# Pediatric Homonymous Superior Quadrantanopia in the Presence of Acute Vertebral Artery Dissection

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**Objective:** Spontaneous dissection of the vertebral artery is a dangerous and often underdiagnosed cause of posterior circulation and occipital lobe stroke in the young. Vertebral artery dissection should be considered in young patients with a history of minor, nontraumatic changes in head position and visual disturbances.

**Methods:** Clinical records for this patient were retrospectively reviewed at Miami Children's Hospital and associated pediatric ophthalmology clinic.

**Results:** A previously healthy 8-year-old boy presented to the emergency department with a 2-day history of headache and blurred vision that developed after going on a high-velocity amusement park ride. Confrontation visual field testing showed a subtle superior visual field deficit superiorly. Computer topography scan of the head without contrast was performed and showed bilateral occipital hypodense lesions consistent with ischemic infarction. A filling defect was seen in the transverse portion of the right vertebral artery at the level of C2 as it exited the right transverse foramen, consistent with an acute arterial dissection at this level. Automated visual field testing showed macular sparing left homonymous superior quadrantanopia.

**Conclusions:** Most patients with vertebral artery dissection present with signs of posterior circulation ischemia consisting of neurological deficits, headaches, or neck pain. This case report highlights the unique clinical features and diagnostic imaging seen in the work-up of this rare disease process and exemplifies the importance of identifying acute visual symptoms in an emergency situation.

**Key Words:** vertebral artery dissection, quadrantanopia, nontraumatic dissection, pediatric dissection, left homonymous superior quadrantanopia

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**S** pontaneous dissection of the vertebral artery is a dangerous and often underdiagnosed cause of stroke in the young.<sup>1,2</sup> In these cases, the angiographic finding on autopsy analysis of vertebral artery dissections predominantly shows injury to the intima at the atlantoaxial joint. The dissection can allow for formation of a thrombotic nidus that can embolize to the posterior circulation and occipital lobe.<sup>3</sup> The occipital lobe is home to the primary visual cortex and posterior circulation ischemia to this area can present with a variety of visual disturbances.

While light enters the eye, it is received by retinal photoreceptor cells and the signal is sent to the brain via the optic nerve. This sensory information is projected posteriorly to the striate cortex of the primary visual cortex in the occipital lobe through the optic tract and optic radiations.<sup>4</sup> The optic radiations of the upper visual field course lateral through the temporal lobe and synapse inferior to the calcarine sulcus in the lingual gyrus of the striate

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The study was conducted at Miami Children's Hospital and Pediatric

Ophthalmology Consultants, 3100 SW, 62nd Ave, Miami, FL 33155. Copyright © 2015 Wolters Kluwer Health, Inc. All rights reserved. ISSN: 0749-5161 cortex.<sup>5</sup> Lesions to this area of the lingual gyrus give rise to loss of the contralateral upper quadrant of the visual field, a contralateral homonymous superior quadrantanopia. The collateral blood supply from the middle cerebral artery to the occipital pole allows for preservation of central vision and the macula.<sup>6</sup>

### METHODS

Clinical records for this patient were retrospectively reviewed at Miami Children's Hospital and associated pediatric ophthalmology clinic. Information obtained for this case report will be free of identifiable information. This study poses no direct risk to the patient and was performed in compliance with the United States Health Insurance Portability and Accountability Act.

#### RESULTS

A previously healthy 8-year-old boy presented to the emergency department with a 2-day history of headache and changes in vision that developed after going on a high-velocity amusement park ride. The patient described the headache as bifrontal that improved with over-the-counter antiinflammatory medications and was associated with 1 episode of vomiting. There was also 1 brief episode of alteration of awareness with complete return to baseline. Patient denied any motor, sensory, speech, or gait impairments during this time. The patient denied true blurred vision but described an inability to see to the left side. This problem was intermittent and not exacerbated with movements. There was occasional diplopia with no associated flashing lights, floaters, or shadows. Physical and neurological examinations were unremarkable with the exception of a confrontation visual field examination that showed left field deficit in both eyes tested separately.

A comprehensive eye examination was performed by a pediatric ophthalmologist. The patient's visual acuity was 20/20 in each eye uncorrected for distance and for near. Stereopsis was appropriate for age and color vision was intact. Pupils were 5 mm reactive with no afferent papillary defect. There was no ptosis or external eye pathology. Motility showed orthophoria at distance and near with full versions and ductions and no manifest nystagmus. Slit-lamp examination was unremarkable with white conjunctivae, clear corneas, deep and clear chambers, clear lenses, and clear vitreous bilaterally. Fundus examination was unremarkable with direct and indirect ophthalmoscopy showing sharp disks, pink rims, small cups, normal vessels, normal macula, and normal periphery bilateral.

Confrontation field testing showed a relative, subtle deficit superiorly to finger counting in each eye tested individually. Formal automated visual fields testing was performed with computerized Humphrey 24-2 Visual Field Analyzer (Fig. 1). The results showed a congruous loss of the left upper quadrant visual field in both eyes with macular sparing, a left homonymous superior quadrantanopia. There was also a subtle incongruous central field deficit on the right in both eyes. The patient cooperated very well and the study was considered reliable.

