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ISOLATED INTRACRANIAL FIBROMUSCULAR DYSPLASIA PRESENTS AS STROKE IN A 19-YEAR-OLD FEMALE

Lee A. Birnbaum, Richard Sherry, Edgard Pereira

ABSTRACT - Isolated intracranial fibromuscular dysplasia is rare and may present with cerebrovascular events. It should be considered as etiology of stroke in otherwise healthy young patients. Though diagnosis is often challenging, characteristic morphologies may be revealed on magnetic resonance and catheter angiography. Cephalocervical fibromuscular dysplasia typically involves the extracranial portion of the internal carotid artery (nearly 95%). This rare case demonstrates isolated intracranial fibromuscular dysplasia in a 19-year-old female with left caudate and genu of internal capsule stroke.

KEY WORDS: fibromuscular dysplasia, stroke, caudate nucleus, cerebral angiography.

Diplasia fibromuscular intracraniana isolada manifestando-se como acidente vascular cerebral em mulher de 19 anos

RESUMO - Displasia fibromuscular intracraniana isolada é rara e pode se apresentar como acidente vascular cerebral (AVC). Ela deve ser considerada como etiologia do AVC em paciente jovem saudável. Embora o seu diagnóstico possa ser um desafio, características morfológicas específicas podem ser reveladas através da angiografia através de ressonância nuclear magnética e por de catéter. A displasia fibromuscular envolvendo a circulação cérvico-cefálica afeta tipicamente a artéria carótida interna extracraniana (95%). O presente caso relata uma rara ocorrência de displasia fibromuscular intracraniana em uma jovem de 19 anos com infarto do núcleo caudado e joelho da cápsula interna à esquerda.

PALAVRAS-CHAVE: displasia fibromuscular, acidente vascular cerebral, núcleo caudado, angiografia cerebral.

Fibromuscular dysplasia (FMD) is a rare, segmental, non-atheromatous and non-inflammatory arterial disease of unknown etiology. The disease typically affects middle age women and involves intermediate-sized arteries throughout the body[1]. FMD is found predominantly within the renovascular (75 %) and cerebrovascular (24%) systems, but also has been reported in the mesenteric, iliac, and subclavian arteries[2]. Although the disease can affect all three arterial wall layers, the media is involved in 90 to 95% of the cases[3]. Pathological changes in the arterial wall result in distinct morphological appearances identified with neuroimaging techniques. Digital subtraction angiography (DSA) typically reveals an alternating pattern of concentric luminal narrowing and mural dilation, referred to as "string-of-beads"[4]. FMD is usually an incidental finding, but advanced disease may lead to arterial dissection, saccular aneurysm, or arteriovenous fistula, all of which, in cephalocervical FMD, may cause subarachnoid hemorrhage, transient ischemic attack, and/or stroke.

Based on a retrospective analysis of approximately 22,000 cerebral angiograms the incidence of cephalocervical FMD ranged from 0.25 to 0.6%[1]. It commonly affects the extracranial portion of the internal carotid artery[4], and in a few cases it can extend to the intracranial circulation. There are rare reports of isolated intracranial FMD, usually with poor angiographic documentation. In a recent comprehensive review, only 4 cases of isolated intradural FMD were identified[1]. The present case describes a 19-year-old woman with caudate stroke due to isolated intracranial FMD, emphasizing the importance of considereing this disease in the differential diagnosis of young patients with stroke, and discusses briefly the clinical aspects of caudate nucleus infarcts.

Departments of Neurology (LB, EP), Radiology (RS, EP) and Neurosurgery (EP), University of Louisville Hospital, Louisville, KY 40202, USA

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Edgard Pereira, MD - Neurointerventional Service - Dept. of Radiology - 530 South Jackson Street, Suite C07 - Louisville, KY 40202, USA - E-mail: edgard.pereira@louisville.edu

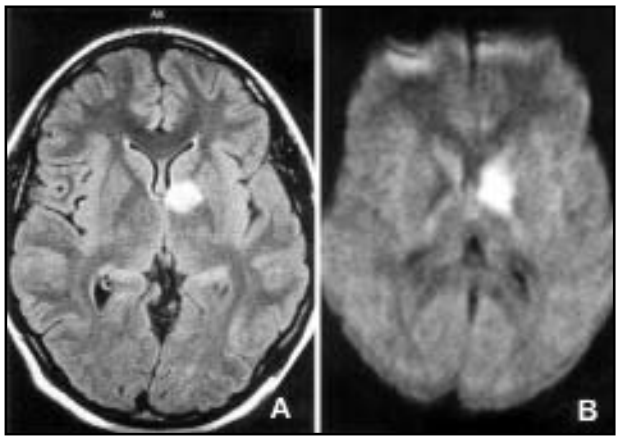

Fig 1. Axial FLAIR MR image demonstrated hyperintense signal in the left caudate nucleus and genu of the internal capsule (A). Diffusion-weighted image suggested acute infarct involving the left lenticulostriate arteries territory (B).

