

Neuroimaging Highlight

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Reversible Wall Enhancement in Pediatric Cerebral Arteriopathy

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A previously well and developmentally normal five-year-old right-handed girl, presented with acute left-sided weakness and dysarthria. Investigations confirmed an acute right lenticulostriate stroke secondary to probable childhood primary angiitis of the CNS (cPACNS), including evidence of vessel wall enhancement (Figure). Despite extensive investigations, no alternative etiology for her stroke was detected. After three weeks of steroids, the child improved and the vessel wall enhancement resolved. Moderate hemiparesis, hemidystonia, and behavioural sequelae are present at 18-month follow-up.

Arteriopathy is the leading cause of childhood arterial ischemic stroke (AIS) and its recurrence.¹ Most common is an acute, unilateral arteriopathy affecting the middle cerebral artery/internal cerebral artery (MCA/ICA) that is thought to be parainfectious and/or inflammatory and has been classified by various terms including cPACNS. Initially described in adults and defined by the Calabrese criteria², cPACNS is an increasingly recognized, diagnostically challenging cause of childhood AIS.³ In addition to the large-medium vessel vasculitis shown here, a small vessel variety is also described.³ Diagnosis of cPACNS includes confirmation of symptomatic AIS with angiographic or histologic evidence of CNS vasculitis without an alternative cause.^{2,3} Non-inflammatory vasculopathies such as arterial dissection and Moyamoya disease are important differential diagnoses.³

Histologic evidence of vasculitis on brain biopsy remains the gold standard but this is usually impossible in large vessel cPACNS, emphasizing the need for better imaging markers. A large pediatric cohort study describing MR-MRA patterns of cPACNS suggests MRA sensitivity is only approximately 70%.⁴ When abnormal, the MRA most commonly showed proximal involvement of the carotid termination and proximal anterior cerebral artery and middle cerebral artery segments as seen in our patient. Cerebral angiography appears to be highly sensitive in large vessel cPACNS, often demonstrating irregular stenosis and dilatation with “banding or striae” of the same vessels (Figure D).

Vessel wall thickening and enhancement has been described in a small PACNS cohort that included eight children.⁵ It appears

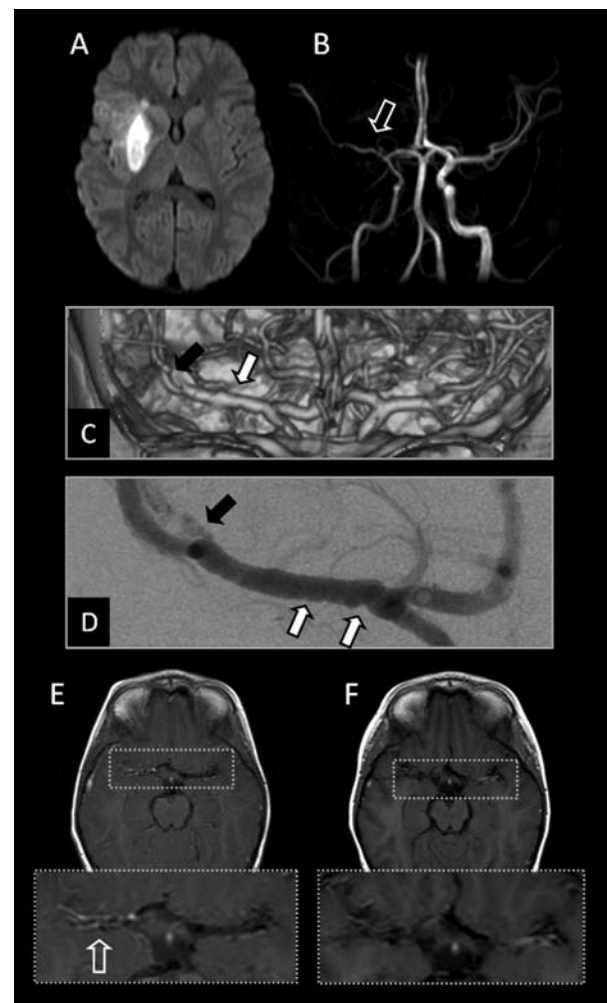


Figure: (A) Diffusion MRI confirms acute right lenticulostriate stroke. Vascular imaging with MRA (B), CT angiogram (C), and conventional angiogram (D) demonstrates proximal right M1 and M2 stenosis and extensive right M1 wall irregularity. Axial T1 with gadolinium (E) shows right M1 wall thickening and enhancement that is resolved 4 weeks later (F).

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to be a relatively valuable diagnostic finding as vessel wall thickening was identified in 25 of 27 patients and vessel wall enhancement was seen in 23 of 27 patients. Whether more advanced “wall imaging” can further delineate disease processes in the cerebral arteries is under investigation.

Treatment of cPACNS is controversial but immunosuppression has been suggested as an alternative.³ We demonstrate a potential correlation with conventional angiography and immunosuppression therapy. In the absence of more advanced wall imaging, we suggest that gadolinium-enhanced T1 sequences be included in acute pediatric AIS imaging to examine possible wall enhancement in the context of conventional angiography and other routine investigations. Establishing non-invasive markers of disease pathophysiology will enable treatment trials in childhood AIS.

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