Pediatric Neurology 53 (2015) 247–252

Contents lists available at ScienceDirect

Pediatric Neurology

journal homepage: www.elsevier.com/locate/pnu

Clinical Observations

Focal Cerebral Arteriopathy: The Face With Many Names

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ABSTRACT

OBJECTIVE: Focal cerebral arteriopathy is a term used to describe unilateral intracranial arteriopathy involving the distal internal carotid artery and proximal segments of the middle and anterior cerebral artery. We describe the disease course of 10 pediatric arterial ischemic stroke patients with focal cerebral arteriopathy from a single quaternary-care center. **METHODS:** We retrospectively reviewed pediatric stroke patients with focal cerebral arteriopathy without lenticulostriate collaterals treated at our institution between 2005 and 2014. Angiography was reviewed by a child neurologist and a pediatric neuroradiologist, and chart reviews were performed. **RESULTS:** Ten individuals with focal cerebral arteriopathy were identified. At the time of stroke presentation, four patients were diagnosed with arterial dissection, two with moyamoya disease, one with embolic occlusion, one with hemorrhagic stroke, and two with arterial dissection or vasculitis. At last follow-up, six patients had a change in diagnosis: four were diagnosed with transient cerebral arteriopathy, two with arterial dissection, and four with moyamoya disease. Four children experienced stroke recurrence. All were administered aspirin, one was administered heparin, two were administered intravenous tissue plasminogen activator, and five underwent surgical revascularization. **CONCLUSIONS:** Among pediatric stroke patients with a similar angiographic appearance, there is variable concordance between diagnosis, prognosis and treatment choice. Improved consensus-based diagnostic criteria and further research is needed to identify disease biomarkers and predictors of arterial progression.

Keywords: carotid artery dissection, pediatric stroke, focal cerebral arteriopathy, moyamoya disease

Pediatr Neurol 2015; 53: 247-252

Introduction

Cerebral arteriopathies are a common cause of stroke in children[1] and are strongly associated with stroke recurrence[2] and poor neurological outcome.[3] The term "focal cerebral arteriopathy" describes an angiographic appearance, with unilateral stenosis or vessel irregularity of proximal large intracranial arteries including the internal carotid (ICA), middle cerebral (MCA), and anterior cerebral arteries (ACA).[4] In the absence of defining angiographic characteristics that lead to a specific diagnosis, the differential diagnosis of patients with these angiographic findings includes post-varicella arteriopathy, transient cerebral arteriopathy, arterial dissection, moyamoya disease, fibromuscular dysplasia, or vasculitis.[5] Focal cerebral arteriopathy is an important subtype of childhood arteriopathies, accounting for 20% of all cases of childhood arterial ischemic stroke.[6]

In 2007, the International Pediatric Stroke Study working group developed the consensus-based Childhood Arterial Ischemic Stroke Standardized Classification and Diagnostic Evaluation (referred to as the CASCADE criteria) to standardize the classification schemes and improve research pertaining to patient outcomes, therapies, and risk of recurrence.[5] Despite these efforts, no definitive diagnostic criteria, biomarkers, or standardized treatments exist.[7] Diagnosis is further complicated by limitations of existing imaging modalities in differentiating vessel wall abnormalities.[1]

Article History:
Received March 10, 2015; Accepted in final form May 15, 2015

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0887-8994/$ – see front matter © 2015 Elsevier Inc. All rights reserved.
http://dx.doi.org/10.1016/j.pediatrneurol.2015.05.008

Treatments for childhood arteriopathies vary depending on suspected etiology and include aspirin, heparin, immunosuppression, and revascularization surgery. No clinical trials have evaluated the comparative efficacy of different treatment options.[8] The absence of definitive diagnostic criteria and treatment regimens complicates clinical care and standardization of research protocols.[1] In this case series, we describe the clinical and angiographic course of 10 pediatric stroke patients with focal cerebral arteriopathy to demonstrate the variability in diagnosis and treatment in patients with similar angiographic characteristics.

Methods

Patients were selected among children presenting to the Lucile Packard Children's Hospital at Stanford between 2005 and 2013 for acute management or follow-up of arterial ischemic or hemorrhagic stroke with abnormal arterial imaging. The Lucile Packard Children's Hospital at Stanford University is a quaternary-care center, which is a popular referral site for revascularization surgery. Included patients had an angiographic diagnosis of focal cerebral arteriopathy, defined as unilateral stenosis or occlusion involving the distal ICA and/or proximal segments of the ACA and MCA, in the absence of etiology-defining angiographic characteristics. Patients with etiology-defining angiographic characteristics such as lenticulostriate collaterals (suggesting moyamoya disease), double-lumen, or intimal flap (suggesting arterial dissection) were excluded, because these features provide more certainty in the etiological diagnosis and were therefore not the focus for this study. A retrospective chart review was performed on each patient. Data collection included age, gender, location and type of stroke, initial angiographic features, initial diagnosis, angiographic course, treatment, stroke recurrence, and final diagnosis. Neuroimaging and angiography review was conducted by consensus between two reviewers: a child neurologist and pediatric stroke specialist (J.E.) and a board-certified pediatric neuroradiologist (K.Y.). This study was approved by the center's institutional review board.

