

Step1:

Answer:

c) Biliary lake

Step2:

▪ Answer:

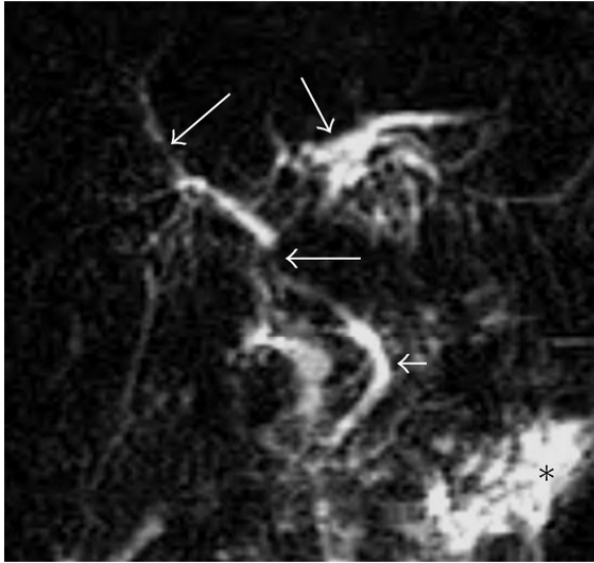
b) Hereditary hemorrhagic telangiectasia

▪ Findings:

- Abdominal CT scan in arterial phase shows :Heterogeneous liver parenchymal enhancement, diffuse small hyper vascular lesions, hepatic vein filling with contrast on arterial phase, enlarged hepatic artery branches and hepatic veins, large cystic mass in the left lobe and smaller one in the right lobe without enhancement.
- Findings are in favor of arteriosystemic shunting and telangiectasis due to Hereditary Hemorrhagic telangiectasia syndrome. Cystic masses are biliary cyst lakes due to ischemic cholangitis in this condition.

▪ Discussion:

- Hereditary hemorrhagic telangiectasia (HHT) or Osler-Weber-Rendu syndrome is an autosomal dominant vascular disorder characterized by spontaneous epistaxis, telangiectasia, and visceral vascular malformations.
- Hepatic vascular malformations are common, though a minority are symptomatic.
- Liver telangiectasis are early manifestations of hepatic involvement in HHT: sub centimeter hyper vascular focal lesions resembling an asterisk.
- Three types of intrahepatic shunts between the major vessels of the liver are possible: arteriosystemic (hepatic artery to hepatic vein), arterioportal (hepatic artery to portal vein), and portosystemic venous (portal vein to hepatic or systemic veins, rare) .
- Arterial-systemic shunting between the hepatic artery and hepatic vein contributes to HOCF, a major cause of morbidity and mortality for HHT patients. Arterial-portal shunting can lead to portal hypertension. Rarely hepatic encephalopathy results from portosystemic venous shunting.
- Liver-specific abnormalities include pseudocirrhosis, FNH, and ischemic cholangiopathy.
- The vascular supply of biliary ducts depends on the hepatic artery branches so shunts by stealing the arterial flow may cause ischemic cholangitis. The biliary disease subtype of liver involvement by HHT is characterized by biliary strictures and dilation and bile cysts.



Ischemic cholangitis in a patient with HHT: MRCP sequence shows contour irregularities multiple focal strictures and dilatation of the biliary tract (arrows) with a “pruned tree” appearance. Contour irregularity of the common bile duct wall is seen (arrowhead).

Mehmet Bilgin et al. “CT and MRI Findings of Hepatic Involvement in Rendu-Osler-Weber Disease”, Case Reports in Radiology, vol. 2012, Article ID 484085, 2012.

▪ **References:**

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“Case courtesy of Faeze Salahshour, MD, Associate Professor of Radiology, Imam Khomeini Hospital, Tehran University of Medical Sciences.”