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CASE REPORT

Hereditary Haemorrhagic Telangiectasia — Osler-Weber-Rendu Disease — with Extensive Hepatic Arteriovenous Malformation

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ABSTRACT

Hereditary haemorrhagic telangiectasia is a rare disease. Hepatic involvement is infrequent. A patient with an extensive hepatic arteriovenous malformation is presented. Characteristic computed tomographic and angiographic findings are described followed by a review of the literature.

Key Words: *Angiography, Arteriovenous malformation, Computed tomography, Hereditary haemorrhagic telangiectasia, Telangiectasia*

INTRODUCTION

Osler-Weber-Rendu disease or hereditary haemorrhagic telangiectasia (HHT) is a rare autosomal dominant disorder characterised by mucocutaneous and visceral telangiectasia. The prevalence of HHT is estimated at 1 to 2 cases per 100 000 people.

CASE REPORT

A 75-year-old woman presented with repeated episodes of melaena and epistaxis. She was referred to the Radiology and Organ Imaging Department of the United Christian Hospital for mesenteric angiogram during an admission to hospital for massive upper gastrointestinal (GI) bleeding.

The patient had congestive heart failure on presentation. Laboratory investigations showed that she had iron deficiency anaemia and mildly deranged liver function. Both upper and lower GI endoscopy were performed but the findings were unremarkable.

Mesenteric angiogram with celiac catheterisation was subsequently performed. Angiography showed dilatation of the hepatic artery, multiple areas of focal dilatations, and marked early drainage through the hepatic veins into the inferior vena cava (Figure 1). Focal tuft of dilated vessels representing telangiectasia were also found in the left lower jejunal and right upper colic arterial branches. However, there was no evidence of active gastrointestinal bleeding.

Dynamic contrast enhanced computed tomography (CT) of the abdomen was also performed because of the deranged liver function test and the suspicion of a hepatic space-occupying lesion. CT showed that there was dilatation of the hepatic artery with early filling of the hepatic veins, suggesting the presence of an arteriovenous shunt (Figure 2). Diffuse heterogeneous enhancement of the hepatic parenchyma, in keeping with telangiectases involving both lobes of the liver, were also found.

DISCUSSION

The frequency of hepatic involvement in HHT varies from 8% to 31% in epidemiological recruited and symptomatic patient populations, respectively.[1,2] However, the exact prevalence of hepatic arteriovenous malformation (AVM) is still unknown.[3]

Hepatic involvement may induce high output cardiac failure. The majority of such patients are postmenopausal woman with hyperdynamic circulation resulting from

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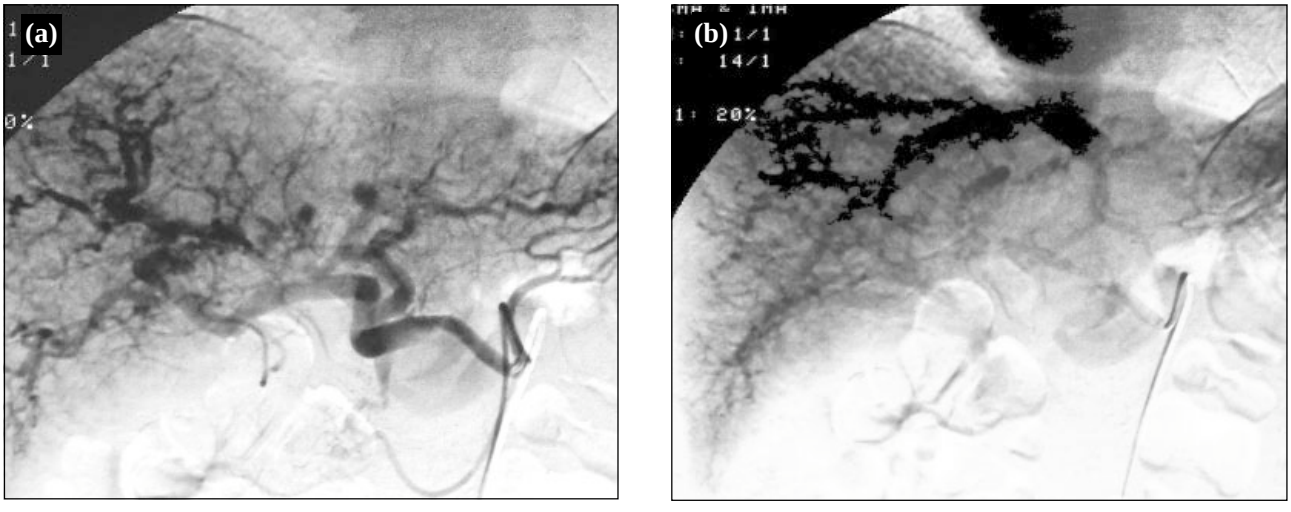

Figure 1. Angiograms of hepatic arteriovenous malformations in hereditary haemorrhagic telangiectasia. (a) Coeliac angiograms obtained during early arterial phase show dilated and tortuous hepatic arteries; (b) early filling of hepatic veins into the inferior vena cava.

arteriovenous shunting, portovenous shunting, or both.[4,5] The patient described in this report probably had this complication. However, if further surgical or embolisation treatment was being considered, right heart catheterisation and haemodynamic studies would be required to confirm and evaluate the degree of shunting.

