement in late arterial phase (red arrow in A) with washout in portal venous and delayed phases (yellow arrows in B and C)

the right hepatic duct branches and reach up to the surface of liver causing mild contour bulge and irregularity (Fig. 2).

MRCP revealed multiple intraluminal papillary masses filling the suprapancreatic common bile duct, extending proximally into the common hepatic duct, primary confluence and further into the right hepatic duct and sectoral ducts of segments V, VI, and VII of the right lobe of the liver causing their aneurysmal dilatation. They appeared intermediate in signal intensity on T1-weighted images, hyperintense on T2-weighted images and showed patchy areas of diffusion restriction. The lesions showed hypoenhancement (with respect to liver parenchyma) on post-contrast images (Fig. 3).

Based on imaging findings, radiological diagnosis IPNB was given. The final pathology report revealed a pancreatobiliary type of IPNB.

Case 2

A 61-year-old gentleman with a prior history of gastroesophageal reflux disease (GERD) came to OPD with complaints of pain over the right upper abdomen and progressive jaundice. He had been taking proton pump inhibitors for GERD for the past 6 years. Upper gastrointestinal (GI) endoscopy with biopsy revealed features of Barrett's esophagus. In view of progressive jaundice and pain in the right upper abdomen, US was advised.

Ultrasound of the upper abdomen revealed hyperechoic masses filling and expanding the common hepatic and common bile ducts. The lesion did not show any significant internal vascularity on color Doppler (Fig. 4). Intrahepatic biliary radical dilatation was noted in both lobes of the liver.

Further evaluation was done with MRCP, which revealed an irregular hypoenhancing soft tissue lesion in the common hepatic duct and proximal common bile duct causing dilatation of the same, appearing hypointense on T1-weighted images and heterogeneously hyperintense on T2-weighted images, with few areas of diffusion restriction within along with no significant post-contrast enhancement with bilobar intrahepatic biliary radical dilatation (Fig. 5).

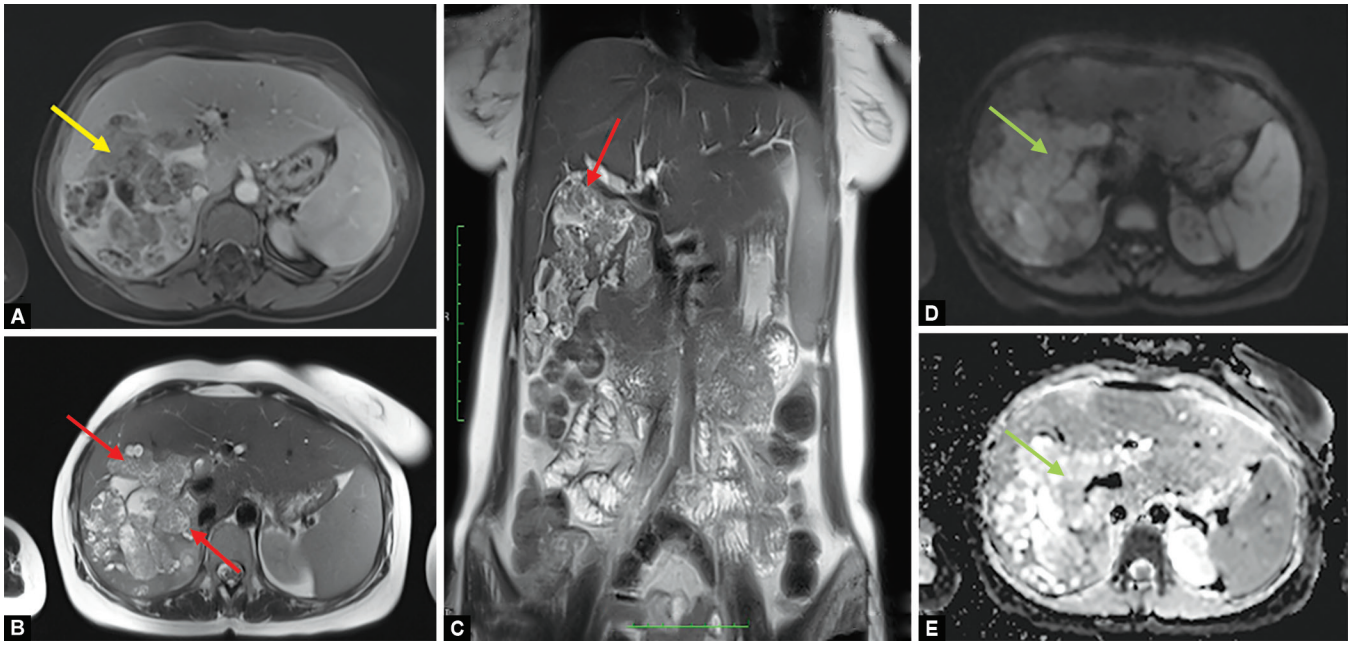
Based on imaging findings, the possibility IPNB was given, with a remote possibility of cholangiocarcinoma. Histopathology was advised, which confirmed the diagnosis of the intestinal type of IPNB.