Computed topography (CT) scan of the head without contrast was performed and showed bilateral occipital hypodense lesions consistent with ischemic infarction (Fig. 2). This study

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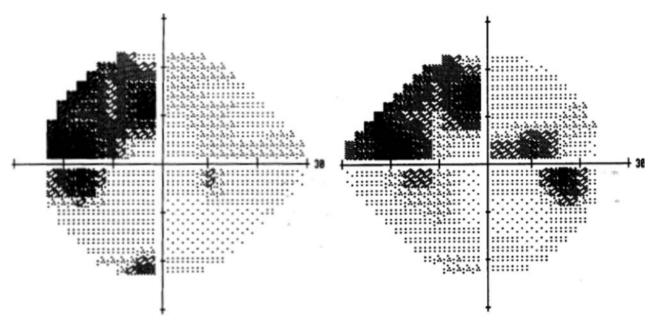


FIGURE 1. Formal automated visual fields were tested with computerized Humphrey 24-2 Visual Field Analyzer. Results showed a congruous loss of the left upper quadrant visual field in both eyes with macular sparing, a left homonymous superior quadrantanopia. There was also a subtle incongruous central field deficit on the right in both eyes.

was followed with magnetic resonance imaging (Fig. 3) and arteriography, which showed restricted diffusion in medial, posterior, and inferior aspects of both occipital lobes. Similar lesions were seen in the thalami and inferior aspect of the right cerebellar hemisphere. No gross vertebral, basilar, or posterior cerebral artery dissections were seen in this study. Magnetic resonance venography showed no evidence of obstruction or thrombosis. Follow-up CT angiography of the brain and neck was performed. Filling defect was seen in the transverse portion of the right vertebral artery at the level of C2 as it exits the right transverse foramen, consistent with subtle dissection at this level (Fig. 4). This image was not seen on previous studies because of significant brace susceptibility artifact. A 21-channel electroencephalogram was also performed and its findings were reported as normal.

Diagnosis of occipital ischemic lesions due to a dissected right vertebral artery embolus was made. Hematologist recommendation of enoxaparin sodium (Lovenox) 1 mg/kg every 12 hours for 3 to 6 months at a therapeutic level of 0.5 to 1.0 IU/mL was initiated. A 3-month follow-up CT angiogram showed similar filling defect in the transverse portions of the right vertebral artery.

#### DISCUSSION

This case highlights the clinical presentation and associated images of a rare cause of cerebral vascular accident in the pediatric population. Most patients with vertebral artery dissection present with signs of posterior circulation ischemia consisting of neurological deficits, headaches, or neck pain. The use of supplemental radiological imaging and ophthalmologic work-up is a valuable tool to confirm clinical suspicion in this rare disease process. In this situation, ophthalmic work-up would be without any gross red flags and would show only a subtle superior defect in visual fields. Formal automated testing highlights the precise loss of the left upper quadrant visual field with macular sparing. By understanding the visual field anatomy, the location of the lesion can be identified as the contralateral occipital lobe. The use of

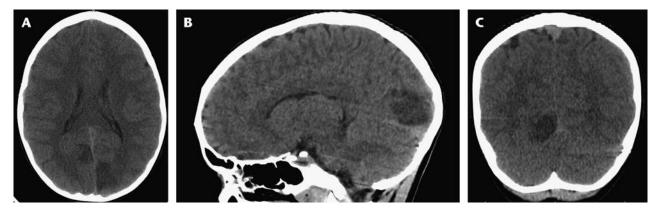


FIGURE 2. Computed tomography scan of the head without contrast showing bilateral occipital hypodense lesions consistent with ischemic infarction shown in axial (A), sagittal (B), and coronal (C) views.

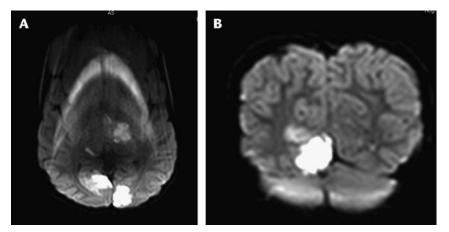


FIGURE 3. Diffusion weighted magnetic resonance imaging showing posterior circulation infarcts to the occipital lobe shown in axial (A) and coronal (B) views.



FIGURE 4. Computed tomography angiography of the brain and neck was performed. Filling defect was seen in the transverse portion of the right vertebral artery at the level of C2 as it exits the right transverse foramen, consistent with subtle dissection shown in axial (A) and coronal (B) views.

confirmatory radiological imaging outlines the posterior circulation anatomy and the unique pathology of the case.

This case also exemplifies the importance of identifying acute visual symptoms in an emergency situation. Patients and doctors may quickly overlook the twists and turns on an amusement park ride as a possible cause of a traumatic dissection. Similarly, a vague complaint of "blurry vision" from a young child can easily go without serious evaluation. The medical literature has shown strokes from vertebral artery dissection occurring from minor, nontraumatic changes in head position in patients of all ages. Often, these are due to abrupt head turning and can frequently be caused by minor automobile accidents or chiropractic manipulation. The clinical presentation of vertebral artery dissection can also be highly variable, presenting with signs of focal deficits, ischemia, or both.<sup>7</sup> Most patients present with signs of posterior circulation ischemia consisting of neurological deficits, headaches, and/or neck pain.<sup>8–10</sup>

This case also discusses 1 treatment option for acute vertebral artery dissection. In children with extracranial craniocervical arterial dissection, anticoagulation is generally recommended, yet there are no randomized controlled trials to compare treatments. In this case, enoxaparin sodium (Lovenox) was initiated at 1 mg/kg every 12 hours for 3 to 6 months at a therapeutic level of 0.5 to 1.0 IU/mL. Consideration of inpatient intravenous unfractionated heparin can also be used as a bridge to long-term oral anticoagulation.<sup>11,12</sup>

## CONCLUSIONS

Vertebral artery dissection should be considered in young patients with a history of minor, nontraumatic changes in head position and visual disturbances. This case report highlights the unique clinical features and diagnostic imaging seen in the work-up of this rare disease process.

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