Neuroimaging Highlight

Editors: William Hu, Mark Hudon

Moyamoya Disease

Submitted by: Ian Fleetwood, Gary K. Steinberg

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A 20-year-old woman had originally presented at age 12 with a focal sensory seizure involving the right hand but had been seizure-free on anticonvulsants. At age 18, she developed right face and hand numbness, right hand tremor and ataxia that resolved without treatment. She had recently developed episodes of right hand and face numbness several times per week.

Her neurological examination identified mild (4+) weakness of proximal right leg muscles and inability to accurately perform fine finger movements with the right hand.

Investigations included MRI (Figure 1) and cerebral angiography (Figure 2).

The angiogram demonstrated bilateral internal carotid artery occlusions with impressive collateral flow through deep perforating arteries. The imaging findings were diagnostic of moyamoya disease.

The patient was admitted to hospital for staged revascularization procedures. She initially had a successful left superficial temporal artery (STA) to middle cerebral artery (MCA) bypass procedure to treat the symptomatic hemisphere. She returned one week later for a contralateral right STA-MCA bypass. Intra-operatively, a suitable recipient MCA branch was not available, and therefore an encephaloduroarteriosynangiosis (EDAS) procedure was performed. The two procedures were well-tolerated, without neurological deterioration, and she was discharged on ASA.

Moyamoya disease is a rare idiopathic condition that results from progressive occlusion of one or both internal carotid arteries or the proximal segments of the MCA or anterior cerebral artery. The incidence in North America is less than in Japan (0.07% per annum), where it was originally described.[1,2] Young patients most commonly present with episodes of cerebral ischemia as their cerebrovascular reserve is depleted, while adults tend to present with intracranial hemorrhage.[3]

This disease exhibits characteristic neuroimaging findings, but can occur with many associated diseases. While CT and MR scans show abnormalities consistent with infarction or

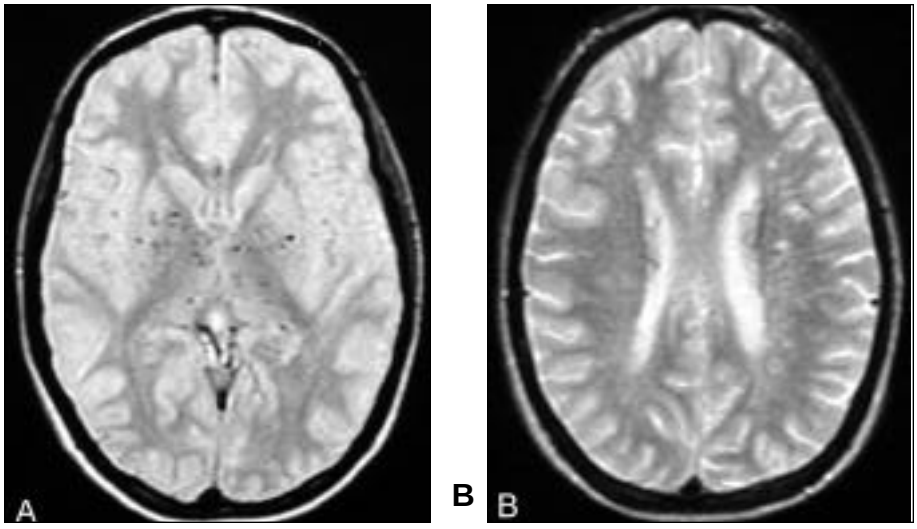

Figure 1: *Intermediate-weighted MR sequences demonstrating (A) multiple flow voids resulting from proliferation of perforating arteries in the thalamus, internal capsule, putamen and globus pallidus; and (B) several hyper-intense areas in the left hemisphere consistent with previous cerebral infarction.*

From the Department of Neurosurgery and Stanford Stroke Center, Stanford University, Stanford, CA, USA
Reprint requests to: Ian G. Fleetwood, Cerebrovascular Surgery Fellow, Stanford University, 300 Pasteur Drive, Stanford, CA94305-5327 USA

