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.1 As illustrated in the vignette, most of these pulmonary AVMs may remain asymptomatic and a neurological catastrophe may be the first manifestation.1 Cerebral cortical infarctions attributable to paradoxical thromboembolism are seen in 14% of such patients with single AVM and is higher with multiple AVMs.2 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.3 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.3,4

Acknowledgments

None.

Disclosures

Dr R. Kolluri, Speaker for GSK, Sanofi-Aventis and The Medicines Company. None for Dr N.K. Singh.
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.
Paradoxical Systemic Embolization in Hereditary Hemorrhagic Telangiectasia
Nishith K. Singh and Raghu Kolluri

_Circ Cardiovasc Imaging_. 2008;1:e11-e12

doi: 10.1161/CIRCIMAGING.108.788570

_Circulation: Cardiovascular Imaging_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2008 American Heart Association, Inc. All rights reserved. Print ISSN: 1941-9651. Online ISSN: 1942-0080

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circimaging.ahajournals.org/content/1/2/e11

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in _Circulation: Cardiovascular Imaging_ can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to _Circulation: Cardiovascular Imaging_ is online at:
http://circimaging.ahajournals.org/subscriptions/