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Clinical Image Open Access

Pulmonary Arterial Hypertension from Hepatic HHT

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Clinical Image

A 42-year female was admitted to our hospital complaining of repeated cough and pant for 2 years. The patient had intermittent nasal bleeding, and her family (including her mother, her aunt) had also similar medical history. Many capillary blood vessels were dilated in her finger and tongue (Figures 1 and 2).

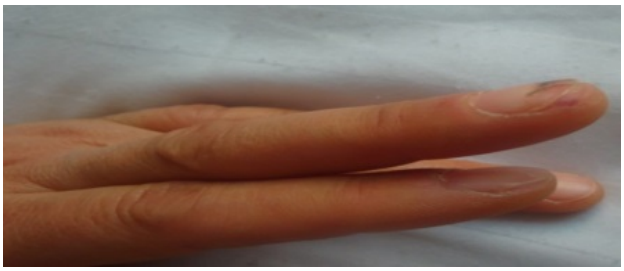

Figure 1: Many dilated capillary blood vessels were noted in her finger

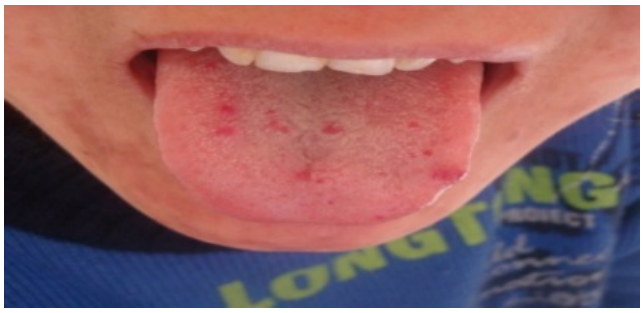

Figure 2: Many dilated capillary blood vessels were noted in her tongue

No heart murmur or rhonchi was audible on the chest auscultation. Hepatosplenomegaly was not noted on abdominal field palpation. Echocardiography suggested right atrium enlargement, right ventricle enlargement, and pulmonary arterial hypertension. Abdominal ultrasound suspected hepatic hemangioma.

Chest radiography showed mild cardiomegaly with increasing pulmonary vascular markings (Figure 3). Contrast-enhanced CT of chest revealed the same cardiac alterations as echocardiography (Figure 4). Hepatic contrast-enhanced CT revealed parenchymal perfusion disorders, arteriosystemic shunts and arterioportal shunts (Figures 5 and 6).

Hereditary Hemorrhagic Telangiectasia (HHT) is mucocutaneous or visceral angiodysplastic lesions, which may be distributed throughout cardiovascular system. Currently, the pathogeneses of the disease could result from genetic mutations that interfere with angiogenesis and its control mechanisms [1].

Hepatic involvement occurs in 30% to 73% of patients with HHT; most of them are symptomatic or have a slight elevation of r-GGT [2]. Hepatic vascular lesions range from tiny telangiectases to transient perfusion abnormalities and large confluent focal vascular masses.

Three different and often concurrent types of intrahepatic shunts are responsible for the parenchymal, vascular and bile duct signs. An arteriovenous shunt can, on rare occasions, be responsible for high-output heart failure as described in our case.

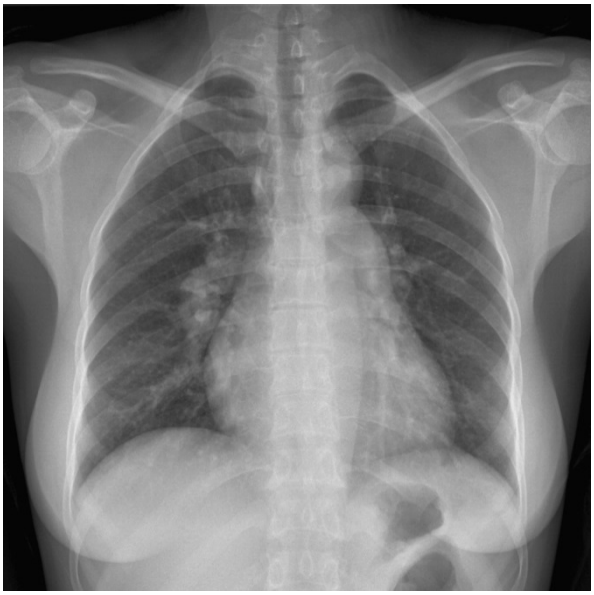

Figure 3: Chest radiography showed mild cardiomegaly with increasing pulmonary vascular markings.