CASE

A 19-year-old woman was admitted to the neurology service with a 4-day history of left side throbbing headache associated with episodic bizarre behavior which included polyphagia, hypersomnia, truancy and visual hallucinations consisting of seeing little girls. Her past medical history was of no significance except for tobacco and social use of alcohol and marijuana. She denied any medication including oral contraceptives. Her physical examination was unremarkable. On neurological examination she was awake and alert but with "inappropriate affect". Language demonstrated hesitancy but normal naming, repetition and comprehension. There was no neglect, apraxia, dyscalculia or finger agnosia. Cranial nerve examination was normal except for left ptosis and mild right central facial weakness. Motor and sensory exams were normal. Gait and coordination were equally normal. The laboratory work up including CBC, electrolytes, calcium, PT, PTT, INR, urine drug screen, HCG, ESR, homocystein, anticardiolipins, RPR, rheumatoid factor and antinuclear antibody was normal. The transcranial Doppler and the carotid duplex were normal. Transesophageal echo was negative for septal defect, PFO or valvular disease. MRI of the brain demonstrated infarct involving the left caudate and genu of the internal capsule (Fig 1). Cerebral angiogram revealed abnormality involving the proximal segments of the left middle and anterior cerebral arteries characterized by regularly spaced areas of concentric luminal narrowing alternating with areas of dilation in a pattern consistent with FMD (Fig 2). MRA of the renal arteries was normal.

DISCUSSION

Arterial wall changes of FMD produce different angiographic findings including focal or tubular stenosis, multifocal stenosis alternating with mural dilation, the so-called "string-of-beads", septations, and aneurysms. The string-of-beads pattern occurs in 80-90% of FMD patients, and is associated with me-

