Case 11157
Intraductal papillary mucinous tumour of the biliary tract

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Section: Abdominal Imaging
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Patient: 73 year(s), female

Authors' Institution

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Clinical History

We report a case of a 73 year-old female patient presenting with recurrent episodes of right upper quadrant abdominal pain and fever. Mild jaundice was inconstantly present. These symptoms were related to recurrent cholangitis and resolved after antibiotic therapy.

Imaging Findings

Abdominal MRI showed marked, regular dilatation of the intra and extrahepatic bile ducts (Fig. 1). Multiple filling defects of intermediate signal intensity on T2-weighted imaging were visible, arising from the bile ducts walls and protruding into the biliary lumen. Lesions were positive on Diffusion-weighted imaging (Fig. 2). The dominant lesion measured 15mm and arose from the lateral left wall, causing a partial obstruction of the bile duct (Fig. 3). Smaller lesions were seen in the confluence of the ducts of segment V and VIII. There were no focal hepatic lesions. A moderate splenomegaly and mild volume of ascites and pleural effusion were also visible.
Endoscopic Retrograde Cholangiopancreatography (ERCP) was performed confirming the regular dilatation of the biliary tree and multiple intraluminal papillary projections (Fig. 4). Mucus plugs

were seen bulging of the papilla into the duodenal lumen. Mucin was drained and the papillary lesions were biopsied during the examination.

Discussion

Intraductal papillary mucinous tumour (IPMT) of the biliary tract is defined as a non-invasive, mucin producing epithelial tumour [1]. It can be considered as the biliary counterpart of the pancreatic IPMT [1]. Some biliary IPMTs produce a large amount of mucin that disturbs the bile flow and causes severe biliary dilatation [2, 3]. Excessive mucin discharge may intermittently and partially impede bile flow causing obstructive jaundice, sometimes complicated by cholangitis or stone formation [3].

Imaging is essential to localize the tumour and to subsequent surgical planning [2]. Bile duct IPMTs appears as small polypoid or flat masses projecting in the lumen of the bile ducts [1, 2]. Small tumours that spread along the bile duct wall may be impossible to visualize [2]. Dilatation of the biliary tree is a common finding [2, 3]. If the tumour is localized in an intrahepatic duct, the bile ducts in the affected lobe are more dilated, sometimes with focal, aneurysmal dilatations containing the tumour [2]. A main duct tumour causes a more generalized dilatation, due to mucin overproduction with obstruction of the ampulla [2]. Mucin may be indistinguishable from papillary proliferation at ultrasound, CT or MRI [2]. ERCP easily confirms the presence of mucin and allows biopsy, confirming the diagnosis [2].

Bile duct IPMT is a low-grade malignancy that grows slowly and tends to spread along the mucosal surface of the bile ducts [1]. It is generally limited to the mucosa, although it can invade the ductal wall in a later phase [1, 2]. It is a non-invasive neoplasm that, in rare cases, can give rise to an invasive carcinoma [1, 4]. Surgery is the treatment of choice [4]. Depending on the location of the tumour it may include bile duct resection, pancreatoduodenectomy and major hepatic resection [1, 3, 4]. Long term survival is expected after surgery [3, 4].

Final Diagnosis

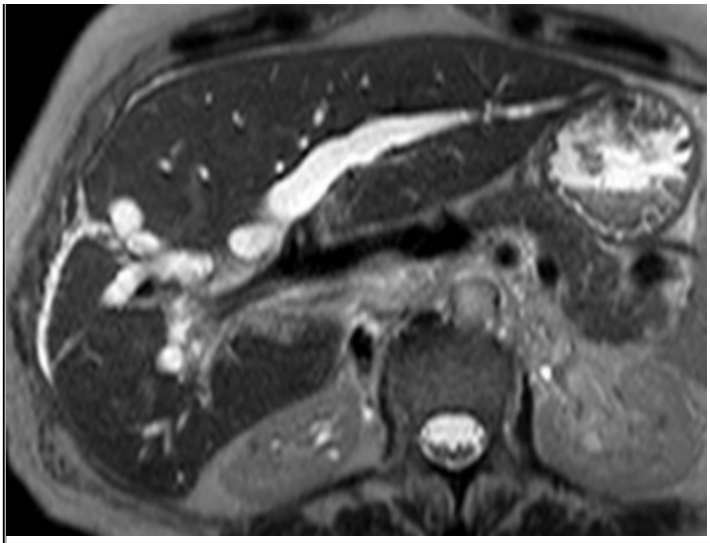
Intraductal papillary mucinous tumour of the biliary tract.

