Rupture of pulmonary arteriovenous fistula in the setting of hereditary hemorrhagic teleangiectasia

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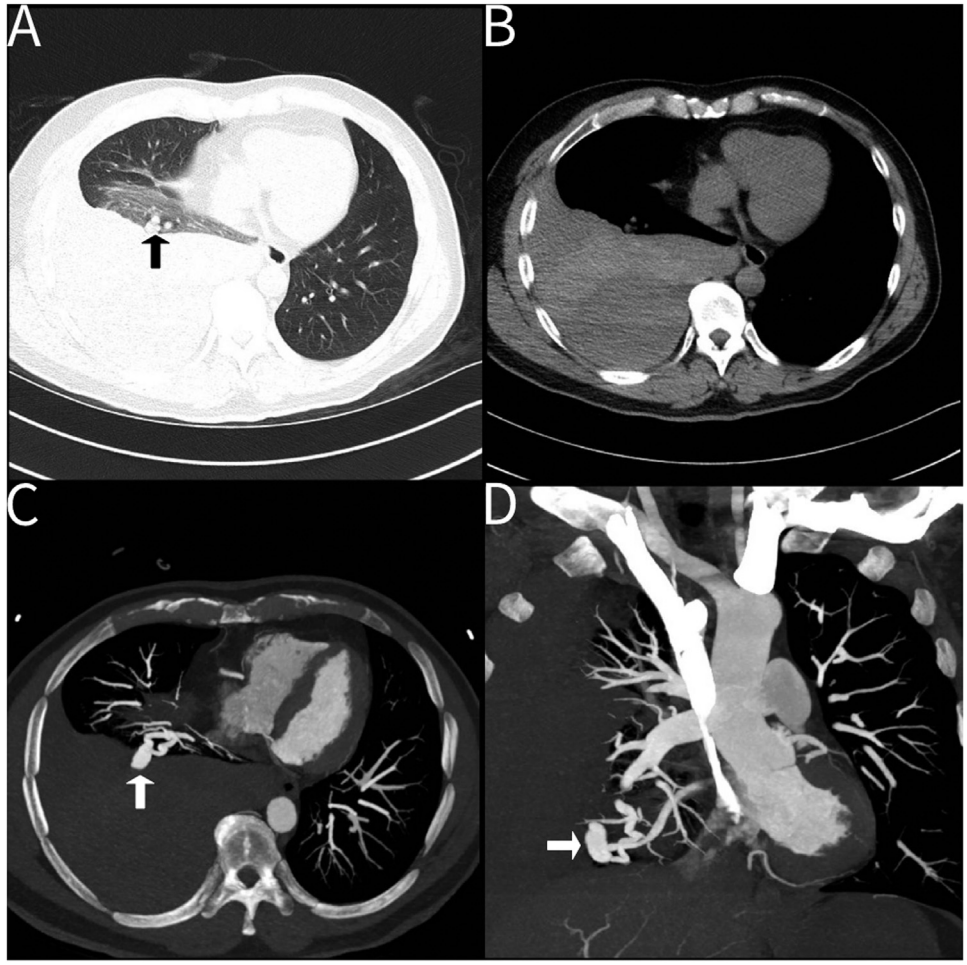

FIG 1.

CASE PRESENTATION

A 49-year-old man presented with a 2-hour history of right-sided chest pain and mental status changes. He reported having spontaneous recurrent nosebleeds, which also occurred in his father. Mucocutaneous telangiectasia was found in his right thigh. His-temperature was 36.0 °C. The blood pressure was 68/43 mm Hg on arrival. Laboratory evaluation revealed the following: white blood cell count, 18.2×10^9/L; red blood cell count, 3.84×10^{12}/L; blood platelet, 180×10^9/L; N%, 95.4%; hemoglobin 116 g/L. C-reactive protein was normal (0.64 mg/L). Tumor markers were normal, including carcinoembryonic antigen and neuron-specific enolase. Chest computed tomography (CT) revealed a large amount of fluid in the right thorax and lung nodules (black arrow) in the right lower lobe (Fig. 1A-B). Chest computed tomography angiography (CTA) showed pulmonary arteriovenous

fistula (PAVF) in the right lower lobe (white arrows), with one feeding artery and one draining vein (Figure 1C-D). He was diagnosed with right hemothorax due to PAVF rupture, and emergency surgery was performed. Approximately, 2500 ml of blood and blood clots were found in the right thoracic cavity. After removal of the blood and blood clots, a PAVF lesion with active bleeding was found in the lateral basal segment, approximately 2.0 cm in diameter. Video-assisted thoracoscopic surgery was performed for pulmonary wedge resection of the right lower lobe. He was discharged 6 days after surgery.

Hereditary hemorrhagic teleangiectasia (HHT) is diagnosed according to Curaçao criteria: recurrent nosebleeds, mucocutaneous telangiectasia, visceral arteriovenous malformations, and an affected first-degree relative.[1] Between 60 and 90% of cases of PAVF are congenital and associated with HHT, which is a heterozygous autosomal dominant vascular disorder.[2,3] CTA is useful for visualization of feeding arteries and draining veins of PAVF.[4] Surgical resection and percutaneous embolization are optional treatment of this condition. In massive hemothorax due to PAVF, surgical resection is considered to be the first choice treatment.[3] Finally, consider rupture of PAVF when patients present with spontaneous hemothorax and pulmonary nodules or masses.

CONFLICT OF INTEREST STATEMENT

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this manuscript.

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