Case 8710

Portal biliopathy
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Patient: 46 years, male

Clinical History:
This is a case of portal biliopathy caused by idiopathic cavernoma in a 46 year old men. Diagnosis of common bile duct compression by the cavernoma was made first by CT scan, then by MRI, and finally managed by endoscopic biliary plastic stenting.

Imaging Findings:
A 46 year old man with a history of idiopathic portal cavernoma (PC) presented with right upper quadrant abdominal pain, jaundice, pruritus, and yellow urine. Physical exam noted hepatomegaly without fever. Laboratory values showed a cholestatic pattern and increased liver enzymes. US showed multiple anechoic tubular structures around the portal vein corresponding to a cavernoma and dilated intrahepatic bile ducts. CT scan and axial MRI confirmed that the common bile duct (CBD) abnormalities resulted from imprinting on the CBD lumen by the veins composing the cavernoma (Fig. 1, 2) and MR cholangiography (MRC) demonstrated well a stricture just underneath the biliary convergence (Fig. 3). Endoscopic retrograde cholangiopancreatography (ERCP) showed grade I esophageal varices and stricture of the CBD at the same level (Fig. 4). Sphincterectomy, balloon dilatation, and plastic stenting were made. Cholestasis and jaundice resolved 6 months after the treatment.

Discussion:
Portal biliopathy (PB) is a recent terminology used to describe changes in the bile duct and gall bladder due to cavernous transformation in patients with portal hypertension [1,2]. The mechanisms of biliary abnormalities in extrahepatic portal vein obstruction are either extrinsic compression by collaterals or peribiliary fibrosis resulting from ischemic or inflammatory changes underlying portal thrombosis [3].

The most common conventional cholangiographic abnormalities of PC include irregular strictures of the extra- and intrahepatic bile ducts, dilated segments with a beaded appearance, ectasia, and pruning of the intrahepatic bile ducts mimicking cholangiocarcinoma or sclerosing cholangitis [4].
Colour Doppler US is a useful first imaging modality, demonstrating also the dilatation of the bile ducts in most instances. MRI shows simultaneously the PB and the bile ducts and may prove useful in both adults and in children [4]. In our case these abnormalities were visualized first by CT scan. Because ERCP is an invasive procedure and US may not give a definite outline of the biliary ductal anomalies, MRC has been found to be helpful for the diagnosis of several biliary tract abnormalities [2]. It can be useful in showing the sites of stenoses and guiding the therapeutic interventions. Grading and progress of the stenosis can also be monitored, helping early treatment of the cholestasis [2].
MRC offers the advantages of being noninvasive and providing all the diagnostic components of PB, including extrahepatic portal vein obstruction, gallbladder varices, bile duct abnormalities, and delineating the extent and severity of bile duct strictures [2,4].
The short- and long-term complications of cholestasis, including biliary cirrhosis, require treatment of these biliary abnormalities as soon as they become clinically significant. Bile duct stricture can be managed by endoscopic
procedures such as sphincterectomy, balloon dilatation, and stenting. Direct surgical approach of the bile duct is not indicated as it carries a risk of severe bleeding [3]. Endoscopic sclerotherapy or ligation of oesophageal and/or gastric varices is often recommended in children in whom surgical decompression of the portal circulation may not relieve cholestasis. There have been, however, a few adult patients in whom portal shunt surgery has failed to completely cure the biliary problem; such patients had biliary strictures that did not regress after shunt surgery [1,3]. According to our observation and case reports published to date, endoscopic therapy of benign strictures seems to be effective and may be a viable alternative to surgical bypass, particularly in patients who are suspected to have biliary strictures secondary to PC [3,4]. Temporary insertion of a plastic endoprosthesis is sufficient to achieve free biliary flow, even after removal of the stent [3]. Surgical management by hepaticojejunostomy bears the risk of complications as a result of intraabdominal collaterals and should only be performed if stenting is not successful [1,3].

Differential Diagnosis List: Portal biliopathy

Final Diagnosis: Portal biliopathy

References:

Description: CT scan after intravenous injection of contrast media. Axial (a,c) and coronal reformatted images (b) show close relation between the portal cavernoma (black arrow) and the common bile duct (white arrow).

Note calcifications of in the portal venous system, splenomegaly and dilated splenic vein. Origin:
Description: CT scan after intravenous injection of contrast media. Axial (a,c) and coronal reformatted images (b) show close relation between the portal cavernoma (black arrow) and the common bile duct (white arrow).
Note calcifications of in the portal venous system, splenomegaly and dilated splenic vein. Origin:
Description: Note the cystic duct (white arrow) and CBD (arrow head) compression by cavernoma. Biliary bladder containing stones (black arrow). Origin:
**Figure 2**

Description: Endoscopic retrograde cholangiography showing common bile duct stricture (arrows).

Origin:
**Figure 3**

(a) Description: Axial T2 weighted images (a-d) showing intrahepatic bile duct dilatation. 

(b) Description: Axial T2 weighted images (a-d) showing intrahepatic bile duct dilatation.
Description: Axial T2 weighted images (a-d) showing intrahepatic bile duct dilatation. Origin:
Description: Axial T2 weighted images (e,f) showing common bile duct (black arrows) and cystic duct (white arrow) stenosis caused by pressure from the cavernoma. The common bile duct lumen is still visible. Origin:
Description: Coronal MR cholangiography showing intrahepatic bile duct dilatation, common bile duct (big arrow) and cystic duct (small arrow) stenosis. Origin: