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Orthotopic Liver Transplantation for Hereditary Hemorrhagic Telangiectasia and Men Type I Syndrome – Case Report and Review of Literature

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Abstract
Introduction: Hereditary haemorrhagic telangiectasia (HHT) is a rare autosomal dominant genetic disorder characterized by arteriovenous malformations (AVMs) mainly affecting the lungs and the liver. In this case AVM's resulted in liver cirrhosis and an indication for orthotopic liver transplantation (OLT).
Case Report: A 59 year-old male patient with HHT who had been previously diagnosed with Multiple Endocrine Neoplasia type 1 Syndrome (MEN 1) was listed for OLT for end-stage liver disease due to hepatic AVMs. During the procedure, a novel type of arterial anastomosis (end-to-side) was chosen because of the mismatch in diameter between the hepatic artery (HA) of the donor and the recipient, respectively. Graft function was normal and repeat Doppler ultrasound studies showed a normally functioning arterial anastomosis. However, the patient died on POD 34 due to an un-related cause (cardiac arrest resulting from myocardial infarction).
Conclusion: To the best of our knowledge this is the first report of an association of HHT and MEN 1. Moreover, this is also the first reported end-to-side arterial anastomosis in an HHT patient during OLT. Our paper shows that the surgical technique we applied is both feasible and safe.
Key words: hereditary haemorrhagic telangiectasia, multiple endocrine neoplasia syndrome type 1, orthotopic liver transplantation