Case Report

Fibromuscular Dysplasia: A Cause for Ischemic Stroke

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Case

A 40-year-old woman presented with two episodes of transient painless vision loss. The first episode involved the right visual field of both eyes and lasted approximately one hour after which there was a complete resolution of symptoms. The same symptom recurred the next day but this time it was associated with neck pain and lasted for four hours. There were no associated headaches, ocular pain, dizziness, vertigo, sensory deficits, or speech disturbances. The patient did not have significant past medical history except for the fact that she had recently recovered from a respiratory tract infection that was associated with violent coughing. She was not on any prescription medications and she denied tobacco, alcohol, or illicit drug use. Her physical examination showed no neurological deficits and a National Institutes of Health Stroke Scale score was 0. A magnetic resonance image (MRI) of the brain was obtained and showed evidence of acute infarction (Figure 1).

Given the history of violent coughing and associated neck pain, the initial concern was for an arterial dissection causing stroke. However, other etiologies to consider as part of the differential diagnosis for this young patient with ischemic stroke include vasculitis, hypercoagulable state, and other arteriopathies (such as fibromuscular dysplasia [FMD]). A cerebral angiogram was performed and showed irregularities within both vertebral arteries suggestive of FMD (Figure 2). The patient was treated with antiplatelet therapy (aspirin) and was discharged home. She had no further events and her modified Rankin Scale at one month was 0. Due to the lack of stenoses in the lesions and potential risk for dissection, the patient did not receive any endovascular therapies.

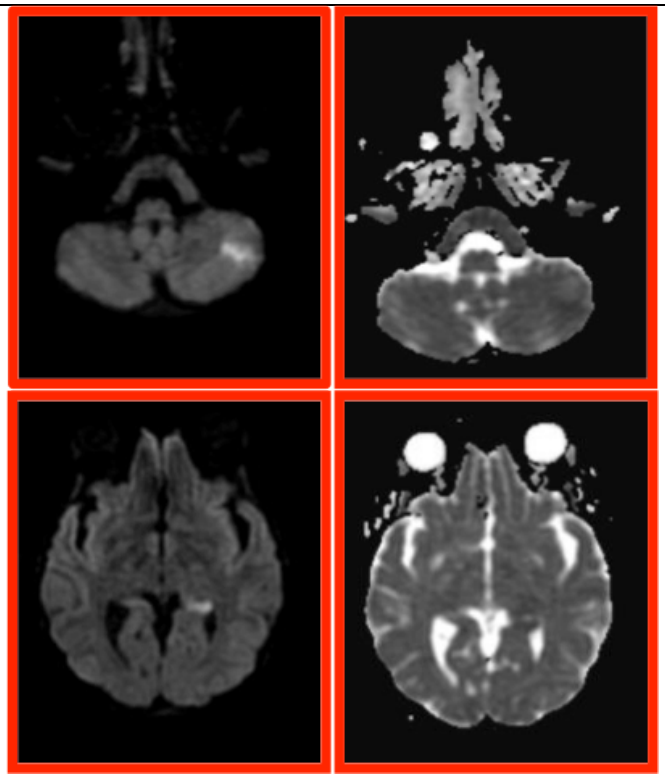

Figure 1: Diffusion weighted images (left) and apparent diffusion coefficient (right) showing two areas of acute infarction, one in the left cerebellum (top row) and one in the left thalamus (bottom row)

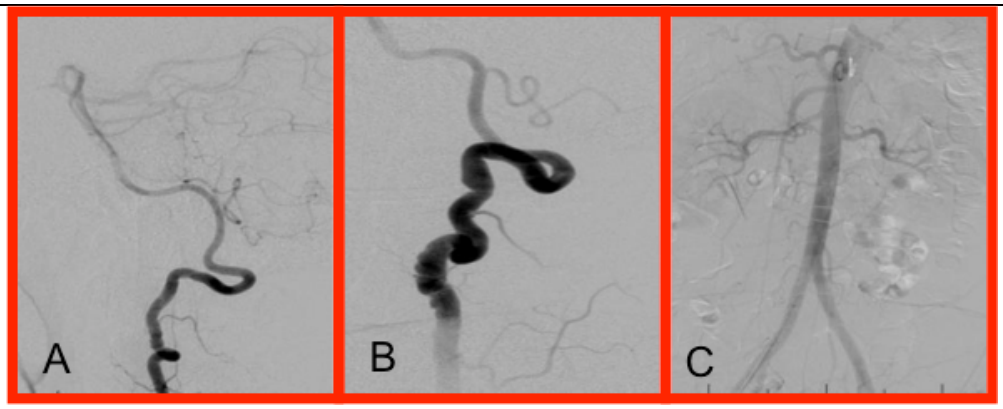

Figure 2: Vascular abnormalities similar to "string of beads" appearance in the right vertebral artery (A) and left vertebral artery (B). There is an absence of vascular abnormalities in the renal vasculature (C).

