Review of Radiologic manifestations in primary and secondary Sclerosing cholangitis

Poster No.: C-1346
Congress: ECR 2016
Type: Educational Exhibit
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Keywords: Inflammation, Cancer, Diagnostic procedure, Cholangiography, MR, Fluoroscopy, CT, Biliary Tract / Gallbladder, Abdomen
DOI: 10.1594/ecr2016/C-1346

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Learning objectives

To review the causes and radiological manifestations of primary and secondary sclerosing cholangitis as well as the complications of each disease entity.

Background

Sclerosing cholangitis refers to an entity of progressive chronic inflammatory disease of the intrahepatic and/or extra-hepatic biliary system characterized by inflammation, fibrosis and multifocal strictures. The most common form of sclerosing cholangitis is the primary form, of which 85% of cases are associated with inflammatory bowel disease.

The secondary form is related to various etiologies, which can be potentially curable. Cholangiocarcinoma can not only develop in 10-15% of the cases of primary sclerosing cholangitis but also in cases of the secondary form. MRI has the highest capability for detection of early cholangiocarcinoma, to evaluate its extent and assess its potential resectability.

Findings and procedure details

Primary sclerosing cholangitis (PSC)

PSC is most likely an immune mediated disease characterized by diffuse inflammation with progressive fibrosis and multifocal strictures of the intra- and extra-hepatic bile ducts, eventually developing into cirrhosis, portal hypertension and liver failure in the majority of patients. Small duct PSC is a disease variant which is characterized by typical cholestatic and histological features of PSC but normal bile ducts on cholangiography (1, 4).

A diagnosis of PSC is made in patients with a cholestatic biochemical profile, when cholangiographic modalities as magnetic resonance cholangiography [MRCP] and endoscopic retrograde cholangiography [ERCP] show the characteristic multifocal biliary strictures alternating with segments of focal dilatations, providing that the secondary causes of sclerosing cholangitis have been excluded (2). Patients, who present with cholestatic liver profile and histopathological results (liver biopsy) compatible with PSC yet have a normal cholangiogram, are classified as small duct PSC (3). Approximately
15% of PSC cases are small duct PSCs and 22% of them may progress to large duct PSC within 8 years, thus, cholangiography should be performed in cases of clinical deterioration of small duct PSC patients (5).

IBD is encountered in 60-80% of PSC patients. Most of these patients (80%) have UC, Crohn's disease (CD) has been reported in up to 13% of PSC patients, IBD may be present at the onset of PSC, preceded it by years or it may even be asymptomatic. Therefore, it is highly recommended that PSC patients undergo colonoscopy and biopsy. IBD patients can develop PSC even years after having undergone total colectomy (5).

The rate of progression of PSC is variable; approximately 49% of symptomatic patients develop biliary cirrhosis and liver failure (6). Medical therapy consists of oral agents such as ursodeoxycholic acid. Endoscopic or percutaneous mechanical dilation of dominant benign strictures can be performed for relief of symptoms. Orthotopic liver transplantation is the only curative therapy for PSC (7).

Cholangiography (MRCP/ERCP) represents the primary imaging modality to evaluate for PSC; however, MRCP is preferred as the initial imaging investigation as it is non-invasive with no radiation exposure. The main characteristic findings are biliary irregularities with strictures, dilatations and peripheral pruning of bile ducts. In 5% of cases, isolated involvement of extra-hepatic bile ducts can be seen (8).

MRCP/ERCP findings in the early stage of the disease include short multifocal intrahepatic biliary strictures alternating with normal or mildly dilated segments giving the characteristic "beaded" appearance of the biliary tree. There is a predilection of the strictures to involve the bifurcation areas. As the inflammatory strictures heal with progressive fibrosis, the peripheral ducts become obliterated and non-visualized giving the "pruning" appearance. Diverticula and webs can also occur in PSC although these findings are not pathognomonic. Pigmented stones can be observed in 30% secondary to biliary obstruction and stasis (7). **Fig. 1 on page 13**

When biliary cirrhosis develops, the diagnostic accuracy of MRCP decreases for the evaluation of strictures due to surrounding parenchymal distortion. In addition, MRCP can be limited in differentiating benign from malignant strictures in PSC; furthermore, some cases of secondary sclerosing cholangitis as well as Caroli's disease can be difficult to distinguish from PSC (9).
Fig. 11: Diagnostic algorithm for PSC.


