

Clinical Communications: Pediatrics

A PRESCHOOL-AGE CHILD WITH FIRST-TIME SEIZURE AND ATAXIA

Joan Papp, MD and Steven T. Dorsey, MD, FACEP

Department of Emergency Medicine, Cleveland Clinic, Cleveland, Ohio

Reprint Address: Steven T. Dorsey, MD, FACEP, Department of Emergency Medicine, E19, Cleveland Clinic, 9500 Euclid Avenue, Cleveland, OH 44195

Abstract—We present a case of a 4-year-old previously healthy child who had a possible first-time seizure at home, and upon a second Emergency Department evaluation was found to have gross cerebellar ataxia suggestive of acute stroke. Initial computed tomography scan and metabolic work-up were unrevealing. Subsequent neuroimaging demonstrated stroke in the left medulla and cerebellum secondary to left vertebral artery dissection. Cervical artery dissection may cause up to 20% of strokes in childhood and adolescence. Unlike typical adult presentations, antecedent or concurrent head and neck pain occurs less often in pediatric dissections. Symptoms of posterior circulation ischemia resulting from vertebral artery dissection may include vertigo, vomiting, ataxia, dysarthria, and seizure. Willingness to utilize newer, non-invasive imaging modalities may lead to earlier recognition of cervical artery dissection when patients have prodromal symptoms or episodes of transient ischemia. Vertebral artery dissection should be included in the differential diagnosis when evaluating children with first time seizure, headache, or neck pain. © 2009 Elsevier Inc.

Keywords—vertebral artery dissection; pediatrics; stroke; anticoagulation; ataxia

CASE REPORT

A 4-year-old child was brought to the Emergency Department (ED) by his mother with a chief complaint of “he keeps falling down.” Seven hours before presentation, she had been called to her son’s bedroom by a

sibling because the patient “could not move.” She found her son awake and lying supine on the floor, sluggish to respond to verbal and tactile stimuli. The brother reported that the patient had abruptly fallen down and had been making “choking sounds.” His mother took him to a local ED, where he was evaluated and discharged home with a diagnosis of seizure. His symptoms had seemed to resolve spontaneously by the time of that evaluation, but one hour before presentation to our ED, his mother noticed that he was falling to the left while ambulating, and seemed sleepier than usual. He also had vomited twice, and was hiccoughing frequently.

On review of systems, the patient had been generally well. The past medical history was significant for frequent otitis media, seasonal allergies managed with loratadine, and hospitalization at age 1 year for pneumonia. He was the product of an uncomplicated pregnancy, but required 5 days of phototherapy after birth for hyperbilirubinemia. The immunizations were up to date.

Vital signs on presentation were: oral temperature 36.9°C, pulse 96 beats/min, respiratory rate 20 breaths/min, and 97% saturation by pulse oximetry. General appearance, hydration status, and cardiorespiratory examination were all normal. On neurologic examination, the child was alert and oriented, and was watching cartoons. The cranial nerve examination was normal except for mild dysarthria. The muscle strength was normal. When asked to ambulate, he was grossly ataxic, began to fall, and had to be caught to prevent him from falling against the wall of the examination room.

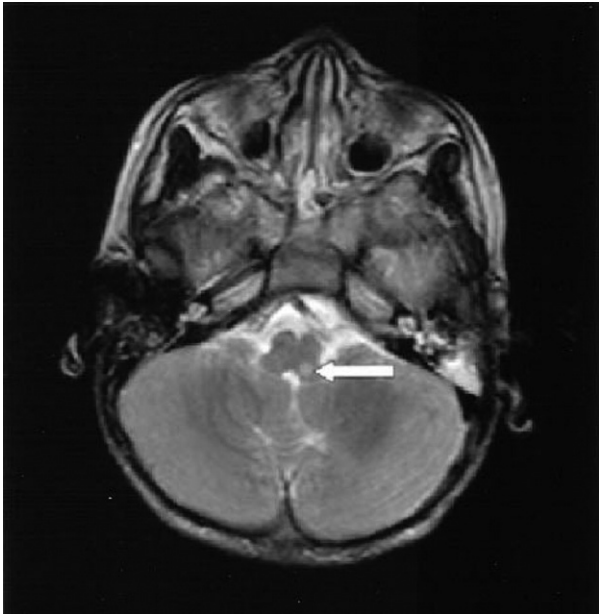


Figure 1. MR image with white arrow demonstrating stroke in the left medulla.