Differential Diagnosis List

Multifocal papillary cholangiocarcinoma, Multiple bile duct stones, Biliary metastasis

Figures

Figure 1 Abdominal MRI



Axial T2-weighted image shows smooth dilatation of the biliary tree, involving both intra and extrahepatic bile ducts.

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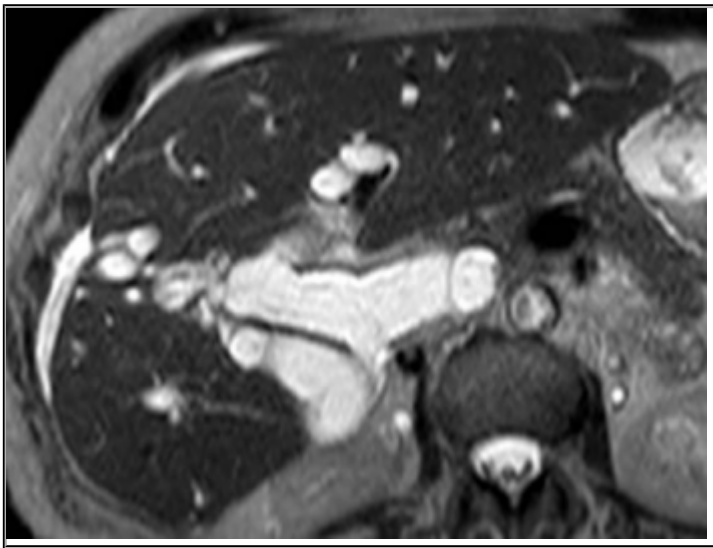
Area of Interest: Abdomen;
Imaging Technique: MR;
Procedure: Diagnostic procedure;
Special Focus: Dilatation;



Axial T2-weighted image shows smooth dilatation of the biliary tree, involving both intra and extrahepatic bile ducts.

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Area of Interest: Abdomen;
Imaging Technique: MR;
Procedure: Diagnostic procedure;
Special Focus: Dilatation;

Figure 2 Abdominal MRI

Coronal T2 weighted image shows papillary projections arising from the bile duct wall and protruding into the biliary lumen.

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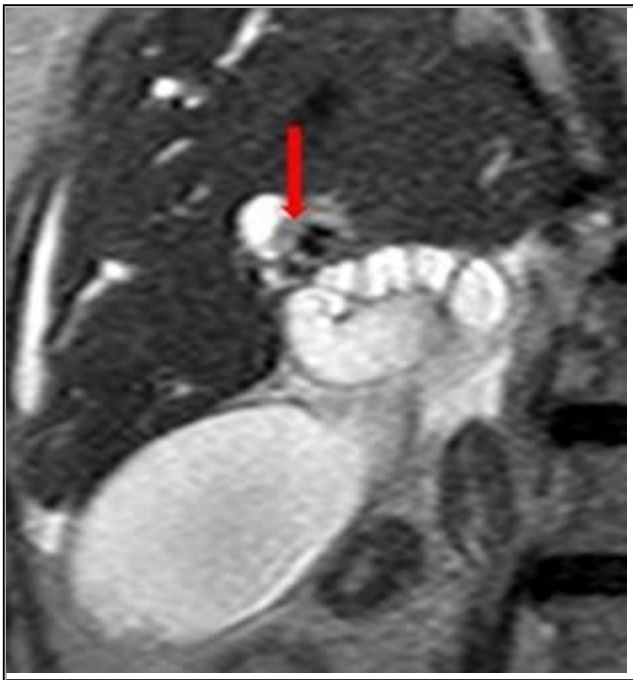
Area of Interest: Abdomen;
Imaging Technique: MR;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;



Coronal T2 weighted image shows papillary projections arising from the bile duct wall and protruding into the biliary lumen.

