

Intraductal papillary mucinous neoplasm of the bile ducts: a rare form of premalignant lesion of invasive cholangiocarcinoma

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Learning objectives

Review the forms of presentation of intraductal papillary mucinous neoplasm of the bile ducts (IMP-N-B) and describe the radiological findings.

Background

Cholangiocarcinoma is a malignant tumor arising from the epithelium of the bile ducts, and tends to have a poor prognosis and high morbidity. Most of these tumors are well-, moderately, or poorly differentiated adenocarcinomas. Is the second most common primary malignant neoplasm (first is the hepatocarcinoma). It accounts for approximately 10-20 % of liver cancers. Incidence is usually in the elderly, with peak prevalence during the 7th decade of life, and has a slight male predilection.

Cholangiocarcinomas can be either intra or extrahepatic. Intrahepatic cholangiocarcinoma has several growth patterns, classified into mass-forming, periductal-infiltrating and intraductal-growing types.

Mass-forming type is characterized by a homogeneous large mass (sometimes more than 15 cm in diameter), with an irregular but well-defined margin and is frequently associated with dilatation of the biliary trees in the tumor periphery.

Periductal-infiltrating type grows along a dilated or narrowed bile ducts without mass formation, and is therefore elongated, spiculated or branchlike. Is rare in intrahepatic cholangiocarcinoma, but are most common at the hilum.

Intraductal growing type are characterised by alterations in duct caliber, usually ductectasia with or without a visible mass. If a mass is visible, are usually small, sessile, or polypoid, often spreading superficially along the mucosal surface and resulting in multiple tumors (papillomatosis).

Intraductal papillary mucinous tumor of the bile ducts is a variant that produces a large amount of mucin. This tumor bears a striking similarity to intraductal papillary tumor of the pancreas.

Both proximal and distal bile ducts to the tumor are dilated because mucin may obstruct the papilla of Vater.

An intraductal tumor can be observed as a papillary mass or as bile duct wall thickening, with sizes of papillary tumors range from 1 to 2 cm.

Other patients show diffuse dilatation of the bile ducts without evidence of mass.

Some intrahepatic papillary mucinous tumors may produce aneurysmlike cystic dilatation of the bile ducts.

These three types of IMPN-B are shown in the following section.

Findings and procedure details

We retrospectively reviewed the images of ultrasound, CT and MRI of 3 patients with diagnosis of intraductal papillary mucinous neoplasm of the bile ducts.

Growth forms are basically divided into three patterns and we bring a patient of each type:

1. Polyps associated with diffuse ductal dilatation (first patient, 73 years old woman) (Figure 1, 2 and 3).
2. Diffuse ductal dilatation without evidence of mass (second patient, 78 years old man) (Figure 4).
3. Cystic dilatation with mural nodule or excrescencies in the cyst wall (third patient, 36 years old woman). (Figure 5 and 6)

IMAGES



Fig. 1: 73 years old woman. US shows a dilated common bile duct with presence of echogenic mass (white arrow in B). The intrahepatic bile duct is dilated (black arrow in C).