Discussion

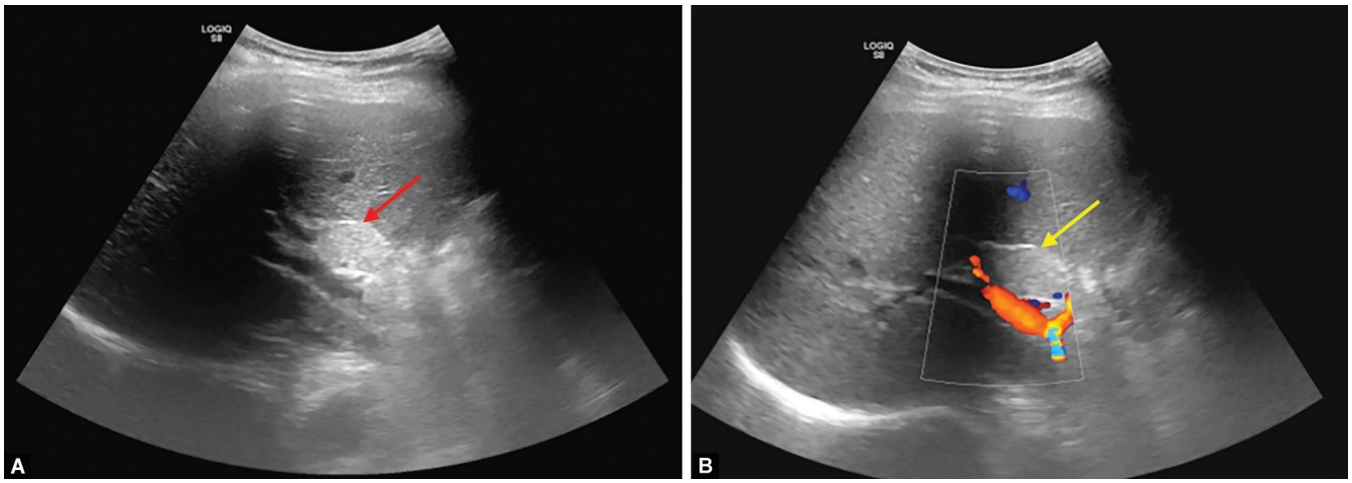
Approximately, 9–38% of all bile duct malignancies are IPNBs.[4] The majority of patients are between the ages of 50 and 70 years, with a mild male predominance. Given that invasive carcinoma, tubular or mucinous adenocarcinoma, or both, are linked to 40 to 80% of IPNBs, IPNB is thought to have a high risk of developing cancer. Both intrahepatic and extrahepatic bile ducts are susceptible to the development of IPNB. Right hypochondriac pain, cholangitis, and obstructive jaundice are the most frequent clinical presentations. Tumor emboli, stones, and macroscopic mucin hypersecretion are possible causes of obstruction in cases of IPNBs. CA19-9 and carcinoembryonic antigen levels may be elevated.

Imaging Appearance

The balance between papillary proliferation and mucin production determines the imaging appearance of IPNB. It appears as an intraductal mass producing upstream biliary system dilatation when there is a predominance of papillary proliferation and minimal



Figs 3A to E: (A) Axial T1 weighted post-contrast and axial and coronal T2-weighted images. (B and C) Intraluminal papillary masses appearing hyperintense on T2-weighted images filling the suprapancreatic common bile duct, extending proximally into common hepatic duct, primary confluence and further into right hepatic duct and sectoral ducts of segments V, VI, and VII of right lobe of liver causing their aneurysmal dilatation (red arrows in B and C) with no significant post-contrast enhancement (yellow arrow in A); (D and E) Diffusion-weighted images with ADC maps show areas of restricted diffusion within the lesion (green arrow)



