

Arteriovenous Malformations in Hereditary Haemorrhagic Telangiectasia



Malformações Arteriovenosas na Telangiectasia Hemorrágica Hereditária

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Palavras-chave: Malformações Arteriovenosas; Telangiectasia Hemorrágica Hereditária; Tomografia Computorizada.

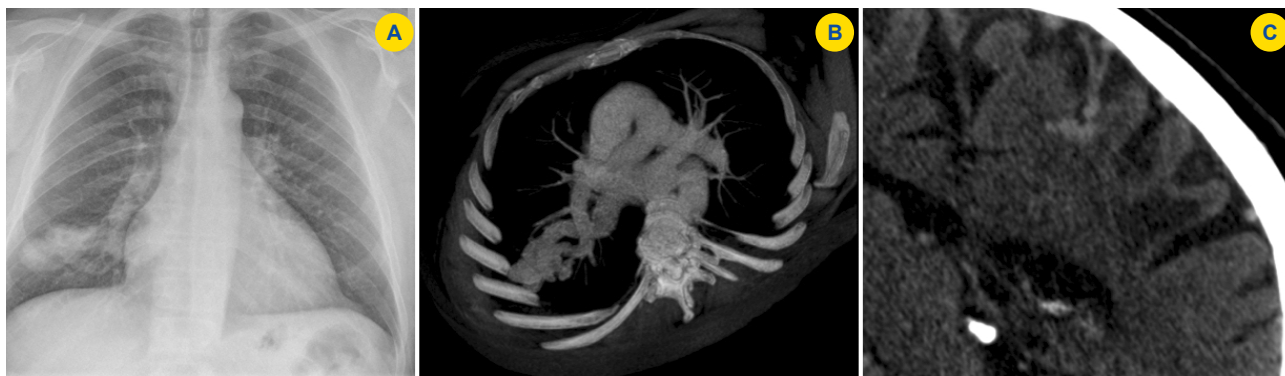


Figure 1 - (A) Rounded homogeneous opacity in the right lower lobe at radiograph frontal view. (B) Inferior view of the chest computed tomography (CT) scans demonstrating the arteriovenous malformations (AVMs) in the right lung. (C) CT study of the head revealing the presence of cerebral AVMs.

A non-smoker 61-year-old man presented with insidious shortness of breath and a history of chronic epistaxis for approximately three decades. The initial investigation revealed right basal crackles at auscultation, very subtle oral telangiectasia and a microcytic anaemia. A chest radiograph (Fig. 1A) demonstrated right basal rounded opacities communicating with the hilum. Further computed tomography scans of the chest (Fig. 1B) and brain (Fig. 1C) have confirmed the presence of multiple arteriovenous malformations (AVMs), suggesting Hereditary Haemorrhagic Telangiectasia (HHT, Osler-Weber-Rendu disease).

HHT is an autosomal dominant inherited disease that affects the vascular endothelium.¹ Patients usually exhibits a positive family history. Due to social issues, this information could not be obtained in the illustrated case. Telangiectasia, AVMs or larger arteriovenous fistulae can either be noticed at physical examination or at imaging assessment.² The developments of pulmonary or cerebral AVMs are known complications in patients with HHT. While endovascular treatment with AVMs embolization is the most adopted intervention, the size and the location of the lesions should determine the best approach.

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