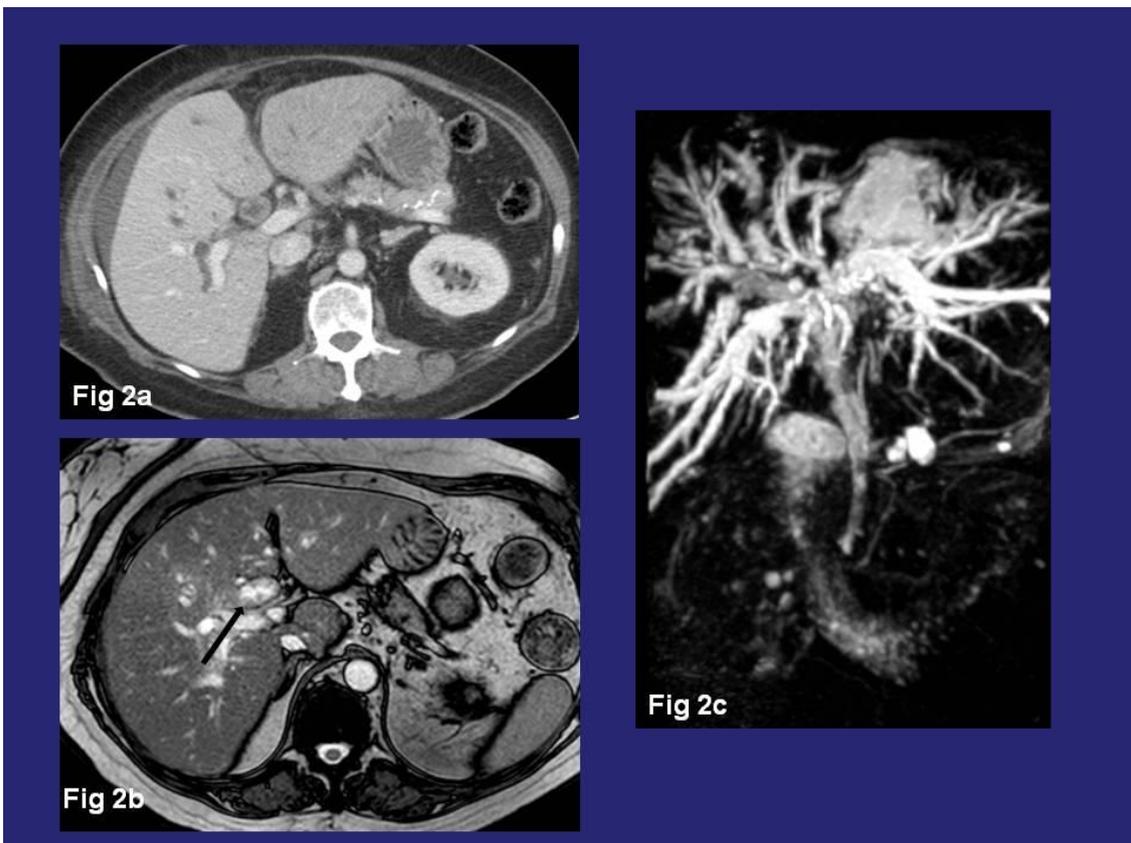


Fig. 2: Patient of figure 1. Contrast-enhanced CT (figure 2a) shows dilated common bile duct, with presence of multiple polyps on their wall; and dilated peripheral bile ducts. T2-weighted MR image (figure 2b) shows multiple polypoid tumors (black arrow). MR cholangiopancreatography (figure 2c) shows markedly dilated intrahepatic ducts.

