INTRADUCTAL PAPILLARY MUCINOUS TUMOR OF THE BILE DUCTS: A Multimodality Pictorial Review



Radiology Department University Hospitals of Coimbra

Director, Prof. Dr. Filipe Caseiro Alves

Rodrigues, L; Antunes, C; Curvo Semedo, L; Carvalheiro, Vítor; Caseiro Alves, Filipe.

Learning Objectives:

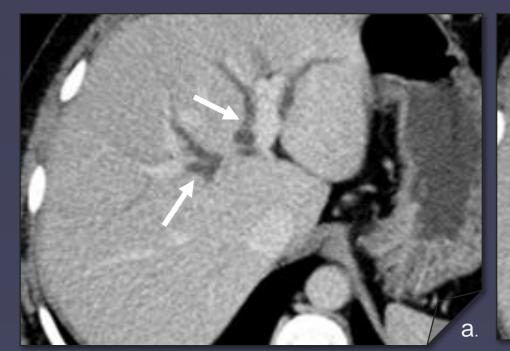
To provide a systemic review of the imaging features of papillary tumors of the bile ducts with emphasis on CT and MRI.

Background:

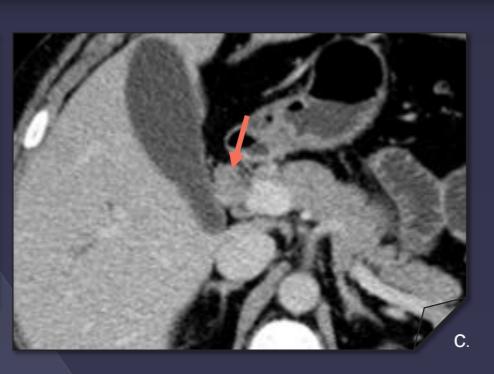
Papillary tumors of the bile ducts are intraductal tumors with numerous frondlike papillary projections. Some of them produce a large amount of mucin that disturbs bile flow and causes severe biliary dilatation. Its preoperative recognition is important in differentiation from high-grade malignant disease.

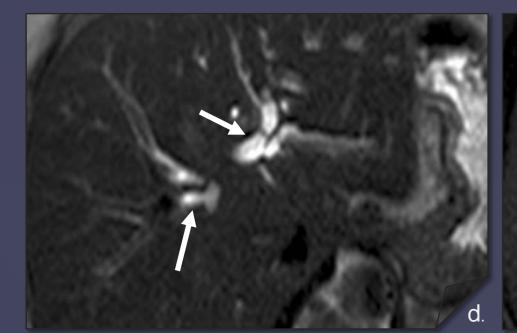
Imaging Findings:

In the presence of an intraductal mucin-hypersecreting papillary tumor, the recognition of the fundamental characteristic imaging features – dilatation of biliary tree, the presence of mucin and the tumor itself – allows for a correct diagnosis to be made. The suggestive findings of this disease will be illustrated through a multimodality imaging approach and analysed for its contribution to refine differential diagnosis.









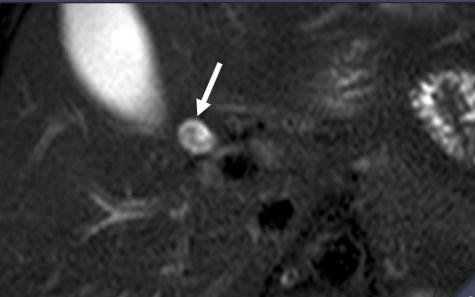


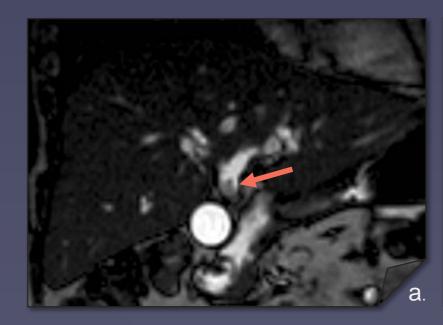


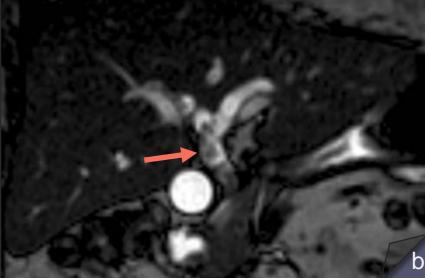
Figure 1 – (a, b, c) Venous phase axial CT images show moderately dilated right and left intrahepatic bile ducts and clear dilatation of upper extrahepatic bile duct (white arrows). A fungating mass occupying the entire lumen of the intrapancreatic common bile duct is visible (orange arrow). (d, e, f) T2-weighted axial fat-sat MR images showing the bile duct dilatation (white arrows) and hypointense signal at the intrapancreatic common bile duct lumen, where the tumoral mass is located (orange arrow).