Electrolytes, complete blood count, and urine toxicology screen were normal. A non-contrast head computed tomography (CT) scan was interpreted as normal. The child was subsequently admitted to the general pediatrics service for neurology consultation and additional neuroimaging.

Upon admission to the pediatric ward, intravenous heparin was initiated. On hospital day 2, magnetic resonance imaging (MRI) demonstrated posterior inferior cerebellar artery (PICA) distribution stroke (Figure 1,) whereas magnetic resonance angiogram (MRA) demonstrated a cut-off of the left vertebral artery (Figure 2.) CT angiography performed on hospital day 3 confirmed left

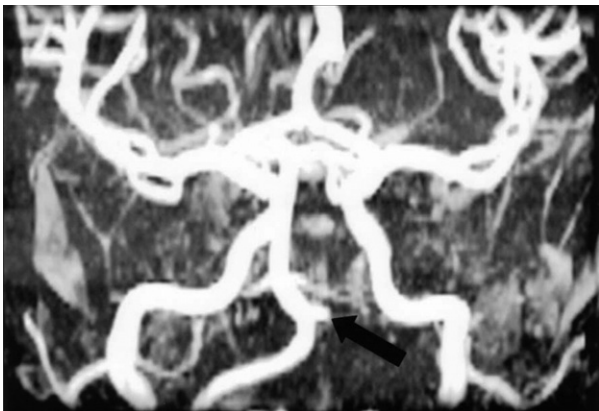


Figure 2. MRA reconstructed image with black arrow indicating terminus of occluded left vertebral artery, visible due to back-filling with gadolinium.



Figure 3. CT angiography image demonstrating similar anatomy to Figure 2. White arrow indicates right vertebral artery. Black arrow indicates terminus of otherwise occluded left vertebral artery.

vertebral artery occlusion secondary to vertebral artery dissection (VAD) (Figures 3, 4.) The patient's ataxia improved substantially over the next several days, and he was transitioned from unfractionated heparin to subcutaneous dalteparin. He was discharged home on levetiracetam for seizure prophylaxis. Other than subsequent ED visits for unrelated complaints, the patient has been lost to follow-up, but the ED records note that anticoagulants have been discontinued and the child's neurologic deficits have resolved.

DISCUSSION

Stroke is rare in childhood. Limited retrospective data suggest an incidence of approximately 2.5 to 3.1 cases



Figure 4. CT angiography image, coronal reconstruction. White arrows demonstrate right vertebral artery. Note absence of similar structure on the left side.

per 100,000 children per year (1). Whereas cervical artery dissection accounts for only 2% of stroke in adults, it may be responsible for up to 20% of stroke in childhood and adolescence (2). As in adults, carotid artery dissection is much more common than VAD (1,3,4). Pediatric cervical artery dissections of both varieties occur more often in boys than girls (1,3,5). This predilection has been attributed to a higher rate of traumatic injury in males, but actually persists when cases of trauma are excluded (5). Although most cases of VAD in children can be attributed to antecedent traumatic injury, many cases are labeled as spontaneous or idiopathic when they occur in a previously healthy child without underlying congenital defect or vasculopathy. Most dissections occur at the level of the first and second cervical vertebrae, an area of relative vulnerability due to rotation at the atlantoaxial junction and lack of bony protection afforded to lower segments, and thus many cases labeled

“spontaneous” may be due to insults so trivial that they are forgotten or go unnoticed (5).