Secondary sclerosing cholangitis

1 - Sclerosing cholangitis in critically ill patient (SC-CIP)
SC in critically ill patients (SC-CIP) is a recently described entity, presumed to result from an ischemic injury secondary to hypovolemic or septic shock. The ischemic insult can also develop in ICU patients receiving positive pressure ventilation for Respiratory distress syndrome with consequent vasoconstriction. The end result is formation of biliary casts with superimposed bacterial and fungal infection, indicative of a very poor prognosis; sepsis is the most frequent cause of death in these patients (10).

MRCP/ERCP are the most useful tools to identify these disease processes, however, the endoscopic removal of biliary casts is of limited efficacy and the only effective treatment option is liver transplantation (6).

2 - Ischemic cholangiopathy

The term ischemic cholangiopathy refers to the ischemic biliary injury with resultant inflammation, ulcers, sludge, and strictures. Ischemic cholangiopathy occurs most commonly in the setting of liver transplant; however, it can be encountered secondary to other causes resulting in alteration of the vascular supply of the biliary tree.

Post-liver transplant ischemic cholangiopathy:

Ischemic cholangiopathy in liver transplant patients mostly occurs within the first 6 months. Ultrasound can detect biliary ductal dilatation, and wall thickening, and possibly echogenic biliary casts secondary to sloughing. Doppler examination of the hepatic artery is essential to identify anastomotic stenosis or occlusion of the hepatic artery (decreased resistive index (<0.5). CT angiography can accurately detect the stenosis at the anastomotic site; MRI/MRCP can identify the biliary strictures and dilatation as well as the biliary casts which appear as filling defects. Transplant liver infarcts can be detected early by CT or MRI.

The term "ischemic cholangitis" refers to the common association of ischemic biliary tract damage with persistent ulcers, inflammation, sludge, and strictures. Apart from the complication of anastomotic stricture, two other types of strictures have been described: 1) Non-anastomotic strictures (NAS) mainly present with diffuse ischemic necrosis of the biliary tree within the first year post-liver transplantation and 2) ischemic type biliary lesions (ITBL) occur later in the setting of a patent hepatic artery (11).

Both types can involve the extra-hepatic ducts, the intrahepatic ducts (including the hepatic confluence), or both. Involvement of the hepatic duct confluence is typical in these injuries. Irregularities are initially seen in the common hepatic duct followed by extension
peripherally to the right and left hepatic ducts. Overtime, progressive multifocal fibrotic healing and strictures formation will eventually develop \( (12) \).

The most common factors resulting in NAS and ITBL are causes resulting in microvascular injury to the liver transplant, including prolonged preservation times of the graft or chronic rejection. Pre-transplantation PSC increases the risk of these injuries as well as the risk of post-transplant recurrence of the disease \( (11) \).

The intrahepatic biliary strictures appear as multifocal tracts of decreased signal alternating with segments of dilation. Findings highly suggestive of an injury that is ischemic in nature are the presence of multiple intraluminal filling defects due to intraductal debris and casts due to sloughed mucosa, as well as intra and extra-hepatic bilomas secondary to bile leakage \( (13) \). Fig. 2 on page 13

Acute rejection of the transplant results in edema of the liver parenchyma with compression and narrowing and possibly obstruction of the bile ducts, whereas chronic rejection presents with progressive obliteration of the ductal system (vanishing bile ducts). The other differential diagnosis for a diffuse pathological process involving the biliary tree post-liver transplant includes recurrence of PSC and diffuse cholangitis. Imaging features on MRCP might be similar with difficult distinction, however clinical and laboratory correlation and sometimes liver biopsy are valuable to differentiate these two entities \( (14) \).

