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Intracranial Dissection Mimicking Transient Cerebral Arteriopathy in Childhood Arterial Ischemic Stroke

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Abstract

Transient cerebral arteriopathy is a frequent cause of childhood arterial ischemic stroke. Differentiating this condition from intracranial carotid artery dissection is challenging but important for initial treatment. We describe 4 cases from the International Pediatric Stroke Study of intracranial carotid artery dissection, initially misdiagnosed as transient cerebral arteriopathy. Presentations were abrupt, with focal neurological deficits in 4, preceding headache in 3, and minor trauma in 1. Infarcts involved the anterior circulation, and magnetic resonance angiography showed unilateral arterial stenosis/occlusion. None had evidence of dissection. All received anticoagulation or thrombolysis. Three died from refractory intracranial hypertension. Intracranial carotid artery dissection was confirmed postmortem (n = 3) and on dedicated MR wall imaging showing intramural hematoma (n = 1). In differentiating transient cerebral arteriopathy from intracranial carotid artery dissection, routine magnetic resonance angiography is unreliable and adjunctive conventional angiography, gadolinium magnetic resonance angiography, or dedicated MRI wall imaging should be considered.

Keywords

stroke, dissection, transient cerebral arteriopathy

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Arteriopathies underlie 50% to 64% of childhood arterial ischemic stroke.[1] The most frequent is unilateral arteriopathy involving the distal internal carotid, proximal middle cerebral, or proximal anterior cerebral artery(s). This condition, originally published as "transient cerebral arteriopathy," is common and presumed to be inflammatory based on postmortem evidence of arteritis in post varicella cases.[2]

Intracranial carotid artery dissection can also involve unilateral intracranial segments of these arteries. In transient cerebral arteriopathy, conventional angiography demonstrates irregular stenosis and banding in affected arteries. In intracranial carotid artery dissection, specific signs include double lumen, intimal flap, and wall hematoma on magnetic resonance imaging (MRI). Risks, including radiation exposure, have led to reduced use of conventional angiography. MR angiography has become the most popular vascular imaging study in childhood stroke.

Transient cerebral arteriopathy and intracranial carotid artery dissection are difficult to differentiate initially, although the subacute course of the stenosis is informative. In transient cerebral arteriopathy, arterial stenosis frequently increases over the initial 3 months, then stabilizes or improves beyond 6 months.[3] Residual permanent stenosis and recurrent stroke are frequent. In intracranial carotid artery dissection, arterial stenosis typically improves over the initial 3 months.[4]

Differentiating transient cerebral arteriopathy from intracranial carotid artery dissection at the time of initial stroke influences choice of initial therapies. On the basis of probable inflammatory etiology, transient cerebral arteriopathy

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Table 1. Initial Radiological and Pathological Findings

Case	Age, y, (sex)	Trauma	Infarct Territory[a]	Vascular Imaging[a]	Initial Diagnosis	Final Diagnosis
1	10 y (F)	No	R MCA[a] (C; BG)	R dICA/ M1[a] hMCA	TCA	Postmortem ICAD
2	6 y (F)	No	R MCA[b] (C; BG)	R M1[b]	TCA	Postmortem ICAD
3	6 y (F)	Minor head bump	L MCA/ACA[a] (C; BG)	L dICA/M1/A1[a] hMCA	TCA	Postmortem ICAD
4	17 y (M)	No	L MCA[a] (BG)	L dICA/M1/A1[b]	TCA	ICAD

[a] Computed tomography, computed tomographic arteriography.
[b] Magnetic resonance imaging; magnetic resonance angiography.
Abbreviations: A1, proximal anterior cerebral artery; BG, basal ganglia; C, cortex; dICA, distal/supraclinoid internal carotid artery; F, female; h, hyperdense; L, left; M, male; M1, proximal middle cerebral artery; R, right; MCA, middle cerebral artery; ACA, anterior cerebral artery; ICAD, intracranial carotid artery dissection.