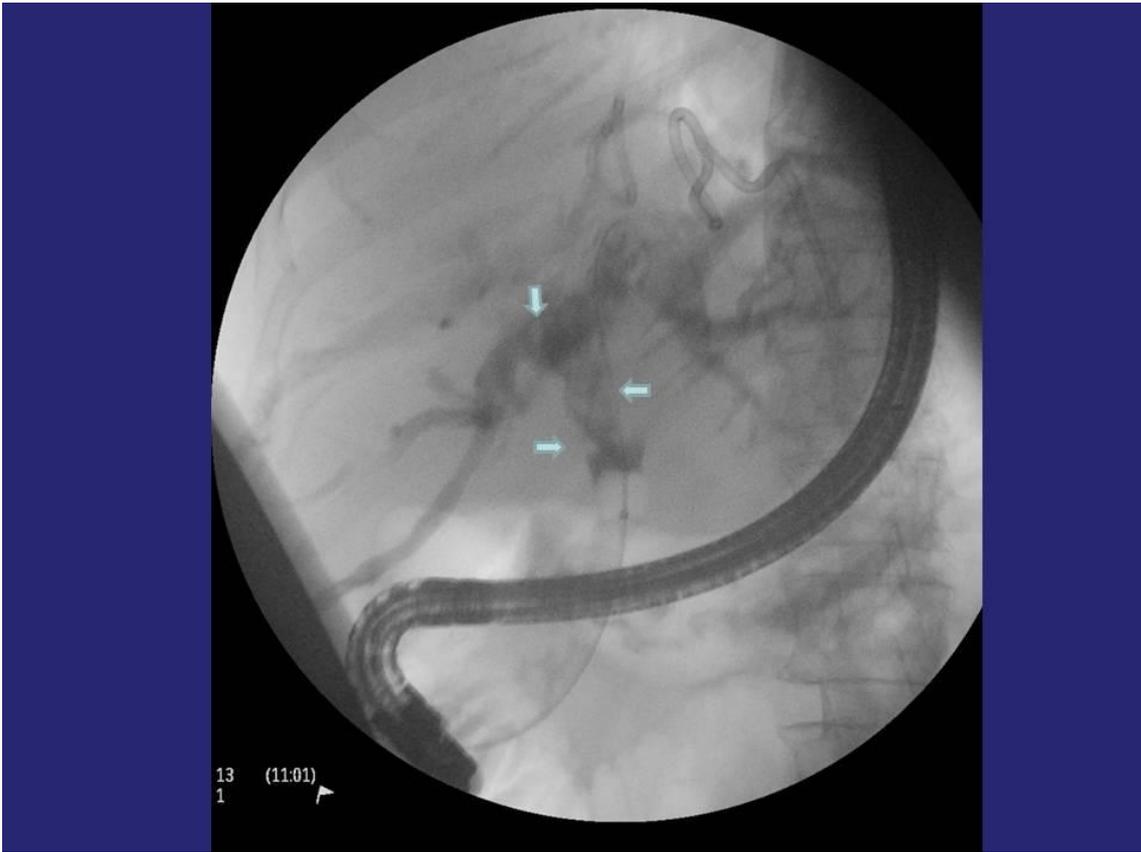


Fig. 3: Patient of figure 1 and 2. Endoscopic retrograde cholangiogram shows multiple filling defects in the bile duct (arrows), representing the polyps.