Chemotherapy induced ischemic cholangiopathy:

Floxuridine (an antimetabolite used in treatment of metastatic adenocarcinoma) is a known cause for ischemic cholangitis, up to 56% of cases has been reported. The pathogenesis is induced vasculitis and vascular thrombosis. Formaldehyde for treatment of hydatid cyst has been also reported as a cause as well as the 5-Fluorouracil \( (15) \).

Other causes of ischemic cholangiopathy:

Systemic vasculitis such as polyarteritis nodosa and giant cell arteritis as well as radiation injury and sickle cell disease are likely aetiologies for ischemic cholangitis with few reported cases \( (2) \). Fig. 3 on page 14

**3- Obstructive cholestasis**

Long stranding obstruction of the biliary tree by any form of obstructive lesions including: stones (choledocholithiasis), gall bladder stones (cystic duct stones with Mirrizi syndrome), neoplastic masses (HCC, cholangiocarcinoma, pancreatic masses, metastasis) and biliary strictures (iatrogenic, post-traumatic), all can result
in development of inflammatory strictures, biliary stasis and secondary sclerosing cholangitis, the development of secondary sclerosing cholangitis in this clinical setting depends mainly on the degree of obstruction and its duration. The pathogenesis involves bile regurgitation from the obstructed duct into the perisunsoidal and periportal spaces with associated inflammation and reactive periportal fibroplastic changes. The relief of biliary obstruction early on the disease process can result in reversal of parenchymal changes, however, if fibrosis and biliary cirrhosis develops, the damage is irreversible (16).

Ultrasound is still the primary imaging modality to identify stones within the biliary tree, appearing as echogenic foci with or without posterior shadowing. However, US limitations include incomplete visualization of the CBD distally and the patient body habitus which may limit adequate evaluation. CT detection of biliary stones depends mainly on their composition and calcium content; MDCT has a moderate sensitivity and high specificity for detection of biliary stones (72-77% and 96% respectively); However, it can readily identify secondary signs of obstruction such as proximal dilation of bile ducts, abrupt termination of the CBD, intra-or extra-hepatic bile collections. In addition, MDCT has the highest capability to identify pneumobilia (17).

MRCP is the best modality to identify choledocholithiasis, appearing as filling defects with low signal surrounded by the high signal of bile (sensitivity and specificity of 91-98% and 88% respectively); However, other causes of filling defects on MRCP may mimic bile duct stones, including air bubbles and blood clots or flow voids from vessels, careful evaluation of 3D MRCP images and axial T2W images is mandatory for differentiation. Additional findings related to long standing choledecholithiasis are well evaluated with MRI. These include the development of biliary strictures and dilatation; biliary casts (can appear as high signal structures in T1WIs) as well as secondary biliary cirrhosis with parenchymal atrophy predominantly involving the lateral segment of the left liver lobe (18).

4- Eosinophilic and Mast cell cholangitis

Eosinophilic cholangitis is a rare benign entity that may lead to biliary obstruction. It is relatively non-specific; Infiltration of the biliary tree by eosinophils can also be related to parasitic, fungal, or drug-induced diseases, also co-existing with primary biliary cirrhosis and hepatic transplant rejection (19). Recently, mast cells were implicated in the progression of hepatic fibrosis in patients with PSC. Cholangitis, although rare, of systemic mastocystosis with ERCP features mimicking biliary duct involvement by PSC. Brush cytology of the bile ducts can reveal the extensive infiltration by mast cells (2).

