Teaching NeuroImages:

Cerebral arteriovenous malformation in hereditary hemorrhagic telangiectasia

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Figure 1 Skin telangiectasias (A) and lip telangiectasias (B)

A 41-year-old woman with a history of recurrent epistaxis presented with left hemiplegia, right gaze deviation, and dysarthria. Physical examination confirmed skin and lip telangiectasia (figure 1). Her son had pulmonary arteriovenous malformation (AVM). Her head CT and cerebral angiography revealed right parietal hemorrhage with parasagittal AVM (figure 2).

Hereditary hemorrhagic telangiectasia (HHT, Osler-Weber-Rendu syndrome) is a disorder characterized by recurrent epistaxis, skin and mucosal telangiectasias, visceral and cerebral AVMs, and positive family history. Diagnosis

is made when three of the above criteria are present.¹ Cerebral AVMs affect 10% of patients with HHT. Routine screening for cerebral AVMs in asymptomatic patients with HHT remains controversial.^{1,2}

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Figure 2 CT (A), cerebral angiography (B), and magnified view of AVM nidus (C)

(A) CT: right parietal hemorrhage. (B) Cerebral angiography: right parasagittal arteriovenous malformation (AVM) supplied by the right distal anterior cerebral artery (white arrowhead), draining to superior sagittal sinus. (C) Microcatheter injection: magnified view of AVM nidus with aneurysmal appearance.

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