Before uniting at the base of the pons to form the basilar artery, the vertebral arteries give off anterior and posterior spinal branches as well as the posterior inferior cerebellar arteries, which supply the medulla and portions of the cerebellum. The vertebral arteries also supply perforating vessels to the medulla (6). VAD causes ischemic symptoms either through limiting bloodflow to branch vessels or through resulting thromboembolic occlusion of distal branch vessels. Patients with VAD may present with a variety of symptoms including vertigo, profound nausea and vomiting, dysarthria, hemiparesis, ataxia, and seizure (4,5,7). A classic Wallenberg syndrome may occur, with loss of temperature and pain perception on the ipsilateral face and contralateral side of the body from the neck down, a unilateral Horner’s syndrome, dysarthria, and vertigo. Prodromal or concurrent occipital headache or ipsilateral neck pain may herald VAD, but occur in less than 50% of affected children; this is in contrast to VAD in adults, where pain occurs in the majority of cases (3–5,8).

Conventional angiography remains the gold standard for diagnosis of cervical artery dissection, and may demonstrate luminal stenosis, aneurysmal dilatation, or the most specific finding, an intimal flap (2,4). Newer non-invasive imaging modalities are now widely available, and are particularly preferable in children. MRI and MRA are most often employed, and were diagnostic in this case, but may miss lesions in the case of posterior circulation dissection (1,3,9). Hyperintensity in the vessel wall on axial MRI imaging, the crescentic hematoma sign, is a pathognomonic finding when corroborated with filling defects on MRA (4). CT angiography may be more readily available at major centers, and has the advantage of speed along with the ability to demonstrate precise anatomy through detailed three-dimensional reconstructed images. With regard to additional work-up beyond diagnosis of the culprit lesion, Ganesan et al. recommend complete cervical spine evaluation, as three patients in their series had hypermobility thought to have contributed to the VAD (3).

Management of VAD in pediatric patients is controversial. Due to the rarity of VAD in the pediatric population, there are very few data available for the practicing clinician regarding therapeutics; in fact, no randomized trial regarding management for acute stroke in children exists (10). Other than anecdotal reports, no investigative data exist on systemic thrombolytic therapy in the setting of pediatric ischemic stroke, and most children present beyond the accepted thrombolytic window (10). Because most ischemic events related to VAD are thought to be due to thromboembolic events, systemic anticoagulation is widely utilized; however, efficacy of anticoagulation

has not been prospectively validated (1,5). In Fullerton et al.'s meta-analysis of pediatric cervical artery dissection, none of the 14 VAD patients presenting before 1990 was anticoagulated, and only three patients were treated with an anti-platelet agent, whereas among 29 VAD patients who presented after 1990, one-third were treated with anticoagulants, one-quarter were treated with anti-platelet agents, and one-fifth of patients were treated with both (5). Mortality and recurrence of dissection were lower among patients receiving anticoagulation. Two of 29 patients who were anticoagulated had serious bleeding complications. Overall mortality from VAD was 2%, as opposed to 33% mortality in the group with anterior circulation dissection (5). In Ganesan et al.'s case series of posterior circulation stroke, three-quarters of the patients were anticoagulated, and over 50% of patients recovered without permanent deficit (3). Limited retrospective data suggest that low-molecular-weight heparins, as utilized in the case presented, seem to be as efficacious as unfractionated heparin in the setting of non-hemorrhagic stroke in children (11).

The Emergency Physician (EP) must include vascular dissection in his or her differential diagnosis when evaluating children with first-time seizure. Crucial components of the physical examination, particularly assessment of gait and coordination, must not be omitted. Although a healthy-appearing child with first-time afebrile seizure can be discharged home with close follow-up and additional outpatient testing, a persistent neurologic deficit requires further investigation (12). Acute frank ataxia is an infrequent finding in children, and the differential diagnosis should include VAD as well as acute intoxication or ingestion. The EP should also consider vascular dissection when evaluating a pediatric patient with a complaint of posterior headache or neck pain, even when there is no history of antecedent trauma. All 136 patients in the case series of Fullerton et al. and Schievink et al. had ischemic signs and symptoms at the time of diagnosis; this may reflect not only a low awareness of the disease

entity, but also a reluctance to obtain neuroimaging studies in afebrile pediatric patients with complaints of head or neck pain but no measurable neurologic deficit (5,7). As noted above, such symptoms are not as common in children as they are in adults with cervical artery dissection, but increased availability of non-invasive imaging modalities, and a more aggressive approach in obtaining them might allow for early diagnosis and treatment in select children with prodromal pain, and might prevent potentially debilitating stroke.

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