5 - AIDS cholangiopathy
AIDS cholangiopathy can potentially occur in HIV patients when the CD4 count is less than 100/mm³. The main cause for the inflammatory process is opportunistic pathogens, most commonly Cryptosporidium, followed by CMV and microsporidium. There is a tendency of these organisms to involve the Ampulla of Vater resulting in biliary stenosis together with involvement of the intra and extra-hepatic biliary tree resulting in multiple strictures. Fig. 4 on page 15

Ultrasound is the initial imaging modality in HIV patients with suspected cholangiopathy. It may reveal bile duct dilatation and thickening. MRCP/ERCP can demonstrate the characteristic ampullary stenosis and long extra-hepatic bile duct strictures. It can also demonstrate a PSC-like pattern of involvement in terms of multifocal intra and extrahepatic strictures. Acalculic cholecystitis can be seen in these patients as it can be caused by the same pathogens (20).

**6 - Recurrent pyogenic cholangitis**

Formerly known as oriental cholangiohepatitis, RPC is a disease occurring almost exclusively in patients from Southeast Asia. Parasitic infestation by liver flukes (Clonorchis sinensis and Ascaris Lumbricoides) with or without gram negative bacterial infection are the main pathogens. The pathogenesis of the disease is injury of the epithelial lining of the bile ducts and stones formation with associated suppurative inflammation, strictures and abscesses formation. The infection usually presents in middle aged patients with acute recurrent bouts of cholangitis. MRCP/ERCP findings include multifocal areas of dilatation with or without intraductal stones, bile ducts wall enhancement, periductal edema, focal bile duct strictures as well as cholangiectasis (bile lakes) and intrahepatic abscesses. Eighty percent of these cases are complicated by the development of pigmented stones (7&21).

The disease can result in long standing biliary strictures resulting in segmental liver atrophy. MRI/MRCP is most sensitive for initial evaluation and follow-up of the disease as well as identifying complicating cholangiocarcinoma. Treatment varies according the disease presentation and includes biliary drainage, stone retrieval, and segmentectomy of involved liver segment (17). Fig. 5 on page 16

**7- Portal biliopathy**

The formation of large venous collaterals at the region of the porta-hepatis in patient with cirrhosis and portal hypertension can result in common bile duct obstruction by extrinsic compression. This can occur following chronic thrombosis of the portal vein with cavernous transformation and consequent dilatation of the paracholedochal veins that protrude into the lumen of the bile ducts causing secondary ischemic bile duct injury.
The term portal biliopathy is a collective term describing biliary tree injury secondary to portal hypertension. Ultrasound with Doppler can reliably identify the portal cavernoma and the dilated venous collaterals appearing as vascular anechoic tortuous channels. MRCP as well as MRI with intravenous contrast can demonstrate the extent of biliary tree involvement, which commonly affects the CBD and CHD at the porta-hepatis. Bile duct wall irregularities, saccular dilatations and filling defects corresponding to the dilated vessels compressing the ducts are also well demonstrated with MRI. Choledocholithiasis is observed in 17% of these patients. In cases of advanced cirrhosis, peripheral pruning of bile ducts can be also observed (22,23). Fig. 6 on page 17

One study using MR cholangiography combined with MR portography in patients with portal cavernoma revealed stenosis of the common bile duct and/or hepatic ducts from a mass effect of the cavernoma in 21 out of 25 patients (24).

8 - Autoimmune IgG4 Cholagniopathy

IgG4 related sclerosing disease involves a spectrum of immune-mediated abnormalities characterized by infiltration of multiple organs by IgG4 plasma cells, in addition to the IgG4 induced cholangiopathy, simultaneous autoimmune pancreatitis, retropertineal fibrosis and multifocal renal disease can be also seen. The diagnosis depends on the constellation of imaging, laboratory (raised levels of IgG4 levels in serum) and pathological findings. The typical involvement of the biliary tree by the disease includes relatively long segments of biliary strictures, thickening and enhancement (25).

