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Case Report

Liver Transplant in Patient with Rendu-Osler-Weber Syndrome

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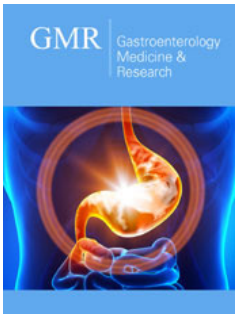

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Introduction

Rendu–Osler–Weber syndrome or "hereditary hemorrhagic telangiectasia", is a rare autosomal dominant disease with a clinical penetrance of 97%. Cutaneous and mucosae telangiectasias are responsible for digestive bleeding and epistaxis. Visceral vascular dilatations perform arterio-venous shunts in different organs. Clinical diagnosis of Rendu-Osler disease is based on the Curacao criteria, but this syndrome must be confirmed genetically (ALK-1, ENG and SMAD 4 genes mutations are found in 90% of patients) and may predict some specific complication such hepatic or cardiac disease. Computed tomography, magnetic resonance imaging and echo-color Doppler are the gold standard for refined research when liver is symptomatic. Rendu-Osler disease in liver can be classified in function of pathophysiologic findings: shunt from the hepatic artery to the hepatic vein (type 1); shunt from the hepatic artery to the portal vein (type 2) or shunt from portal to hepatic vein (type 3. In biopsies, pseudo-cirrhosis is the most frequent histologic finding. Patients can suffer high-output right cardiac failure, ischemic cholangitis and portal hypertension. It depends on the shunt's size. When the liver is the "symptomatic" organ, mainly associated with biliary complications (recurrent cholangitis, biliary necrosis or liver abscesses), or portal hypertension (ascites, encephalopathy, variceal bleeding, etc), the best treatment is liver transplant, although bevacizumab has been used in the treatment of epistaxis, melaena and anemia with encouraging results, showing a reversal of the need for liver transplantation in certain patients [1,2].

Case Report

62-year-old woman diagnosed with hereditary hemorrhagic telangiectasia (Rendu Osler Weber syndrome). The initial study revealed high-flow heart failure, secondary to the presence of multiple intrahepatic arteriovenous fistulas (A and B). The initial management of heart failure was performed with sildenafil and bevacizumab, without clinical response, so it was decided to undergo liver transplantation.

In the preoperative study, the echocardiogram showed moderate dilatation in the atria and right ventricle, dilatation and eccentric hypertrophy of the left ventricle, and a cardiac output of 7.1 l/min. Pulmonary hypertension was not evident. During liver transplantation, a liver with abundant arteriovenous fistulas and a large diameter (12mm) hepatic artery (C) was found, performing arterial revascularization through the patient hepatic artery left branch (D). The postoperative course was without complications and heart failure returned in a few days. In patients with Rendu Osler Weber syndrome, the enlarged hepatic artery diameters can complicate arterial anastomoses in liver transplant procedure, particularly in the presence of small donor arteries. In these conditions, the recipient hepatic artery right or left branch should be considered to be a suitable vessel for the arterial anastomosis in these patients (Figure 1).

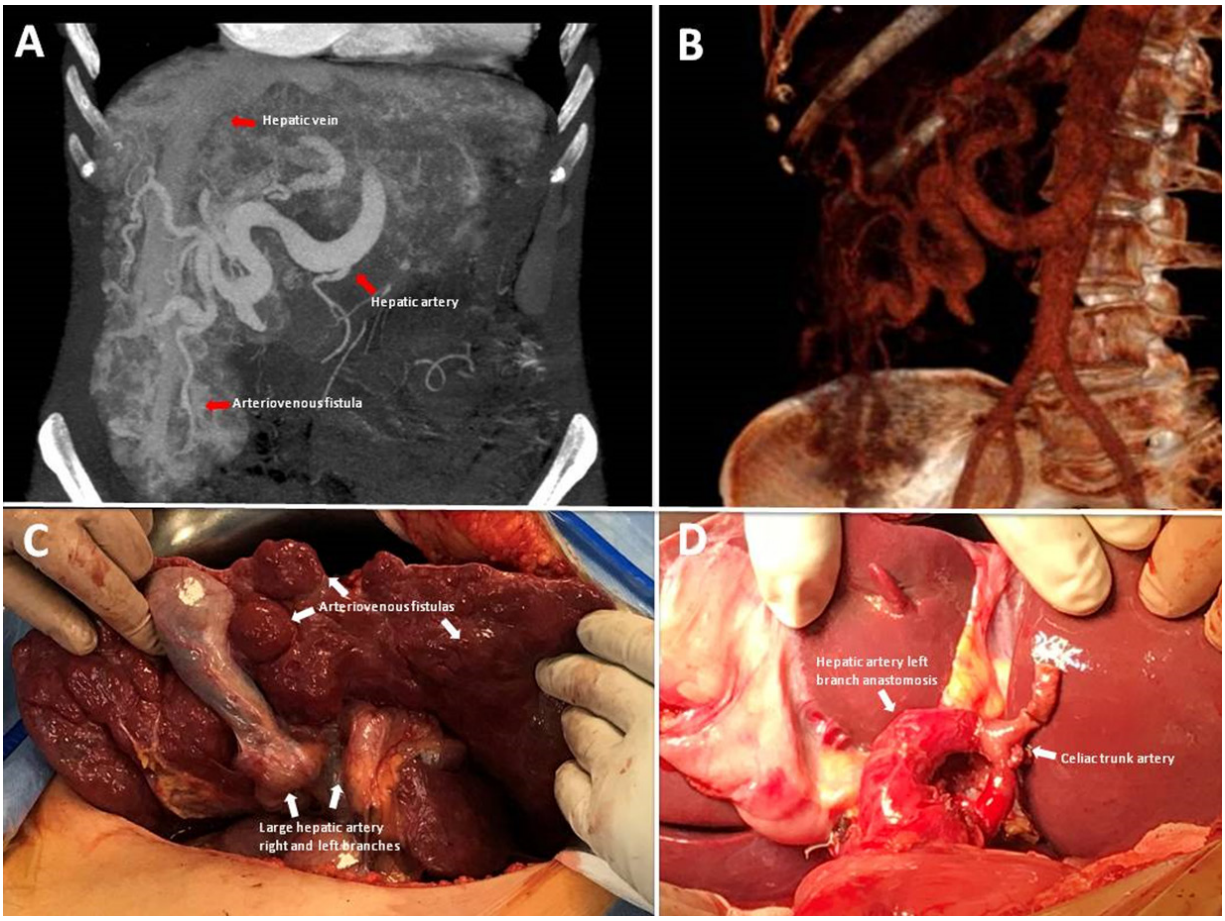

Figure 1

Conflict of Interest

There is no conflict of interest statement for all authors.

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