Hepatopulmonary syndrome with large pulmonary arteriovenous malformations: CT findings with emphasis on its association with a mosaic pattern of the lung parenchyma

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INTRODUCTION

Hepatopulmonary syndrome (HPS) is defined as the triad of liver disease, arterial oxygenation defect, and evidence of intrapulmonary vascular dilatations. The physiopathologic mechanism is not completely understood, but an excessive production of vasodilator substances has been implicated. These vasoactive substances could explain the formation of anomalous communications between pulmonary vessels and pulmonary arteriovenous malformations (PAVM) (1).

CASE REPORT

We present the case of a 54-year-old patient with cirrhosis, progressive dyspnea, and platypnea. Thoracic computed tomography (CT) showed multiple PAVM, confirming the diagnosis of HPS (Fig. 1 A and B). Besides precisely identifying the number and location of PAVM, CT also demonstrated a striking mosaic pattern of the lung parenchyma, characterized by the presence of alternating geographic areas of low attenuation (showing pulmonary vessels with a decreased diameter) with regions of relatively increased attenuation (showing pulmonary vessels with a normal diameter). This mosaic pattern of the lung parenchyma has scarcely been described in patients with HPS since it is not always present and usually requires a post-processing of the CT images in order to increase the contrast between the low attenuation areas (representing hypoperfused regions) and the areas with a relatively increased attenuation (representing better perfused regions) (Fig. 1C). The decision was made to embolize the major PAVM, achieving an improvement of both the oxygen partial pressure and the patient’s symptoms. This improvement allowed the patient to become an acceptable candidate for liver transplantation.

We believe that, unlike other radiological signs of HPS, the mosaic pattern has not been sufficiently described in the scientific literature. If the association of the mosaic pattern on CT with HPS is confirmed in larger studies, it could become a useful sign for detecting hypoperfused pulmonary areas related to small nonvisible PAVM (2,3).

REFERENCES


Fig. 1. A. Axial MIP (maximum intensity projection) CT image shows a large pulmonary arteriovenous malformation (PAVM) (asterisk) in the lingula. B. Coronal MIP CT image shows a smaller PAVM (asterisk) in the right lower lobe. C. Coronal minIP (minimum intensity projection) CT image demonstrates the mosaic pattern of the lung parenchyma, characterized by the presence of alternating low attenuation geographic areas (reflecting hypoperfusion) (arrows) with areas of increased attenuation (reflecting better perfused regions). Note that hypoperfused regions are mainly located in the basal areas of both lungs.