Discussion

FMD is a non-atherosclerotic and non-inflammatory arteriopathy that affects medium-sized arteries. FMD is thought to be rare, and the actual prevalence is unknown. One Austrian study in 2000 suggested that the prevalence is 0.5-0.7% of

cases that are diagnosed by cerebral angiography.[1] It occurs most commonly in the renal and carotid arteries. [1-3] It affects the renal arteries in 60-75% of cases with 35% cases occurring bilaterally. [2,4] Extracranial involvement of the cerebrovasculature (including both cervical carotid and vertebral arteries) occurs in 25-30% of cases and most commonly at the C1 and C2 levels.[2,4,5] There is an association with intracranial aneurysms in 7-50% of these patients.[2,4]

While most patients with FMD are asymptomatic, patients have the potential to develop stenoses, aneurysms, dissections, or occlusions within the affected vessels.[2] There is also a noted association with increased tortuosity of the carotid arteries without abnormalities suggestive of FMD.[2] Symptomatic FMD patients can complain of whooshing sounds in their ears, headaches, vertigo, lightheadedness, or syncope.[4] Ischemic strokes can occur in approximately 20% of cases of FMD. [6]

There has not been an evidence-based establishment of risk factors for FMD. Estrogen may play a role, as the female-to-male ratio for this condition is 9:1. Tobacco may also play a role.[2] The occurrence among twins suggests a genetic component for this condition; however, several studies have been completed without any conclusive evidence of genetic patterns.[2,4] There has been suggestion that a congenital or acquired deficiency of the vasa vasorum may play a role in the development of FMD in the carotid arteries.[1,7] Other conditions that have been found to be associated with FMD include alpha-1-antitrypsin deficiency, Ehlers-Danlos syndrome, Alport's syndrome, pheochromocytoma, Marfan's syndrome, and Takayasu's arteritis.[1,4]

FMD has been compared to atherosclerosis and vasculitis, but it is important to note the differences among the conditions. Atherosclerosis is common in the older population and affects proximal areas of arteries while FMD affects younger populations and involves the middle-to-distal portions of arteries.[2,5] Vasculitis is an inflammatory disorder associated with elevated inflammatory markers and fibrinoid necrosis while FMD is not.[2]

Olin et al. provides a pathologic classification system for FMD. Eighty to 90 percent of cases are classified as medial fibroplasias with alternating thinned media and thickened collagen-containing ridges that give the classic beading appearance. Approximately 10% are classified as intimal fibroplasias where collagen deposition within the intima is complicated by fragmented internal elastic lamina. Rarer forms include perimedial fibroplasia, medial hyperplasia, and adventitial fibroplasia.[2]

Conventional angiogram is the diagnostic test of choice.[3] There is little evidence to support the use of computed tomography or magnetic resonance angiogram imaging

as diagnostic measures.[5] Conventional angiogram shows the classic "string of beads" sign that is common with FMD.[1,3-7]

Once FMD is diagnosed appropriately, secondary risk factor prevention must be undertaken. First line agents to prevent complications from FMD include antiplatelet agents.[2,7] Graduated internal dilatation or angioplasty is the preferred intervention in symptomatic patients, such as those who present with headache, transient ischemic attack, or stroke; however, there are no large, randomized trials to support one treatment over another.[2, 5-7] Stenting is reserved for patients who develop dissection during angioplasty or those with aneurysms; carotid endarterectomy can be used in very special circumstances.[1,2]

Teaching points

1. FMD is a non-atherosclerotic and non-inflammatory arteriopathy that commonly affects the renal and carotid arteries.
2. FMD most commonly affects females.
3. FMD carries a good prognosis.
4. FMD can only be diagnosed accurately by conventional angiogram.
5. Once diagnosed, patients with FMD should be started on antiplatelet agents to prevent complications such as stroke. Symptomatic patients can undergo graduated internal dilatation or angioplasty.

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