This pattern of involvement of the biliary tree helps to differentiate IgG4 disease from PSC which is mandatory for appropriate clinical management. Long biliary strictures with post-stenotic dilatation are commonly seen in IgG4-related cholangiopathy, whereas the PSC pattern of short strictures with beaded appearance of the bile ducts are not generally observed (26). In a study using MRCP as the primary imaging modality, it was suggested that CBD wall thickening > 2.5mm, non-interrupted continuous strictures and absence of liver parenchymal changes are useful findings to differentiate IgG4 related disease from PSC. In addition, involvement of other organs such as the pancreas and kidneys can be seen in IgG4 sclerosing disease (27). Fig. 7 on page 18

In a recent study including 18 patients (9 IgG4 cholangiopathy, and 9 PSC) it was found that MRCP was useful to differentiate between the two disease processes, as the length of stricture was longer in IgG4-SC (>10mm) while in PSC it was shorter (<5mm). There was a more defined clear margin of the involved bile ducts in PSC when compared to IgG4 cholangiopathy (28).

9 - Post-traumatic cholangiopathy
Post-traumatic cholangiopathy is a relatively rare complication following major traumatic incident. It can result in ischemic injury of the biliary ducts with consequent stasis, inflammation and strictures formation. Most of the reported cases were in long term ICU patient following burns, electrical injuries and major accidents with hypovolemic status. The cholangiographic findings revealed a PSC-like pattern of diffuse biliary tree involvement (29). Fig. 8 on page 19

Iatrogenic strictures are the most common benign biliary strictures, and most commonly affect the CHD or proximal CBD. They can occur secondary to surgery or endoscopy. MRCP/ERCP studies are useful to differentiate iatrogenic strictures which are more regular and show gradual tapering of the involved bile ducts when compared to malignant strictures (14).

An effective way to summarize the causes of secondary sclerosing cholangitis according the underlying etiology are divided into four categories (16); Table 1:

<table>
<thead>
<tr>
<th>Underlying aetiology</th>
<th>Causes</th>
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<tbody>
<tr>
<td>Chronic Obstruction</td>
<td>Biliary stones</td>
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<td></td>
<td>Biliary strictures (Iatrogenic or secondary to chronic pancreatitis)</td>
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<td></td>
<td>Anastomotic strictures in liver transplant</td>
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<td></td>
<td>Neoplasms</td>
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<td></td>
<td>Portal biliopathy</td>
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<tr>
<td>Infectious</td>
<td>Recurrent pyogenic cholangitis</td>
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<td></td>
<td>HIV related pathogens (cryptosporidiosis - Microsporidiosis - Cytomegalovirus)</td>
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<tr>
<td>Immunologic</td>
<td>IgG4 related cholangitis</td>
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<tr>
<td></td>
<td>Eosinophilic cholangitis</td>
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<tr>
<td></td>
<td>Mast-cell cholangiopathy (very rare)</td>
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<tr>
<td>Ischemic cholangiopathy</td>
<td>Post-liver transplant hepatic artery thrombosis</td>
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<td></td>
<td>Chemotherapy infusion biliary injury</td>
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<td>Trans-catheter arterial embolization</td>
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<td></td>
<td>Vasculitis</td>
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<td>Radiation injury</td>
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<tr>
<td></td>
<td>Portal biliopathy</td>
</tr>
<tr>
<td></td>
<td>Critically-ill patients (SCI-CIP)</td>
</tr>
<tr>
<td></td>
<td>Major trauma, burns, electrical shock</td>
</tr>
</tbody>
</table>

Risk for cholangiocarcinoma
The risk of Cholangiocarcinoma in patients with PSC is approximately 5-15%. It is crucial to diagnose cholangiocarcinoma in patients with sclerosing cholangitis prior to liver transplantation because they tend to have a poor prognosis post-transplantation. The diagnosis of cholangiocarcinoma by MRI/MRCP can be difficult because of the co-existent benign biliary strictures in sclerosing cholangitis. The diagnosis is usually achieved by a combination of clinical, laboratory and imaging features. Carbohydrate antigen 19-9 (CA 19-9) is the most sensitive and specific tumor marker (78.6% and 98.5% respectively) with a cut-off value of 129 U/mL (30).

A dominant biliary stricture can develop in 45-58% of PSC patients during follow-up MRCP studies, which raises the suspicion for Cholangiocarcinoma. A dominant stricture is defined as stenosis with a diameter of less of 1.5mm in the CBD or less than 1mm in the CHD. Benign strictures are far more common than malignant strictures (31).