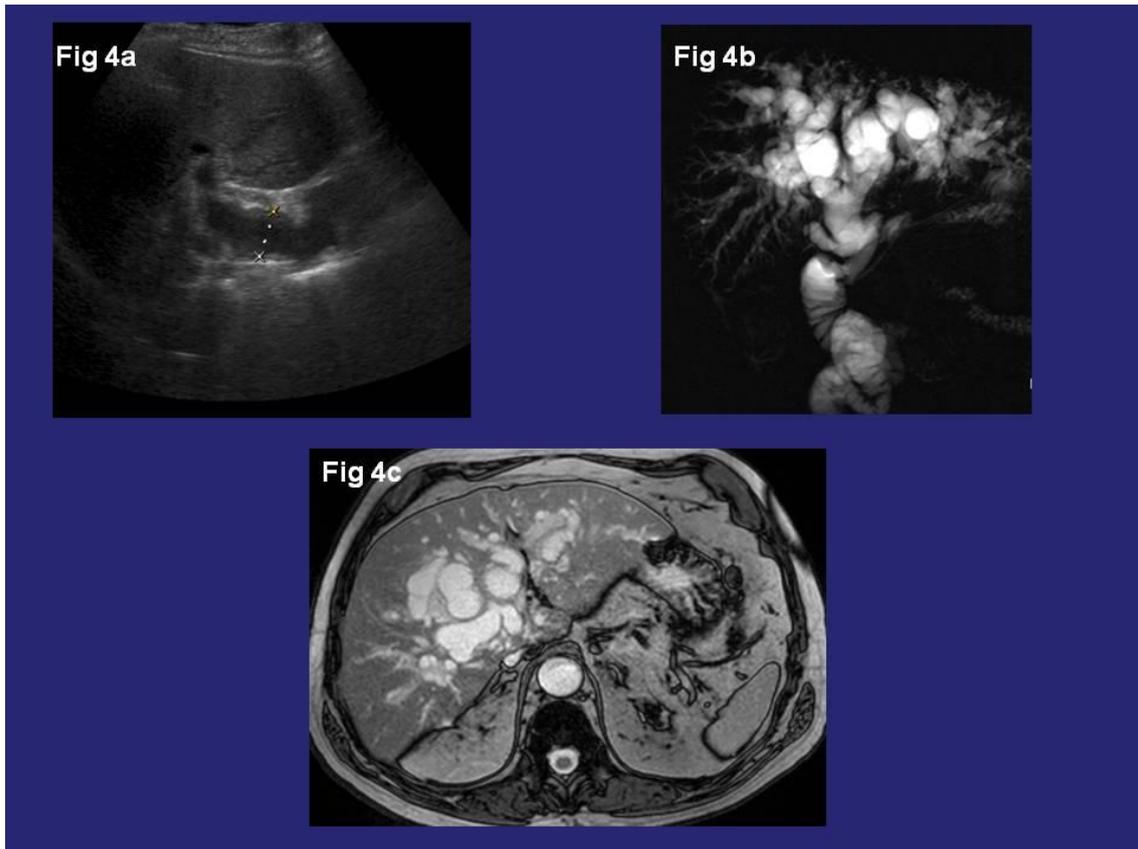


Fig. 4: 78 years old man with diagnosis of IPMN-B. US shows a dilated common bile duct (figure 4a). MR cholangiopancreatography (figure 4b) shows a massively dilated bile duct without evidence of obstructing lesion. T2-weighted MR image (figure 4c) shows the same findings as in a and b.