Characteristic imaging features of this disease are well demonstrated with angiography. Angiography shows the following features:

- the dilatated and tortuous hepatic artery and its branches
- widespread parenchymal blushes of vascular telangiectasia
- early opacification of hepatic veins, signifying arteriovenous shunting.[6]

Contrast enhanced CT scan commonly shows a prominent hepatic artery, which is associated with dilatated hepatic and/or portal veins. Dynamic study may also demonstrate the presence of an arteriovenous or arterioportal shunt. Telangiectases in the hepatic parenchyma could be demonstrated as diffuse heterogeneous enhancement of the hepatic parenchyma.[7] CT readily confirms the diagnosis of hepatic involvement in HHT, and has been reported to show the comparable diagnostic yield with coeliac angiography.[4] However, angiography remains useful for the diagnosis of associated vascular lesions of the mesenteric tree.

The management options for symptomatic hepatic AVM include hepatic artery embolisation, ligation, or liver transplantation. Results from recent reports are

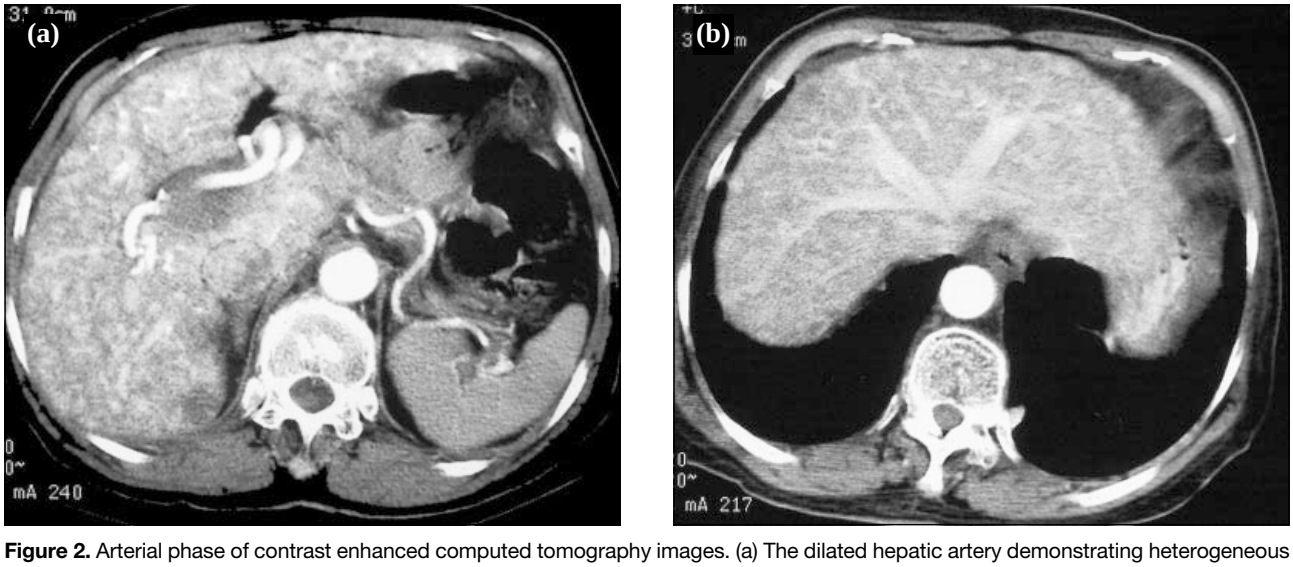

Figure 2. Arterial phase of contrast enhanced computed tomography images. (a) The dilated hepatic artery demonstrating heterogeneous enhancement of the hepatic parenchyma, suggesting diffuse telangiectasis; (b) early filling of the hepatic veins, indicating the presence of an arteriovenous shunt.

controversial.[8-10] Hepatic artery embolisation to alleviate heart failure due to massive shunting through hepatic arteriovenous shunts has been successfully tried. However, significant morbidity or mortality for up to 41% of the patients occurrs.[10] The long term outcomes for patients with successful embolisation or transplantation are still largely unknown.

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