

Acute Pontine Ischemic Stroke in a Healthy Child With Intracranial Vasculopathy

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Abstract

Here we report the case of a previously healthy 8-year-old boy who presented with altered mental status, right facial droop and right-sided hemiplegia the day after playing in an inflatable bouncer. No head trauma was reported by the patient nor witnessed by the parents. Urgent magnetic resonance imaging (MRI) demonstrated acute ischemic infarction in the left pons; computed tomographic angiography excluded arterial dissection but identified a small hyperdense filling defect in the basilar artery, later confirmed to be a calcification at the origin of a perforating artery. Pediatric National Institutes of Health (PedNIH) Stroke Scale score was 15. Infectious, inflammatory, hypercoagulable and additional vascular causes were excluded. Although the cause of the calcification remains obscure, we speculate that, similarly to mineralizing microangiopathy, a minor trauma led to stroke in this child. To our knowledge, mineralizing microangiopathy, the well-described entity affecting perforating arteries of the anterior circulation in young children leading to basal ganglia stroke following minor head traumas has not been described in the posterior circulation or in previously healthy school-age children.

Keywords

pediatric stroke, pontine infarct, ischemic stroke, trauma, mineralizing microangiopathy, intracranial calcifications, MRI, magnetic resonance imaging, hemiplegia

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A previously healthy 8-year-old boy presented with a chief complaint of “difficulty walking.” On the day prior to presentation, the child was on vacation with his family. He was playing in an inflatable bouncer and later complained of acute onset left eye pain, headache and nausea. No head trauma was reported by the patient or seen by the parents. He was able to ambulate without difficulty and later brought to his room uneventfully. The following morning on the day of presentation, he was unable to walk and fell to his right. Given the acute presentation and progressive nature of these symptoms, he was brought to a local hospital emergency department. Initial evaluation showed right facial weakness in addition to right upper and lower extremity flaccid weakness. The PedNIH Stroke Scale was 15.¹

A brain magnetic resonance image (MRI) was obtained and was concerning for a left pontine ischemic stroke (Figure 1A, B). With the finding of an acute ischemic posterior circulation stroke in a previously healthy child we suspected vertebral dissection possibly related to unwitnessed head/neck trauma. As infectious and inflammatory events have been increasingly associated with pediatric stroke,² we were also concerned about the possibility of focal or generalized vasculitis affecting the small arteries of the posterior circulation. We additionally considered thromboembolic events, a hypercoagulable state

and congenital stenosis or malformation of extra- or intracranial vasculature.

The patient was transferred to the Pediatric Intensive Care Unit at Children’s National Health System for close monitoring and hourly neurologic evaluations. Given the history of playing in an inflatable bouncer despite no reported history of head trauma, there was a high level of suspicion for a dissection of the intra- or extracranial vasculature. Computed tomographic angiography (CTA) of the head and neck was obtained and findings were consistent with a pontine ischemic stroke but no dissection was identified. Neurologic examination was significant for altered mental status, right facial droop, bulbar

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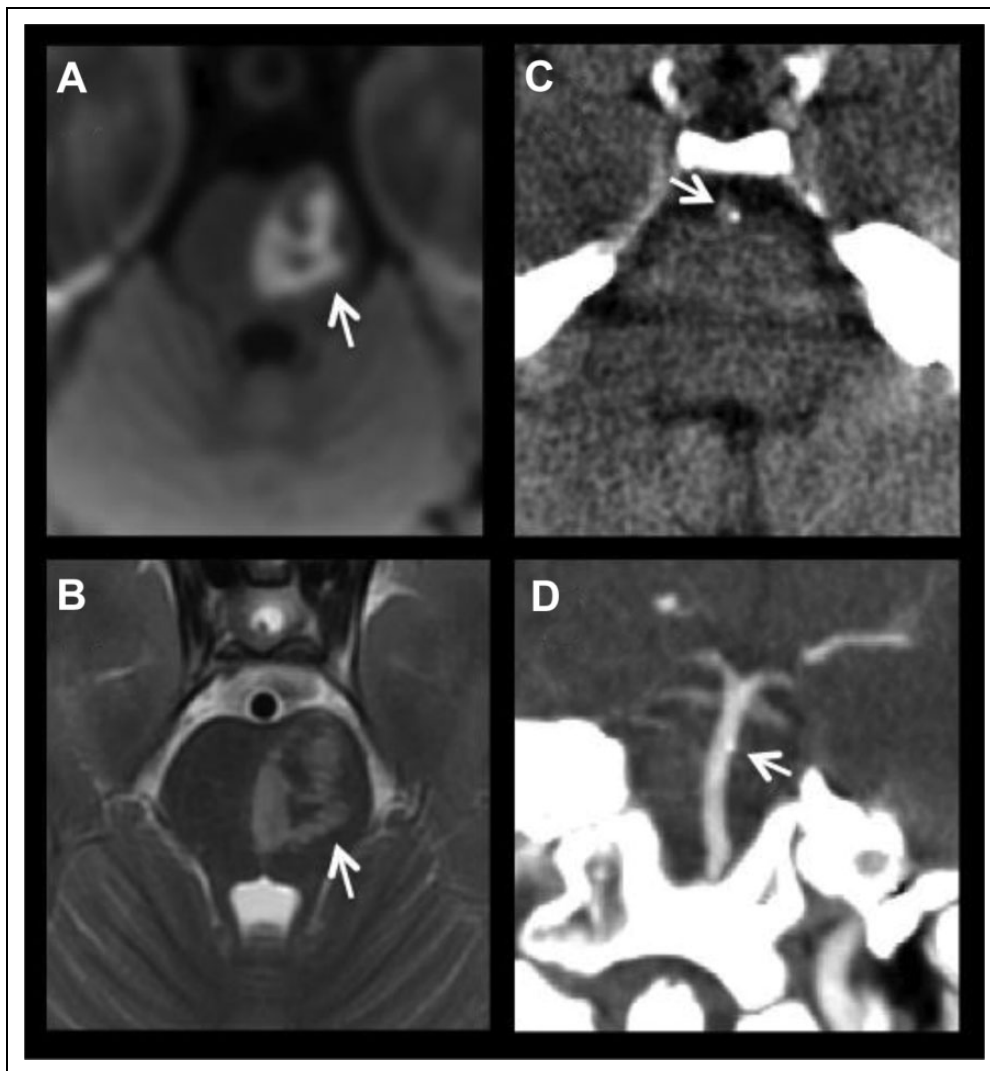