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Area of Interest: Abdomen;
Imaging Technique: MR;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;

Figure 3 Abdominal MRI

Coronal T2-weighted image shows a lesion arising from the left wall of the main bile duct, causing partial obstruction.

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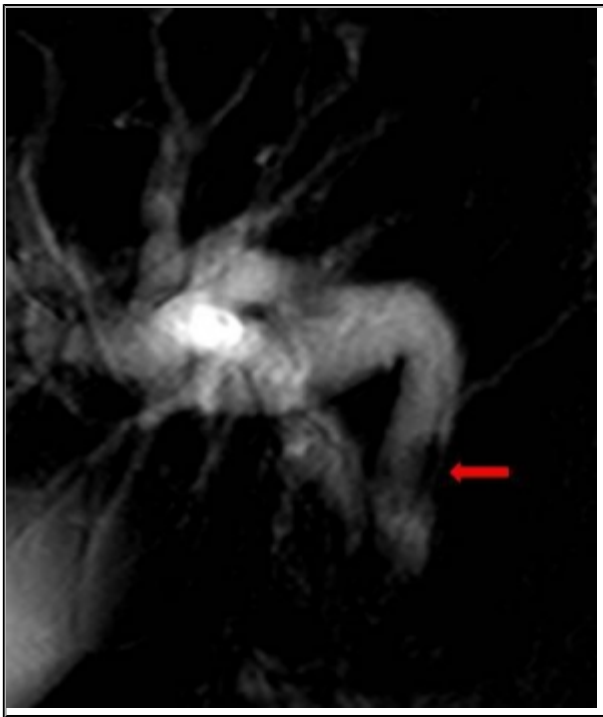
Area of Interest: Abdomen;
Imaging Technique: MR;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;



MRCP projection shows a lesion arising from the left wall of the main bile duct, causing partial obstruction.

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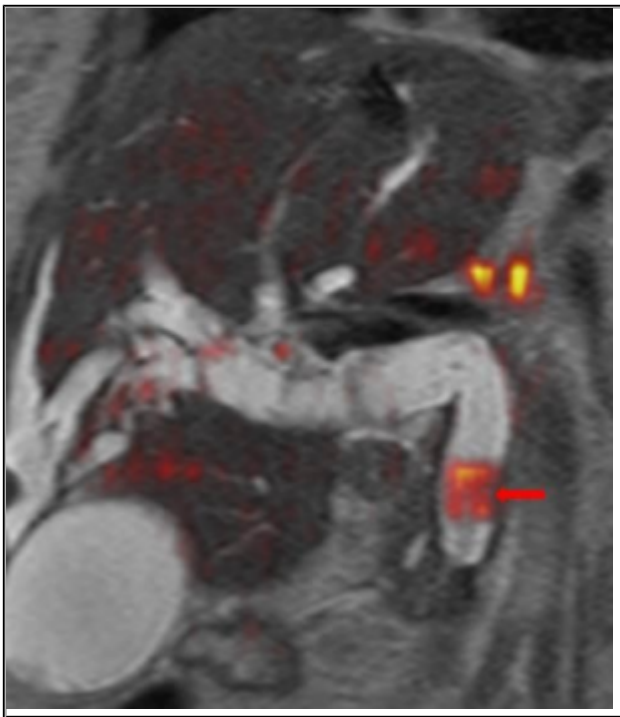
Area of Interest: Abdomen;
Imaging Technique: MR;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;



Diffusion-weighted imaging fused with coronal T2- weighted image shows a lesion arising from the left wall of the main bile duct, causing partial obstruction.

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Area of Interest: Abdomen;
Imaging Technique: MR-Diffusion/Perfusion;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;

Figure 4 ERCP

ERCP shows the dilated biliary tree and multiple filling defects in the bile ducts walls, corresponding to the projecting papillary tumours.

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Area of Interest: Abdomen;
Imaging Technique: Digital radiography;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;

ERCP shows the dilated biliary tree and multiple filling defects in the bile ducts walls, corresponding to the projecting papillary tumours.

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Area of Interest: Abdomen;
Imaging Technique: Digital radiography;

Procedure: Diagnostic procedure;
Special Focus: Neoplasia;

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Citation

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