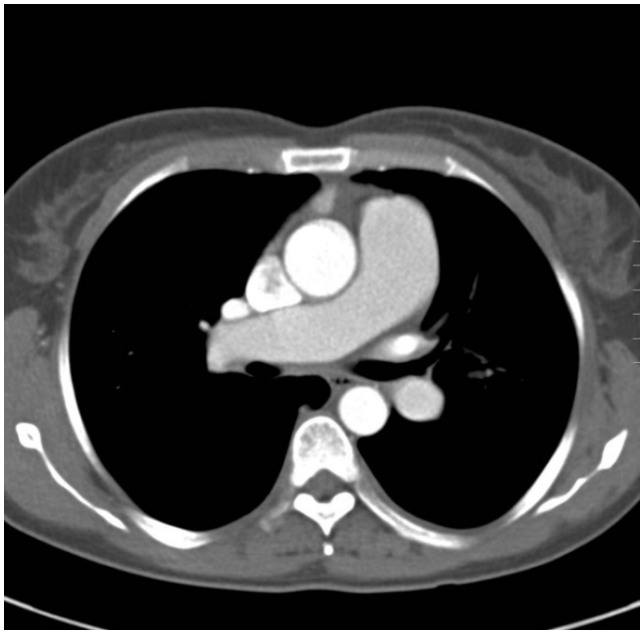

Figure 4: Contrast-enhanced CT of chest revealed pulmonary arterial hypertension.

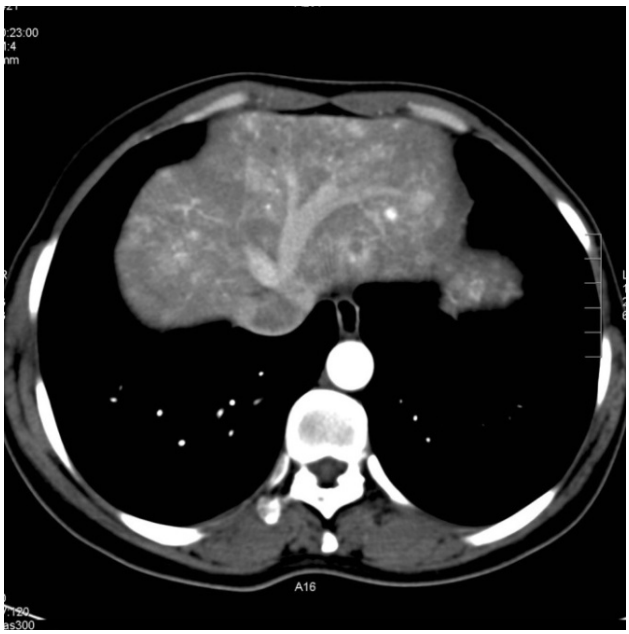

Figure 5: 5-Axial CT in the arterial phase earlier shadow of hepatic veins suggesting of arteriosystemic shunt.

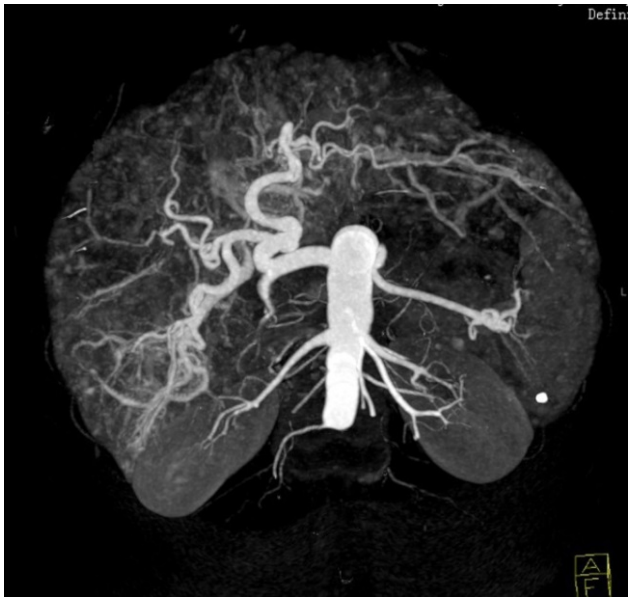

Figure 6: Maximal intensity projection of contrast-enhanced CT scan revealed arterioportal shunts.

References

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2. Buscarini E, Danesino C, Olivieri C (2015) A liver involvement in hereditary haemorrhagic telangiectasia or Rendu-Osler-Weber disease. Dig Liver Dis 37: 635-645.