Figure 1. Acute pontine stroke in a healthy child with intracranial vasculopathy. (A) Diffusion-weighted imaging and (B) T2 fluid attenuated inversion recovery showing acute left-sided pontine stroke (arrow). (C) Head and neck CT angiography showed an intracranial calcification at the junction of a perforating artery, in axial view (arrow). (D) View of the vertebral artery in coronal section showing the calcification (arrow).

weakness with hypophonia and right-sided hemiplegia. Reflexes were reduced on the left and normal on the right. Coordination was normal on the left but difficult to test on the right due to weakness. Gait examination was deferred due to right-sided hemiplegia.

Review of the initial brain MRI and CT angiography by a pediatric neuroradiologist was concerning for a hyperdensity with a small filling defect in the basilar artery, later confirmed to be a calcification at the origin of a perforating artery (Figure 1C, D). Brain MRI confirmed the acute left pontine ischemic stroke. An extensive workup for the etiology was performed. Complete blood count, basic metabolic profile, liver function tests, lipid profile, erythrocyte sedimentation rate, C-reactive protein, thyroid-stimulating hormone, and prothrombin time and partial thromboplastin time were all within normal limits on admission. Blood cultures were negative for any occult infection. Thrombophilia workup was negative. Genetic testing

for factor II (G202 10A) and factor V (Leiden) mutation testing was also negative. Homocysteine and lipoprotein A levels were within normal limits. Antinuclear antibody screen, anticardiolipin, and B2 glycoprotein I antibody testing were negative. Cardiac echocardiogram was negative for a thrombus or intracardiac shunt. Therefore, cardioembolic, hematologic and autoimmune etiologies were not identified.

The patient was placed on heparin on the day of admission (approximately 36 hours after last known normal) and this was continued for about 2.5 days before the patient was transitioned to daily aspirin on the fourth day of the admission. He did not have any further neurologic deterioration while in the pediatric intensive care unit and he was later transferred to the Neurology Ward. He steadily improved during the admission and was discharged to inpatient rehabilitation, where he received intensive physical, occupational, speech and recreational therapy. He was continued on aspirin at time of discharge.

A complete neuropsychological profile was obtained on admission and in follow-up after discharge from the hospital. Prior to the injury the patient was a very good student in the second grade. An initial limited evaluation showed limitations in temporal orientation, expressive language and executive function. On follow-up evaluation 6 months after the stroke, a detailed neuropsychology evaluation demonstrated word-finding difficulties in addition to weakness with fine motor speed and dexterity, working memory and sustained attention, processing speed, reading decoding and reading fluency. He was diagnosed with a mild neurocognitive disorder due to the stroke and a specific learning disability that was characterized as mild to moderate in severity. The patient received an individualized educational plan with environmental modifications in school.