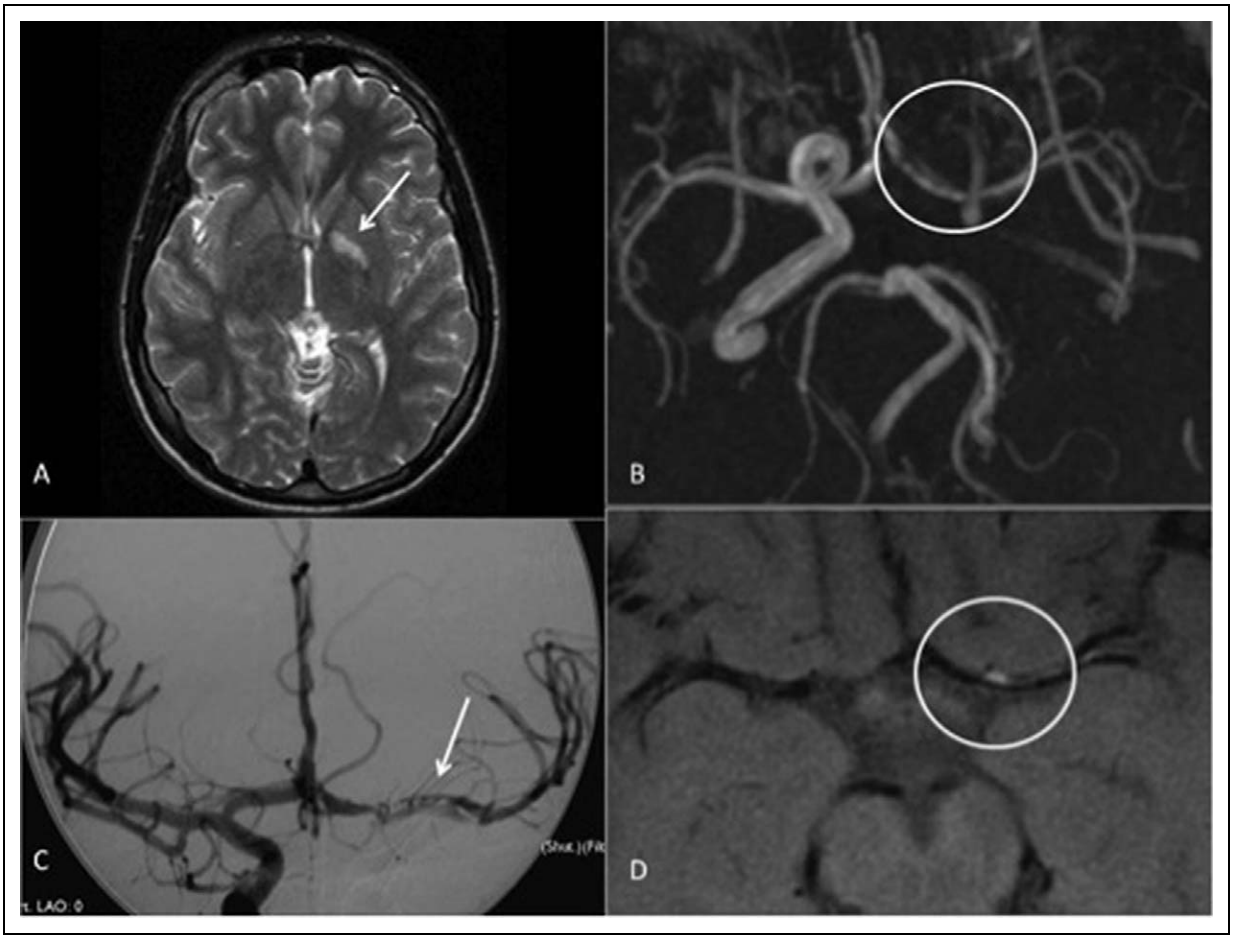

Figure 1. Case 4. A, Magnetic resonance imaging (MRI; arrow) showing left basal ganglia diffusion restriction. B, MR angiography nonvisualization of distal internal carotid artery and (circle) signal irregularity in left anterior cerebral artery and middle cerebral artery. C, Conventional angiography (arrow) left middle cerebral artery contrast filling defect suggestive of recanalizing thrombus or double lumen. D, MR vessel wall signal hyperintensity (circle) suggestive of intramural thrombus.

is increasingly treated with immunosuppressive medications.[5] For intracranial carotid artery dissection, adult and pediatric stroke guidelines recommend avoiding anticoagulation, although this is controversial.[6,7]

We describe 4 children initially diagnosed with transient cerebral arteriopathy but later determined to have intracranial carotid artery dissection. We explore challenges and solutions to initial diagnosis and management of these conditions.

Methods

International Pediatric Stroke Study[8] members pooled 4 cases of childhood arterial ischemic stroke initially diagnosed with transient cerebral arteriopathy and later discovered to have intracranial