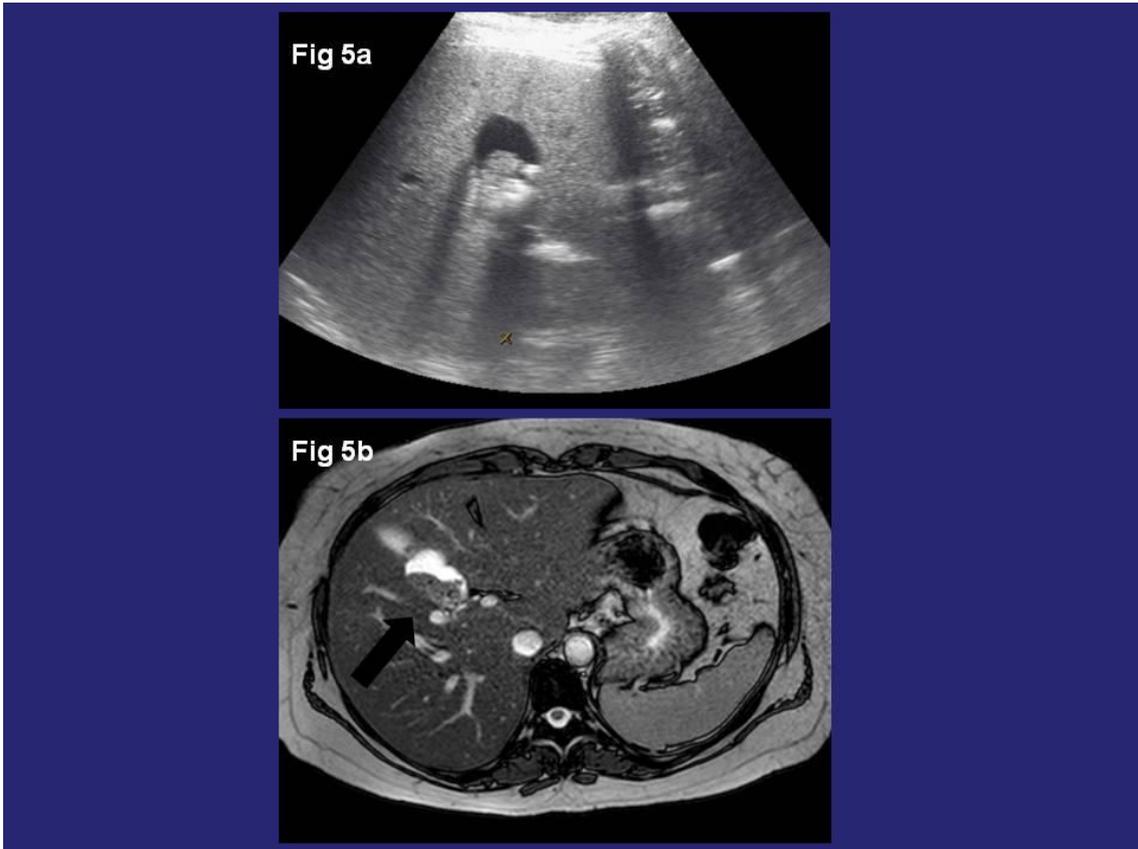


Fig. 5: 36 years old woman. US image (figure 5a) shows a complex-cystic lesion, with presence of an echogenic mass. T2-weighted MR image (figure 5b) shows a cystic dilatation of the bile duct, whit a polypoid tumor (black arrow) in the wall of the cyst.

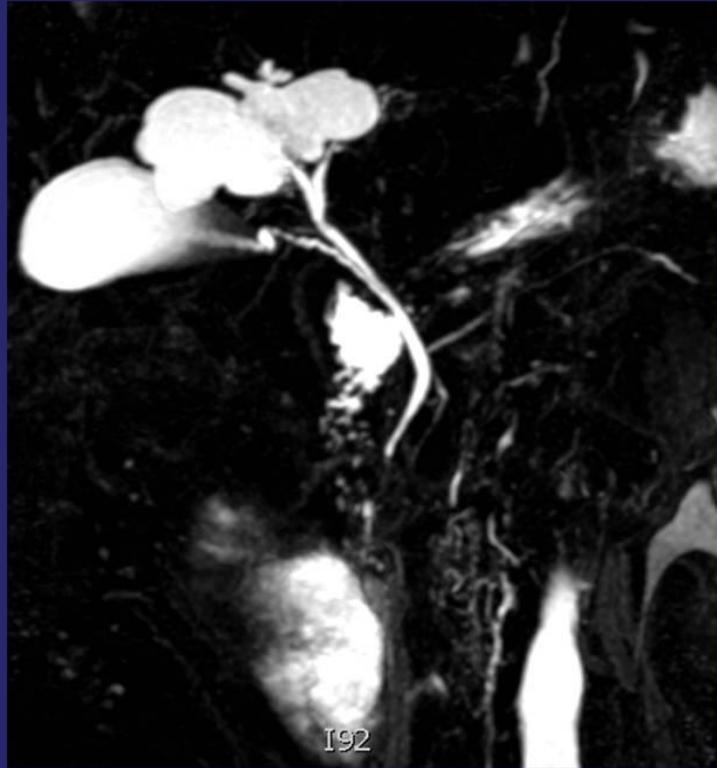


Fig. 6: Patient of figure 5. MR Cholangiopancreatography show cystic dilatation of the bile duct. The other biliar tract is normal.

Conclusion

The mucinous variant of intraductal papillary tumors of the bile duct are a rare variant presentation of premalignant lesions of cholangiocarcinoma. They are characterized by high production of mucin (they bears a similarity to pancreatic IPMT), have a slow growth and a low rate of invasive forms. Knowledge and characterization of this entity are important because it has a better prognosis than other forms of cholangiocarcinoma (mass-forming and periductal-infiltrating).

References

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