We continue to assert that MRI offers important advantages over CT scanning in the diagnosis of ICA dissection, and we believe this to be the investigation of choice in situations where this condition is suspected.

References

Suspected Isolated Angiitis
Causing Stroke in a Child

To the Editor:
Cravito and Feigin originally described isolated angiitis of the central nervous system in 1959. Today, it is a rare etiology for stroke with only seventy-eight cases described in a most recent literature review. This disease has been reported in a 7-year-old boy infected with herpes zoster. We report the suspected occurrence of isolated central nervous system angiitis in a child without apparent systemic disease. The case illustrates that isolated angiitis must be considered in the evaluation of all childhood strokes. A 5-year-old boy was admitted to the hospital in September 1988 for evaluation of recurrent right hemiparesis. He developed transient right-sided weakness and slurring of speech lasting 30 minutes during the afternoon prior to admission. During that evening, a recurrent right hemiparesis developed, which persisted and precipitated hospitalization. Two months earlier, he had suffered a 45-minute episode of transient right hemiparesis. On admission, he was alert, with a mild expressive aphasia. The general physical examination was unremarkable, with vital signs, skin, cardiovascular, and lung exams entirely normal. Neurologic exam revealed central right facial nerve weakness, mild right hemiparesis, and decreased fine motor movement in the right hand.

Magnetic resonance imaging (MRI) revealed two high-signal areas in the left basal ganglia, suggesting infarction (Figure 1). The sedimentation rate was mildly elevated at 17 (10-13 mm/hr). Laboratory studies including general blood chemistry, PT, PTT, lipid profile, SPEP, hemoglobin electrophoresis, and complement studies were normal. Chest x-ray, EKG, echocardiogram, and lumbar puncture were normal. Other negative studies included RF, VDRL, ANA, anti-SS ANA, ENA, lupus anticoagulant, lupus-like inhibitor, protein C and S, and urine, blood, and CSF cultures.

Bilateral carotid angiography performed on the second hospital day (Figure 2) showed beading on the left middle cerebral artery that was suggestive of vasculitis and associated infarction. We seriously considered a diagnosis of isolated angiitis of the central nervous system when no secondary cause of vasculitis was uncovered.

We started prednisone 40 mg daily (2 mg/kg) and discharged the patient on the ninth hospital day. Prednisone dosage was tapered slowly on an outpatient basis and discontinued after four months when his motor deficits resolved. Selective left carotid angiography performed in May 1989 revealed improvement in the narrowing of the left middle cerebral artery. To date, this child has remained symptom-free.

The proposed criteria for diagnosis of isolated angiitis of the central nervous system include: 1) headaches and multiple neurologic dysfunctions present for at least 6 months or the acute onset of a major deficit, 2) cerebral angiography showing luminal narrowing, 3) absence of an inflammatory or infectious disease, and 4) a leptomeningeal biopsy revealing vascular inflammation. Our case fulfilled the first three criteria. However, brain biopsy is the definitive diagnostic test, although it is only positive 50% of the time. We were unable to justify a brain biopsy in a 5-year-old child who was improving and whose angiogram had no detectable secondary cause for vasculitis. Thus, we have only a very high suspicion for isolated angiitis.

At this time, we cannot comment on possible treatment for affected children although this patient did appear to improve while on prednisone. In the adult, cyclophosphamide and alternate day prednisone for at least 1 year have been recommended for rapidly progressive or steroid-resistant cases. There are no guidelines for the diagnosis and treatment of this disease in children. However, this case illustrates that isolated angiitis of the central nervous system may affect children and could be a potential cause of pediatric stroke. Thus, as pediatric cases of isolated angiitis are confirmed by biopsy, appropriate studies and comparison to the adult variant of this disease can be performed.

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Suspected isolated angiitis causing stroke in a child.
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Stroke. 1990;21:1380
doi: 10.1161/01.STR.21.9.1380

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