Repeat imaging 6 months after the initial presentation was obtained. Brain MRI showed interval radiographic maturation of the pontine infarction with development of coagulative necrosis with increased conspicuity of Wallerian degeneration affecting the left corticospinal tract. There was no evidence of acute or interval infarction since the initial presentation. A small focal cortical dysplasia with gray matter heterotopia involving the cortex and subcortical white matter of the left inferior frontal gyrus was also identified but is thought to be an incidental finding. CT angiography and MR angiogram showed no evidence for vasculitis, *moya moya* or other focal cerebral arteriopathy on initial or follow-up imaging.

Follow-up evaluation with Neurology and Physical Medicine & Rehabilitation 8 months after the initial presentation for acute stroke showed that he was able to hold a pencil and write with his right hand and that his handwriting was closer to baseline. He was independent with all mobility, including stairs and was back to running. He continued to have some difficulty with processing speed and completed outpatient physical, occupational, and speech therapy programs. School-based occupational therapy continued and was focused on improving handwriting. He had a gradual return to school and completed the second grade. His neurologic examination was significant for 4+ to 5/5 strength on the right (movement possible against some resistance by the examiner) and 5/5 on the left (normal strength). Tone was significant for a mild spastic catch to the right ankle. He also had slightly slowed finger-nose-finger on the right side, and 3 beats of clonus with right ankle dorsiflexion. He was continued on aspirin 81 mg daily. He was cleared to return to soccer and for riding a bicycle and skateboarding with a helmet. We recommended avoiding contact sports.

On follow-up with Neurology 16 months after presentation the patient was in the third grade, no longer receiving occupational therapy in school. He continued to have an individualized educational plan. He was noted to have a subtle limp on the right side but played soccer and was able to keep up with his peers. On neurologic examination, strength continued to improve with only mild weakness in the proximal right upper and lower extremities. Follow-up imaging at the time of this evaluation showed stable cystic encephalomalacia in the left hemi-pons related to prior insult and no new evidence for acute

or interval stroke. MRA of the head was normal, as in prior. Patient was continued on daily aspirin for secondary stroke prevention. On follow-up with the family 18 months after the initial presentation, the patient's mother reported that he was "doing amazing! He is a typical young boy with a lot to look forward to."

Discussion

With the clinical picture of headache and hemiparesis following possible neck/head trauma (from jumping), arterial dissection was initially suspected. However, after the CT angiography revealed a small calcification in the arterial wall we believe that a mechanism of endothelial damage with possible clot formation was the cause of this child's stroke similarly to mineralizing microangiopathy. This is supported by the patient's presentation as he was initially normal and deficits were found the following morning after playing in the inflatable bouncer. Mineralizing microangiopathy is a well-described entity affecting perforating arteries of the anterior circulation in children less than 2 years of age leading to ischemic stroke in previously healthy infants and toddlers after minor head trauma. Imaging typically reveals unilateral basal ganglia infarction and lenticulostriate artery mineralization.³⁻⁶ This entity can also be secondary to combined radiation and chemotherapy treatment in school-age children with central nervous neoplasms.⁷⁻¹⁰ However, our patient was older and did not have any identifiable risk factors. Our patient had altered mental status, which has been described before for mineralizing microangiopathy.¹¹ This may suggest an underlying genetic predisposition that contributed to the formation of the arterial calcification. Head trauma was not reported by the patient nor the parents but minor head trauma is difficult to rule out definitively. Further genetic testing, including whole exome sequencing, should be considered for similar cases in the future. To our knowledge, mineralizing microangiopathy has not been reported to affect the posterior circulation or to occur in school-age children.

In addition, lenticulostriate vasculopathy is a well-described entity of the anterior circulation, known to occur in 0.4% of all live-born neonates and in 1.9% to 5.8% of ill neonates in association with variety of congenital and acquired disorders.¹² A potential role for hypoxic-ischemic perinatal conditions has additionally been postulated.¹³ Lenticulostriate vasculopathy is characterized by thickened hypercellular vessel walls with intramural and perivascular mineralization. This process generally regresses over time although the association between lenticulostriate vasculopathy and mineralizing angiopathy leading to basal ganglia stroke has been demonstrated.⁴

To our knowledge, a similar entity to lenticulostriate vasculopathy has not been described in the posterior circulation. Use of head CT as the preferred imaging modality in children with stroke particularly in the youngest patients may have contributed to underrecognition of a similar entity affecting the posterior circulation and leading to acute stroke. Indeed, it was

demonstrated that routine head CT with 5-mm sections fails to detect the vascular mineralization in infants.⁴

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Author Contributions

YAK wrote the first draft of the manuscript. JM, JB, MD and PP reviewed and edited the manuscript, and edits were reviewed and incorporated by YAK. PP supervised the work. All coauthors reviewed and approved the final version of the manuscript.

Declaration of Conflicting Interests

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Ethical Approval

This is a retrospective case report and does not qualify as human subjects research requiring IRB review and approval. Protected Health Information was not disclosed but permission to share the case was obtained from the patient's parents.

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