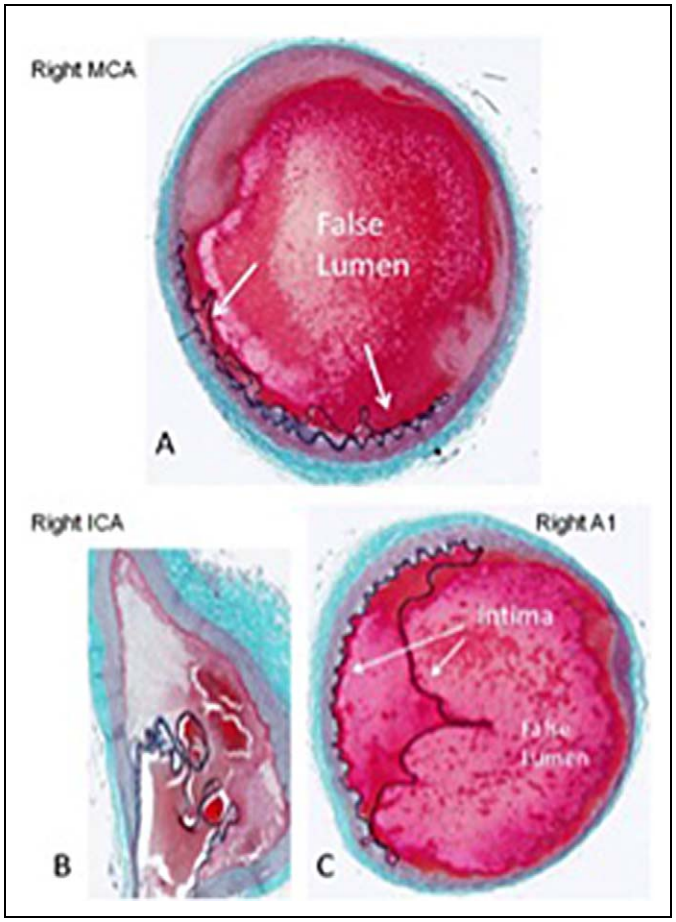

Figure 2. Case 1: Elastic trichrome stained postmortem sections demonstrating dissection with intima and internal elastic lamina (arrows) stripped from smooth muscle media.

carotid artery dissection. Intracranial carotid artery dissection was diagnosed postmortem in 3 and antemortem in 1 child through dedicated wall imaging MR techniques. Clinical, imaging, and pathology data from each case were reviewed centrally at The Hospital for Sick Children. Institutional Review Board approval and parental consent were obtained for all patients.

Results: Summary of Case Features

At presentation, all children had acute focal neurological signs, preceded by acute headache in 3 and minor head trauma in 1. Neuroimaging demonstrated acute anterior circulation infarcts, including basal ganglia. Children were all initially diagnosed with transient cerebral arteriopathy with typical infarct and vascular imaging findings. Three had additional cortical infarction (Table 1). Conventional angiography, in 1 child, showed questionable wall hematoma (Figure 1). Treatment comprised anticoagulation in 3 and thrombolysis in 1; however, 3 children developed fatal malignant infarct edema despite emergency neurosurgical interventions.

Postmortem pathology demonstrated unexpected intracranial carotid artery dissection in 3 children, without inflammatory arteriopathy (Figure 2). Dedicated MR wall imaging demonstrated wall hematoma in the fourth, confirming intracranial carotid artery dissection (Figure 1).

Discussion

Our cases demonstrate that intracranial carotid artery dissection can be misdiagnosed as transient cerebral arteriopathy if initial vascular imaging lacks characteristics specific to dissection. Under-recognition and delay to diagnosis present significant barriers to optimal early management of childhood intracranial carotid artery dissection.

Clinical presentation does not reliably differentiate transient cerebral arteriopathy and intracranial carotid artery dissection. Our children presented with mainly nonspecific features, including hemiparesis. Headache, developing over minutes to hours in 3 cases, might suggest intracranial carotid artery dissection.[9] However, one-third of children with arterial ischemic stroke have headache at onset.[10]

Radiographic presentation does not reliably differentiate transient cerebral arteriopathy and intracranial carotid artery dissection. Infarcts in transient cerebral arteriopathy typically involve striatocapsular structures, present in our 4 cases.[3] Additional cortical infarction in 3 can point to intracranial carotid artery dissection.[11]Arterial stenoses involving the distal internal carotid, proximal middle cerebral, or proximal anterior cerebral artery(s), present in all cases, typifies both intracranial carotid artery dissection and transient cerebral arteriopathy. Not surprisingly, MR angiography uniformly failed to demonstrate dissection in our cases. Time-of-flight MR angiography misses 50% of carotid dissections that are confirmed by conventional angiography.[12] In a single case, dedicated arterial wall imaging with MRI made a diagnosis of intracranial carotid artery dissection by demonstrating wall hematoma.

Diagnosis of intracranial carotid artery dissection requires a high index of suspicion and either conventional angiography or MRI demonstrating wall hematoma.[13] Atherosclerosis is rare in childhood arterial ischemic stroke; therefore, segmental gadolinium arterial wall enhancement and absence of wall hematoma provide presumptive evidence of inflammation and transient cerebral arteriopathy.

Conclusion

Our case series demonstrates challenges and potential solutions in differentiating intracranial carotid artery dissection from transient cerebral arteriopathy in childhood anterior circulation stroke. Diagnosis in life presents challenges with traditional imaging techniques, and specific diagnosis leads to different treatment considerations. Therefore, we suggest that conventional angiography or dedicated MRI arterial wall imaging techniques be considered in children with unilateral intracranial arteriopathy to assist in specific diagnosis.

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Author Contributions

ND, GAdV, HJF and MTM contributed to the study concept and design. ND acquired the data and drafted the manuscript. JLF, MTM, MS, CH, HJF and GAdV performed critical reviews of the manuscript. GAdV acted as supervisor and approved the final manuscript.

Declaration of Conflicting Interests

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Ethical Approval

Institutional Review Board approval and parental consent were obtained for all patients.

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