Paradoxical Systemic Embolization in Hereditary Hemorrhagic Telangiectasia

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A 53-year-old man presented with sudden onset left-sided weakness. His medical history included hereditary hemorrhagic telangiectasia with recurrent epistaxis. On examination, he was hypoxemic at rest and had left-sided flaccid hemiparesis. He was noted to have multiple lower lip telangiectasias (Figure 1). Computed tomography of chest revealed bilateral segmental and subsegmental pulmonary emboli (Figure 2, arrows) and a left lower lobe arteriovenous malformation (AVM) (Figure 2, arrowhead). MRI of brain and computed tomography of abdomen showed bilateral cerebral (Figure 3), splenic, and renal segmental infarcts. A venous duplex revealed a calf vein deep venous thrombosis that was thought to be secondary to a recent preceding knee surgery. We concluded that the patient had pulmonary embolization followed by paradoxical systemic embolization through the AVM. Patient eventually underwent transcatheter coil embolization of the malformation (pulmonary angiography, Figure 4a and b, arrows).

Pulmonary AVMs are aneurysmal, direct, low-pressure, artery-to-vein connections that result in a right-to-left shunt and are seen in 15% to 35% of patients with hemorrhagic telangiectasia. As illustrated in the vignette, most of these pulmonary AVMs may remain asymptomatic and a neurological catastrophe may be the first manifestation. Cerebral cortical infarctions attributable to paradoxical thromboembolism are seen in 14% of such patients with single AVM and is higher with multiple AVMs. It is recommended that hemorrhagic telangiectasia patients and their family members undergo periodic screening contrast echocardiography and chest radiograph, followed by chest computed tomography for AVMs. All patients with AVMs should receive prophylactic antibiotics before dental and other surgical procedures to minimize risk of cerebral embolic abscesses and those with symptomatic large AVMs should be considered for treatment with transcatheter coil embolization.

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Disclosures

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References


Figure 3. T2-weighted MRI of brain showing multiple bilateral infarcts.

Figure 4. (a) Pulmonary angiogram showing left AVM. (b) Pulmonary angiogram after catheter embolization of left AVM showing coil in situ.
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