Results

Patient 1

A previously healthy 16-year-old girl developed sudden-onset, left-sided face, arm, and leg weakness and severe headache. There was no history of trauma; however, she was active in cheerleading. Magnetic resonance imaging (MRI) demonstrated a right MCA infarct involving the cortex and basal ganglia (Fig 1A). On initial magnetic resonance angiography (MRA), there was an abrupt cutoff at the right supraclinoid ICA. She was diagnosed with right ICA

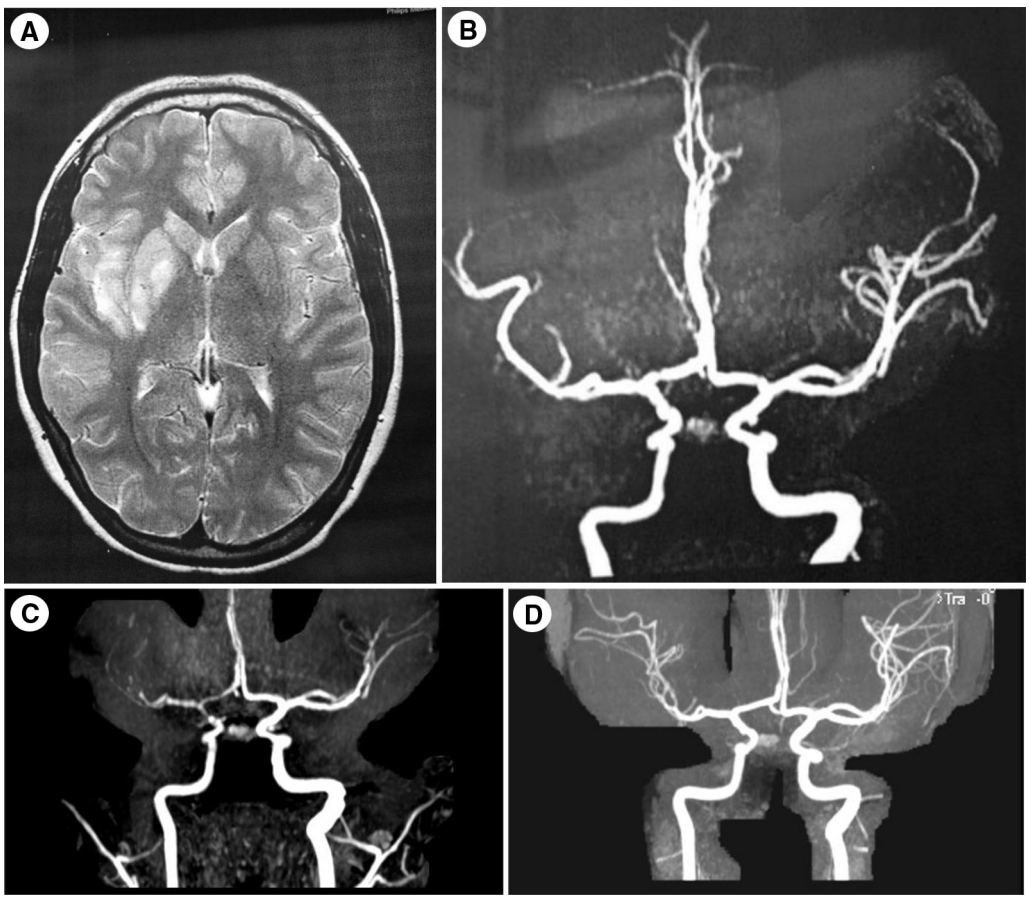

FIGURE 1.
(A) Baseline T2-weighted magnetic resonance imaging (MRI) demonstrating right MCA infarction involving the basal ganglia and adjacent cortex. (B) Magnetic resonance angiography (MRA) at the time of stroke recurrence demonstrating irregular segmental narrowing of the distal internal carotid artery (ICA), M1, and A1 consistent with beading, diagnosed as arterial dissection. (C) MRA at 3 months poststroke demonstrating progression with reduced flow in the distal ICA, and proximal middle cerebral artery (MCA) and anterior cerebral artery, diagnosed as Moyamoya arteriopathy. (D) MRA at 2 years poststroke demonstrating near complete resolution of arterial abnormalities, diagnosed as transient cerebral arteriopathy.