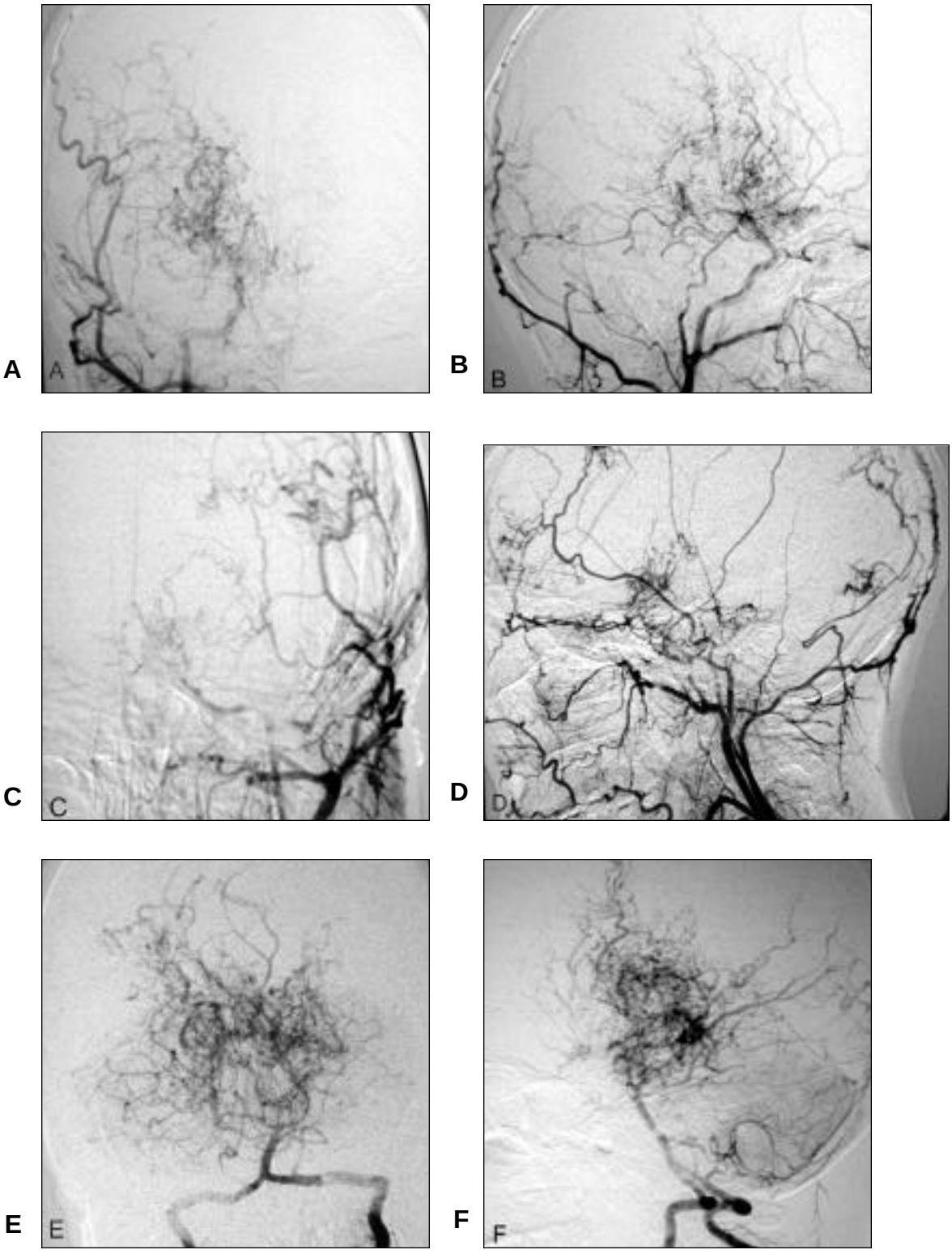

Figure 2: *Cerebral angiogram demonstrates the classic "puff of smoke" appearance of moyamoya disease. On the (A) APand (B) lateral right carotid artery projections, there is no filling of the MCAor ACAand proliferation of lenticulostriate perforators. On the (C) AP and (D) lateral left carotid artery projections, the findings are similar with less notable proliferation of perforators. This likely explains the previous cerebral infarctions in the left hemisphere. The (E) AP and (F) lateral vertebral injections demonstrate remarkable proliferation of thalamoperforating vessels.*

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hemorrhage, catheter cerebral angiography is diagnostic. Due to the stenosis or occlusion of the major intracranial vessels, patients develop deep collateral flow through dilated and tortuous perforating arteries. The "wavering puff of smoke" appearance is referred to in Japanese as moyamoya.[1]

Medical therapy for moyamoya patients is limited to the use of anti-platelet agents and vasodilators as patients become symptomatic.[4] Surgical treatments include direct anastomosis via extracranial – intracranial (EC-IC) bypass (e.g. STA-MCA bypass) and indirect anastomosis through EDAS, encephalomyosynangiosis or omental transpositions.[5] Work from our institution has shown that direct anastomosis is well tolerated and improves the natural history, angiographic appearance and cerebral blood flow abnormalities associated with moyamoya disease.[5]

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