Bartonella presenting as branch retinal artery occlusion in a child

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Branch retinal artery occlusion (BRAO) is rare in children. Bartonella is a known cause of branch retinal artery occlusion in adults, but it is typically not considered in the differential diagnosis for pediatric BRAO. We present the case of a 12-year old boy with a BRAO caused by a *Bartonella henselae* infection. This is the youngest such case reported in the literature. Although rare, Bartonella infection may be an important and underrecognized cause of pediatric BRAO.

Case Report

A 12-year-old white boy presented at Casey Eye Institute at Oregon Health & Science University with acute, painless onset of a grayish-black line in the inferonasal portion of the visual field of his right eye 2 weeks prior. He denied photopsias, floaters, or dark curtain in his vision. Review of systems revealed that 1 week prior to the onset of his visual symptoms he had experienced fevers with intermittent bilateral knee pain, which spontaneously resolved. He had no associated cough or cold symptoms, and review of systems was otherwise negative. He was previously healthy, was not taking medications, had no history of migraines, and had no relevant family history of eye conditions. Social history was notable for several pets at home, including 2 dogs, 2 cats, and 2 guinea pigs.

On examination, his uncorrected visual acuity was 20/25-2 (pinhole, 20/20-1) in the right eye and 20/20-2 in the left eye. Confrontational visual fields demonstrated an inferonasal field defect in the right eye only. Color vision and formal visual fields were not tested. He was also found to have a 1 + relative afferent pupillary defect in the right eye. He had normal stereovision, alignment, motility, intraocular pressure, cycloplegic refraction, and anterior segment examination.

Dilated fundus examination of the right eye was significant for a large, superior cotton wool spot, with inner retinal fovea-sparing whitening along the superior arcade (Figure 1). This was associated with superior optic disk edema, a single, small targetoid chorioretinal lesion superonasal to the optic nerve, and several punctate intraretinal yellow deposits throughout the temporal macula thought to be partial macular star exudates. The left eye was significant for 2 distinct, small round targetoid chorioretinal lesions in the nasal midperiphery and similar punctate intraretinal yellow deposits in the macula. Fundus examination of the left eye was unremarkable.

Fluorescein angiography revealed a superotemporal branch retinal artery occlusion (BRAO) in the right eye, with localized vascular leakage along the superior arcade with staining of the nasal chorioretinal lesion and mild optic disk leakage. Optical coherence tomography of the right eye confirmed retinal nerve fiber layer edema in the location of the large cotton wool spot and early inner retinal atrophy, with hyperreflectivity and blurring of the inner retinal laminations in the distribution of the superotemporal BRAO (Figure 2).

The patient was diagnosed with bilateral acute neuroretinitis, multifocal chorioretinitis, with a BRAO in the right eye. Workup included complete blood count, comprehensive metabolic panel, urinalysis, quantiFERON Gold, FTA, RPR, ANA, and serology for toxoplasma, toxocara, Lyme disease, HIV, and *Bartonella henselae*. Given that this BRAO was accompanied by neuroretinitis and multifocal chorioretinitis without visible embolus, we did not feel that an extensive hypercoagulability workup was indicated. Bartonella IgG and IgM results were positive, with titers of 1:1024 and 1:32, respectively. The remainder of his workup was unremarkable. The patient confirmed that one of his cats was 9 months old and frequently scratched and bit him. The patient was treated with oral doxycycline 100 mg twice daily for 21 days and a tapering course of oral prednisone at 30 mg daily starting 3 days after initiating doxycycline. We recommended that both cats be tested for Bartonella and, if positive, that they be removed from the household or treated. Both household cats tested positive for Bartonella by serology (1:128 for both cats) and blood culture and were treated.

At 3 months' follow-up, the patient retained lack of perfusion in the distribution of the BRAO, the macular star exudates had resolved, and the chorioretinal lesions appeared atrophic. Visual acuity remained unchanged.

Discussion

BRAO in children is exceedingly rare, with an estimated incidence of <1 per 50,000 outpatients <30 years of age.[1] Most reported cases of BRAO in children have been deemed idiopathic.[1-3] The differential diagnosis for BRAO in a child typically includes hypercoagulable states (sickle cell, increased factor VIII, Fabry disease, homocystinemia), cardiac defects (dextrocardia, patent foramen ovale, atrial myxoma), and trauma. Less commonly reported etiologies are Susac's