In patients with dominant stricture suspicious for cholangiocarcinoma by MRCP and elevated CA 19-9 > 129 U/mL in serum analysis, ERCP and biliary brushings should be performed with FISH (Fluorescence in situ hybridization) for cytogenetic analysis of the obtained specimens. The initial MRI should be performed with contrast-enhanced sequences to evaluate for possible masses and to determine its relation to the vessels. If the MRI/MRCP is negative for masses or dominant strictures and CA 19-9 value is more than 129 U/mL, ERCP with biliary brushing should be performed, if the cytology is negative, the stricture is mostly of benign nature. If the MRI scan is negative and the cytology results are still suspicious for cholangiocarcinoma, serial CA 19-9 levels as well as the MRI and biliary brushing should be repeated every 6 months. PET-CT scan can be performed if there is a high clinical suspicion for Cholangiocarcinoma. Finding, however, can be non-specific in the presence of inflammatory changes, and should be correlated with the patient’s clinical status (31).

Cholangiocarcinoma can be detected in various morphological forms; the mass-forming type, the periductal infiltrative form and the intraductal form. The intraductal type of cholangiocarcinoma can present with marked ductectasia with or without papillary mass or can present with focal biliary stricture-like lesion with proximal dilatation. In MRI, The mass forming type of cholangiocarcinoma presents a mass lesion with high signal in T2WIs and low signal in T2WIs. In the post-contrast sequences, the mass typically shows peripheral faint enhancement with gradual increase in enhancement being most avid in the delayed phase. It may be associated with capsular retraction or surrounded by satellite nodules. In C, the mass appears homogenous in the precontrast images and follows the same pattern of post-contrast enhancement as MRI. Dilated regional bile ducts may or may not be visible (32). Fig. 9 on page 20

In both CT and MRI, The intraductal pattern of cholangiocarcinoma can present as diffuse ductal dilatation with papillary or plaque-like masses. However, in the second pattern of
intraductal cholangiocarcinoma, only the diffuse ductectasia can be seen without visible masses. The third type can be seen in imaging as focal or segmental duct dilatation with or without visible intra-ductal lesion. Klatskin tumor preferentially involved the common hepatic duct bifurcation with diffuse infiltrative stricture (32). \textbf{Fig. 10 on page 21}

A laboratory and Radiological algorithm to follow when there is clinical suspicion for cholangiocarcinoma in patients with sclerosing cholangitis is detailed in \textbf{Fig. 12 on page 22}.

\textbf{Fig. 12}: Diagnostic algorithm for Cholangiocarcinoma in patient with primary sclerosing cholangitis.
Images for this section:

**Fig. 1:** Primary sclerosing cholangitis. Coronal thick slab MRCP, 3D MRCP, axial T2WI and post-contrast T1 WI with fat suppression. Classic multifocal intra and extrahepatic biliary strictures alternating with segmental dilatation (orange arrows). A Lengthy stricture in the mid-CBD as well as a short stricture at the CHD (green arrows). Diffuse periductal enhancement involving the left lobe segment II hepatic duct in keeping with active cholangitis (blue arrow).

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Fig. 2: Post-liver transplant ischemic cholangiopathy. US/Doppler for a patient with liver transplant demonstrating low resistive index in the main hepatic artery which suggests antegrade stenosis. Evidence of hyperechoic cast material within the bile ducts (thin arrow). This is highly concerning for ischemic cholangitis. Post-contrast T1W MRI images revealed narrowing of the central intrahepatic biliary duct and CHD with dilatation of right and left peripheral intrahepatic biliary ducts and filling defect within the main hepatic artery corresponding to an intraluminal thrombus (thick arrow).

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**Fig. 3:** Patient with Sickle cell disease. 3D MRCP, Axial T1WI post-contrast and axial T2 WI. Multiple focal strictures in the intrahepatic bile ducts causing mild saccular multifocal dilatations; there is also concentric thickening of the distal CBD (arrow). Infarction of the posterior one-third of the spleen with associated haemorrhage (two arrows).