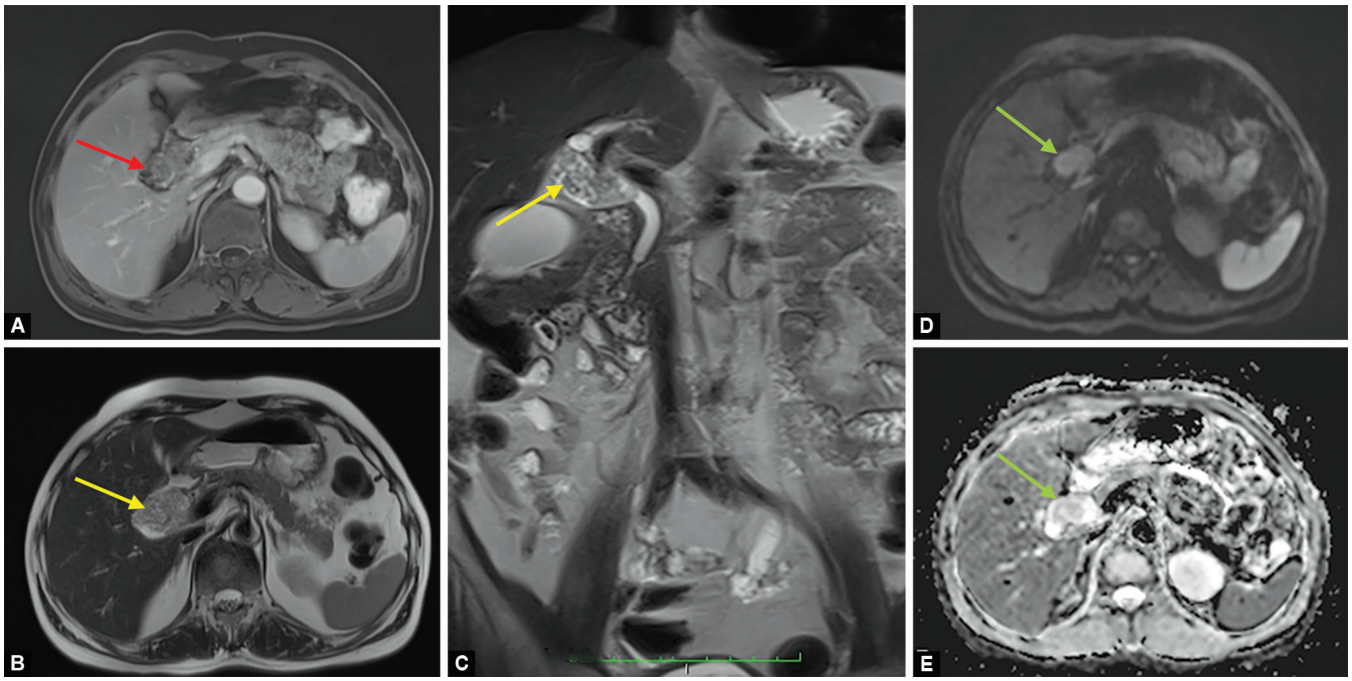
Figs 4A and B: Ultrasound of the upper abdomen. (A) Hyperechoic mass filling and expanding the common hepatic and proximal common bile duct (red arrow); (B) on color Doppler, the lesions no significant internal vascularity (yellow arrow)

mucin formation. IPNB produces diffuse bile duct dilatation in cases of high mucin production and low papillary proliferation with no detectable mass on imaging.[3] However, an intraductal tumor with proximal and occasionally distal bile duct dilatation is how IPNB most commonly manifests. Bile duct dilation can be tubular, fusiform, or cystic in nature. According to some authors, a characteristic sign of IPNB is aneurysmal dilatation of a bile duct branch.[7]

There are five imaging patterns that can be seen with IPNBs.[8] Type I is the most common pattern, in which there is diffuse duct dilatation along with an intraductal tumor. The 2nd common pattern is type II pattern, which exhibits diffuse duct dilatation without any obvious visible mass. Type III demonstrates localized duct dilation along with an intraductal papillary mass. Type IV is the least common pattern and demonstrates cast-like intraductal lesions that completely fill the lumen. Type V shows minimal proximal ductal dilatation and a focal stricture-like lesion.

On ultrasonography, in up to 41% of patients with IPNB, an intraductal hypoechoic or hyperechoic lesion might be seen. Since mucin and bile are both anechoic, US may not be able to detect their presence. When compared to the surrounding hepatic parenchyma, the intraductal mass is typically either iso- or hyperattenuating on computed tomography (CT) and may enhance on the late arterial phase.

On T1-weighted images, IPNB appears iso- to hypointense, and on T2-weighted images, IPNB generally shows signal intensity that



Figs 5A to E: (A) Axial T1 weighted post-contrast and (B and C) axial and coronal T2-weighted images show irregular hypoenhancing (red arrow in A) soft tissue lesion in common hepatic duct and proximal common bile duct causing dilatation of the same (yellow arrow in B and C) appearing hyperintense on T2-weighted images; (D and E) Diffusion-weighted images with ADC maps show areas of restricted diffusion within the lesion (green arrow)

is higher when compared to liver parenchyma. In the late arterial phase, IPNB frequently exhibits iso or hyperintensity, but it does not continue to be hyperintense in the portal venous and delayed phases.[9] Diffusion restriction may be seen in the solid component of the mass. Because mucin looks similar to bile on CT and magnetic resonance imaging, it can be challenging to detect mucin in patients with IPNB.

The two main differential IPNBs include hepatic mucinous cystic neoplasm (HMCN) and cholangiocarcinoma.[2] HMCN is a cystic epithelial tumor that lacks communication with the bile ducts and has a typical ovarian-type stroma. Cholangiocarcinoma patients are older age groups (>60 years) and have a male predominance.

Conclusion

In spite of advancements in imaging techniques, the radiological diagnosis of IPNB remains difficult. In instances where the imaging findings are similar those as described above, there should be a high level of suspicion for this condition, and a preoperative diagnosis using cytology and molecular techniques should be attempted.

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