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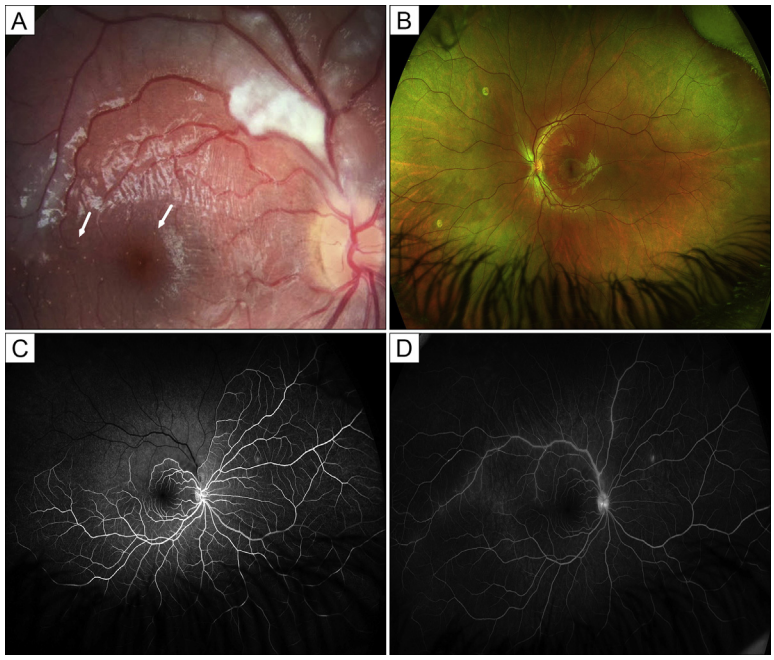

FIG 1. A, Fundus photograph of the right eye showing a superior branch retinal artery occlusion with large cotton wool spot, superior optic disk edema, and punctate intraretinal yellow deposits (hard exudates, arrows) in the macula. B, Fundus photography of the left eye showing multifocal targetoid chorioretinal lesions. C-D, Fluorescein angiography of the right eye revealing delayed filling of a superotemporal branch arteriole with concomitant delayed venular filling designating a branch retinal artery occlusion, late leakage of the superior optic disk, and a hyperfluorescent targetoid lesion in the superonasal midperiphery.

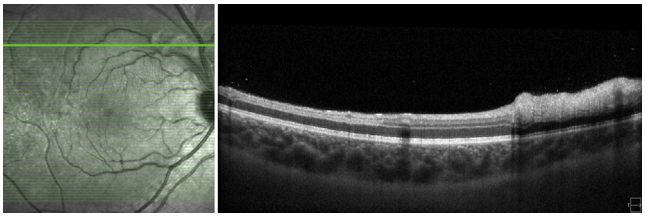

FIG 2. Optical coherence tomography of the right eye showing a large cotton wool spot with corresponding nerve fiber layer edema in the superonasal macula and early inner retinal atrophy in the superotemporal macula in the distribution of the superotemporal arteriole.

disease, posterior uveitides (toxoplasmosis, tuberculosis, West Nile virus, and Lyme disease), and even prolonged carotid compression from a cell phone.[4,5]

The global annual incidence of cat scratch disease is estimated at 4.5 per 100,000 population, with ocular involvement occurring in 5%-10% of patients.[6] The most common presentation of ocular bartonellosis is Parinaud oculoglandular syndrome. Posterior segment manifestations include neuroretinitis, localized or multifocal chorioretinitis, and vasculitis. BRAO is a known manifestation of ocular Bartonella in adults, having been reported in one series of 6 cases[7] as well as individual case reports.[8-10] However, only once has this finding been reported in a child, and this was in a 15-year-old girl with neuroretinitis.[11] Our case is novel for two reasons. First, this is the youngest patient to have been reported in the literature to suffer a BRAO secondary to Bartonella. Second, while the partial macular stars, small afferent pupillary defect, and subtle disk leakage were indicators of neuroretinitis, the BRAO was the most striking finding in this patient 2 weeks after onset of symptoms. Given that optic disk edema and macular star exudates can resolve spontaneously, while a complicating BRAO could persist, we feel that it is important for pediatric ophthalmologists to recognize Bartonella as a potential cause of branch retinal artery occlusion in a child, particularly because actively infected pets could cause recurrent exposure. Of note, the "idiopathic" cases of pediatric BRAO reported to date have not included Bartonella as part of their workup, and thus it is possible that this condition has been underdiagnosed and underreported to date.[1-3] We recommend that a social history is obtained during evaluation of a pediatric BRAO, and if corroborative for cat ownership or interaction, Bartonella titers should be tested.

Management of ocular bartonellosis remains controversial. Immunocompromised individuals often require systemic antibiotics to control their infection, whereas infection in immunocompetent patients is often self-limited, and treatment is not necessarily warranted. Given the severity of disease and location of vascular occlusion in this patient, we elected to treat. Guidelines for management of the feline culprit of ocular Bartonella do

not exist, but in this particular case of possibly high reexposure potential, we decided recommend cat testing and treatment.

Literature Search

PubMed was searched on April 1, 2018, without date or language restriction, using the following terms: *pediatric retinal artery occlusion* AND *bartonella retinal artery occlusion*.

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