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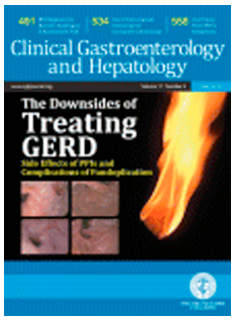

Upper Gastrointestinal Bleeding Due to Hereditary Hemorrhagic Telangectasia

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Title

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Abbreviations

AVMs - arteriovenous malformations; HHT - Hereditary hemorrhagic telangiectasia.

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Key words

Hereditary hemorrhagic telangiectasia; hepatic arteriovenous malformations; Osler - Weber -Rendu disease

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A 50 year old Caucasian woman presented with hematemesis. She had past medical history of asthma. There was no family history of coagulopathies. Physical examination revealed pale conjunctiva with a single oral telangiectasia. Her laboratory analysis showed hemoglobin 103 g/dL, platelet count 97 x 10^9/L, serum albumin 43 g/L, bilirubin 15 µmol/L, alkaline phosphatase 147 U/L, aspartate aminotransferase 28 U/L, gamma-glutamyl transferase 70 U/L, alpha-fetoprotein < 3 ng/mL and negative liver screen. The oesophagogastroduodenoscopy revealed four columns of oesophageal varices with stigmata of recent bleeding. She was treated with band ligation.

CT imaging demonstrated heterogeneous enhancement of the liver (Figures A, B) with multiple arteriovenous malformations (Figure A), portal venous malformations (Figure B) and established portal hypertension with splenomegaly up to 15 cm. There was no biliary duct dilatation. The portal vein and hepatic veins were patent and there was no evidence of cirrhosis. This was in keeping with portal hypertension secondary to Hereditary Hemorrhagic Telangiectasia (HHT).

HHT, also known as Osler-Weber-Rendu disease, is an autosomal dominant vascular disease characterised by mucocutaneous or visceral angiodysplastic lesions (telangiectases and AVMs) affecting the skin, brain, lungs and gastrointestinal tract [1]. HHT is associated with elevated blood levels and tissue expression of vascular endothelial growth factor (VEGF). HHT is rare (prevalence 1/100,000-2/100,000) although hepatic involvement occurs in up to a third of cases [2]. Given the dual blood supply to the liver, hepatic arteriovenous malformations give rise to three types of shunts: arteriovenous (hepatic artery to hepatic vein), arterioportal (hepatic artery to portal vein), and portal venous (portal vein to hepatic vein) [3]. The most common being arteriovenous shunts [4]. Approximately 5% of patients with hepatic involvement of HHT are symptomatic. Patients often present with signs of congestive cardiac failure, portal hypertension, biliary ischaemia and portosystemic encephalopathy [4].

The definitive diagnosis of hepatic involvement in HHT is confirmed by imaging studies. The hallmark findings are intrahepatic hypervascularisation and an enlarged common hepatic artery (Figure C, white arrow). These abnormalities can be demonstrated by various imaging modalities: angiography, doppler ultrasonography, computer tomography and magnetic resonance imaging [3].

No treatment is recommended for asymptomatic patients. In patients with symptomatic liver involvement, measures to reduce shunting such as arterial embolisation of large AVMs can be hazardous and risk the development of new shunts within the liver [5]. Bevacizumab, a VEGF inhibitor, has been shown to be

helpful in reducing complications of HHT [6]. It was initially found to be very helpful in management of epistaxis in patients with nasal lesions. Treatment with VEGF inhibitors has been associated with reversal of cholestasis, resolution of cardiac failure, ascites and marked reduction in liver vascularity[7]. At present, liver transplantation is the only definitive cure for hepatic involvement in HHT [3].

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