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Fig. 4: HIV Cholangiopathy. Axial post-contrast T1WI with fat-saturation, coronal T2 WI and Coronal oblique MRCP images. Known patient for HIV. Multiple areas of focal dilatation and stricture with abrupt cut-off of the proximal pancreatic duct and proximal CBD at the level of ampulla of Vater (long arrow). These findings are in keeping with AIDS cholangiopathy. Patchy ill-defined areas of arterial enhancement in the liver with dilated intra-hepatic bide ducts, in keeping with perfusion defects due to underlying cholangiopathy. Mild thickening of the wall of the gallbladder with surrounding oedema (two arrows).

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Fig. 5: Oriental cholangiopathy. Pre and post-contrast CT images, T2W and MRCP images. Marked intra and extrahepatic biliary dilatation with intrahepatic, common hepatic and distal CBD stones appearing as intraluminal hyperdense structures in CT (blue arrows) and low signal filling defects in MRI (orange arrows).

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Fig. 6: Portal Biliopathy. A case of portal hypertension with cavernous transformation of the portal vein. Axial post-contrast T1W, MRCP and coronal reconstructed post-contrast CT images. Multiple collateral vessels at the porta-hepatis are encircling and compressing the mid and distal CBD with proximal mild dilatation of the central intrahepatic bile ducts (orange arrows). There are small extrinsic indentations upon the dilated CBD in the MRCP images caused by the collateral vessels (blue arrows).

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**Fig. 7:** Pathologically proven IgG4 Cholangiopathy. Contrast enhanced CT study revealing bilobar intra-hepatic bile ducts dilatation together with non-dilated thickened wall CBD. The pancreas is thickened with peripancreatic pseudocyst secondary to autoimmune pancreatitis. Bilateral hypodense multiple renal parenchymal lesions.

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Fig. 8: Post-traumatic cholangiopathy. Axial T1W post-contrast, post-contrast T1W subtraction images, ERCP and MRCP images. Patient with history of major abdominal trauma and liver laceration. Segmental intrahepatic biliary dilatation and wall thickening secondary to liver laceration bile duct injury in segment VIII (green arrows). Clear communication between a perihepatic collection (Biloma) and the bile duct (blue arrows). The patient developed afterwards diffuse cholangitis with Irregularities identified in the biliary tree (black arrows) as well as a stricture in the CBD (orange arrows).

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Fig. 9: Primary sclerosing cholangitis complicated by cholangiocarcinoma. 3D MRCP, Axial T2W, Axial T1W LAVA pre-contrast and delayed post-contrast T1 WI images. Beaded appearance of the biliary tree as a result of alternating multifocal strictures and dilatation (blue arrows). Soft tissue tumour within the common hepatic duct proximal to the cystic duct, extends into the proximal intrahepatic bile ducts, mainly on the left side with avid delayed post-contrast enhancement, compatible with cholangiocarcinoma (orange arrows).

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**Fig. 10:** Klatskin tumor. Axial T2W, Post-contrast axial T1W and MRCP images. Tumour mass predominantly in the left common hepatic duct with extension into the right hepatic, common hepatic, and proximal common bile duct. The mass is causing obstruction with bilateral intrahepatic biliary duct dilatation.

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Fig. 12: Diagnostic algorithm for Cholangiocarcinoma in patient with primary sclerosing cholangitis.

Conclusion

The pattern of biliary tree involvement on MRI/MRCP is a helpful differentiating feature between primary and secondary sclerosing cholangitis in correlation with other imaging features, as well as the clinical and laboratory findings. It is important for the radiologist to be familiar with the various imaging manifestations of each disease entity, as well as the early features of a complicating cholangiocarcinoma, in order to improve the long term prognosis of the patient via liver transplantation.

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31. Development of dominant bile duct stenoses in patients with primary sclerosing cholangitis treated with ursodeoxycholic acid: outcome after