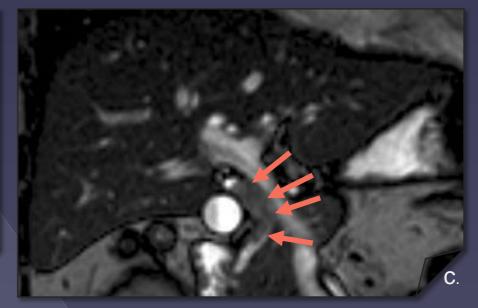
Compared with other types of cholangiocarcinoma, intraductal papillary mucinous tumor is a low-grade malignancy that is generally limited to the mucosa. Clinical symptons are recurrent (episodic biliary pain, fever, chills, jaundice) and related to the recurrent obstruction of the bile ducts by excessive mucin or by tumor-induced stenosis.

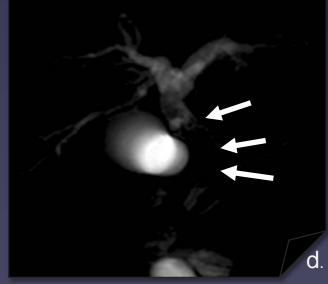
- A) Tumor The intraductal papillary mucinous tumor may appear as a small and flat or fungating mass in the dilated bile ducts (Figs 1c, 1f, 2) and are depicted at US, CT, ERCP or MR cholangiopancreatography. However, tumors that are relatively small or sessile or that spread along the mucosal surface are difficult to visualize with imaging of any modality. Tumor localization is essential for surgical planning.
- Mucin At US, mucin is usually anechoic, like bile. The attenuation of mucin at CT and its signal intensity at MRI is the same as that of water, making mucin impossible to detect by these imaging modalities. At ERCP or PTC, however, mucin lumps may be depicted as multiple elongated linear, ovoid or amorphous filling defects in the dilated bile ducts (Fig 3c).

Figure 2 – (a, b, c) T2-weighted coronal MR images of the same patient show multiple endoluminal hypointense areas#... (orange arrows). (d) MIP reformatted image showing the lengthy endoluminal filling defect caused by a the fungating tumor (white arrows).









c) Bile Duct Dilatation – The biliary tree becomes dilated when either tumor or mucin impedes the bile flow through the bile ducts. In the presence of mucin over-production, the entire biliary tree is markedly dilated (Fig 1), irrespective of the location of the primary mucin-producing tumor, because the hepatopancreatic ampulla is obstructed by a large amount of viscous mucin. When a tumor develops in a hepatic lobar bile duct, the bile ducts in that lobe are disproportionately more dilated than the ducts in contralateral lobe (Figs 3a, 3b, 3d, 3e). Bile ducts that contain a tumor also may show focal dilatation like that of an aneurysm (Fig 3c). Dilated peripheral intrahepatic bile ducts have a propensity to rupture, causing the formation of a mucin collection or the implantation of tumor cells in the peritoneal space (pseudomyxoma *peritonei*).





Figure 3 – (a) Venous phase axial CT image shows severe dilatation of the left and right intrahepatic bile ducts and marked dilatation of the extrahepatic bile ducts but no tumor. Note the disproportionately greater dilatation of the left hepatic bile ducts. (b) ERCP image shows, once more, severe dilatation of the biliary tree, especially the left hepatic ducts and multiple cordlike filling defects (white arrow) that represent mucin in the extrahepatic bile ducts. Note also the oval filling defect (black arrow), which indicates the presence of a nodule in the lateral segmental bile duct of the left hepatic lobe. (c) Venous phase axial CT image show aneurysmal dilatation of the lateral segmental bile ducts of the left hepatic lobe containing multiple fungating masses. (d) Venous phase axial CT image and (e) axial US image show a nodular mass (black and white arrows) causing severe dilatation of the lateral segmental bile ducts of the left hepatic lobe.

Conclusion:

On the basis of this characteristic pattern of biliary dilatation and intraductal tumor depicted at US, CT, ERCP or MR cholangiopancreatography, one can make a diagnosis of intraductal papillary mucinous tumor of the bile ducts and avoid unnecessary aggressive surgical therapy.