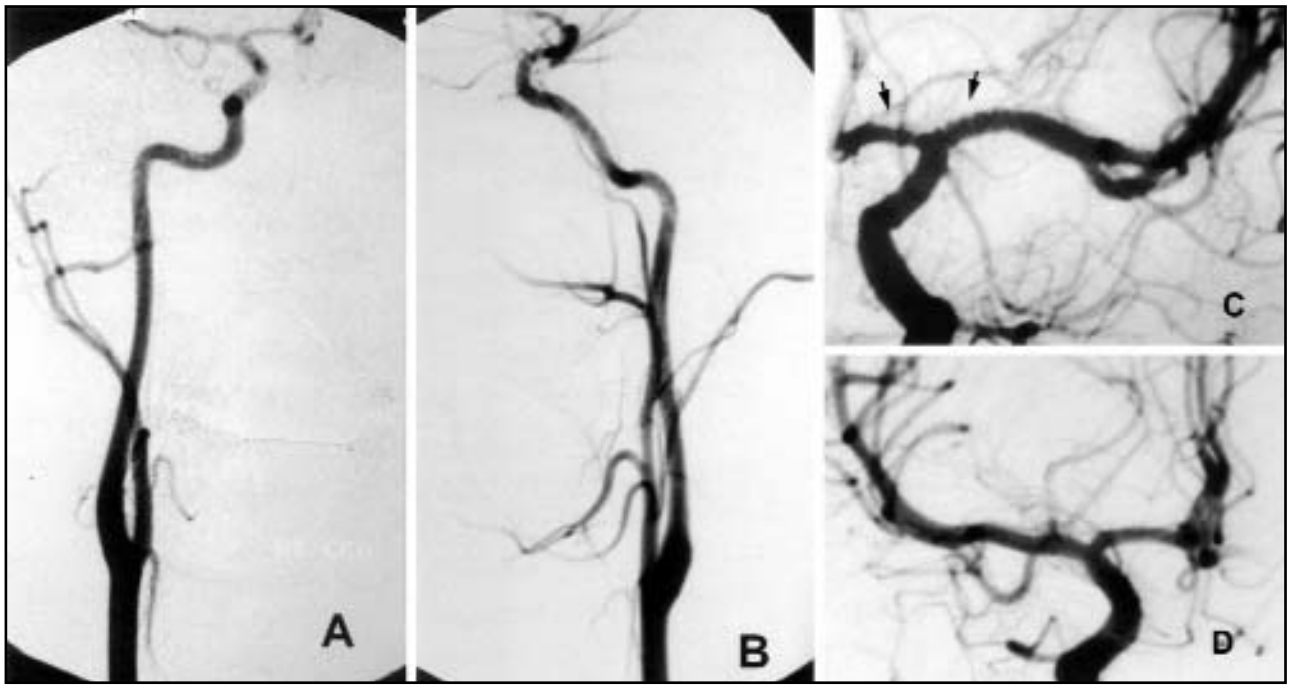

Fig 2. Digital subtraction angiography of the right (A) and left (B) common carotid arteries revealed normal vessels. Left internal carotid artery injection, in frontal view, clearly demonstrated luminal irregularities (arrows) in the proximal left anterior and middle cerebral arteries (C). Angiogram of the right internal carotid circulation, in frontal projection, showed normal vessels (D).

dial fibroplasia, the most common histologic type[5]. This characteristic pattern results from concentric bands of fibrous proliferation and smooth muscle hyperplasia that leads to medial thickening and destruction of the elastic lamina[5]. The rare intimal and even less common adventitial histologic types exhibit less specific appearances on DSA. Nevertheless, distinguishing FMD from other vascular diseases such as atherosclerosis and Vasculitis can be challenging. Of some help is that the areas of dilation in FMD are wider than the normal lumen[5]. Other vascular diseases exhibit normal lumen diameter at areas of relative dilation. The location of the lesion can also prove helpful in diagnosis. Atherosclerosis is commonly found in the proximal internal carotid artery (ICA), FMD is usually seen in the ICA adjacent to the C1-2 level[5]. Finally, multiple strokes make FMD a less likely diagnosis since the risk of recurrent strokes is low[6]. The angiogram performed in this case revealed a mural irregularity consistent with medial FMD. In our patient the angiographic findings of "string of beads" and her age led us to diagnose FMD rather than atherosclerosis.

Stroke in children and young adults is uncommon. At least one third of childhood strokes have no clear etiology[7]. The most frequently noted etiology for childhood stroke is cardiac malformation, followed by sickle cell disease, coagulopathies, metabolic derangements, and infectious processes[7]. FMD should additionally be considered in this differential. Compounding the rarity of this case is our patient's unusual presentation of episodic bizarre behavior, including overeating, excessive sleeping, missing school and work, and visual hallucinations. Nonetheless, the MRI showed an isolated caudate stroke, which explained the behavioral abnormalities. Lesions in the caudate have been associated with confusion, apathy, depression, and hallucinations[8].

Once FMD has been diagnosed, the course of treatment, which is controversial, must be considered. Options include no treatment, anti-platelets, anti-coagulants, angioplasty, and surgery. Most authors agree that an asymptomatic arterial segment of FMD should not be treated. However, symptomatic lesions, as in our case, present a dilemma. Although our patient was discharged on anti-coagulant therapy based on the results of retrospective studies of intracranial atherosclerotic disease favoring the use of warfarin over aspirin[10], the WASID trial, recently concluded, may determine the best therapy for stroke prevention in patients with intracranial stenosis[10].

The degree of stenosis and probability for aneurysm formation or rupture direct procedural intervention. Endovascular intervention, including angioplasty, stent placement, and/or coil embolization, has improved outcomes in symptomatic lesions[9]. For evolving ischemia in which endovascular treatment is not possible, bypass surgery may be an option.

In summary, FMD is a rare, non-artheromatous and non-inflammatory angiopathy of unknown etiology. FMD is a challenging diagnosis and may angiographically resemble atherosclerosis. FMD may distinguish itself by its characteristic "string-of-beads" appearance on angiogram, affinity for the ICA adjacent to the C1-2 level, as well as areas of dilation that are wider than the normal lumen. Treatment preferences are unique to each case and with the evolving capabilities of endovascular procedures outcomes for symptomatic FMD may improve.

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