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RESEARCH ARTICLE

CENTRAL SCOTOMA IN A YOUNG ADULT MALE WITH FLU LIKE SYMPTOMS

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INTRODUCTION

Systemic Bartonella henselae infection, commonly known as Cat Scratch Disease is a rare zoonotic infection occurring at a rate of around 15,000 cases per year in the United States.^[1] Ocular manifestations of cat scratch disease most commonly either Parinauds Ocular-glandular involve Syndrome, consisting of lymphadenopathy and follicular conjunctivitis, or neuroretinitis. The latter most often involves lymphadenopathy and granulomatous inflammation of the retina and optic nerve. Other sequelae of Cat Scratch Disease are rarely reported and can include branch retinal artery occlusions as reported here.

Case Report: A 35-year-old male with no significant past medical history presented to the emergency department reporting an acute onset of a gray blurry spot in his central vision in the right eye for 2 days duration. He reported a prior history of headache, intermittent fevers, chills and myalgia for the past few weeks and he was treated with Augmentin for an enlarged inguinal lymph node during this period. At the time of presentation, he was asymptomatic and denied any other medical issues. On exam his Snellen visual acuity was 20/20 OD when looking up and 20/25 OS. Intraocular pressure was 12 mm Hg OD and 15 mm Hg OS.

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ABSTRACT

Purpose: To briefly discuss a young, otherwise healthy Caucasian male who presented to an emergency department with the complaint of acute scotoma. Observations: External ocular and anterior segment examination were unremarkable. A dilated fundus examination of the right eye showed a wedge-shaped area of retinal whitening and cotton wool spots in the inferior macula between the optic nerve head and fovea. Conclusions and Importance: After a thorough work up including a limited coagulopathy screening and rheumatologic and infectious panels, it was determined the most likely cause of presentation was systemic Bartonella infection contracted from the patient's pet cat. The patient was treated with oral doxycycline 100 mg BID for 4 weeks and maintained stable vision on subsequent follow ups with no recurrent arterial occlusions. We hope this report provides clinicians with an insight into an uncommon ophthalmologic presentation of Bartonella henselae infection, as well as a reasonable template for successful treatment if such a case is encountered.

> Other than blurry central vision OD, visual field was grossly normal in both eyes. Pupils were round and reactive without afferent pupillary defect. External ocular and anterior segment examination were unremarkable. A dilated fundus examination of the right eye showed a wedge-shaped area of retinal whitening and cotton wool spots in the inferior macula between the optic nerve head and fovea as well as a small white retinal lesion located in peripheral temporal retina. There was no evidence of heme or Hollenhorst plaques on biomicroscopy. (Figure 1A) Examination of the left eye revealed a small white lesion in the supero-nasal quadrant. There was no evidence of vasculitis, vitritis or optic nerve edema. Fluorescein angiography of the right eye demonstrated vascular occlusion and blockage of fluorescein signal corresponding to the area of retinal pallor and edema consistent with branch arterial occlusion. (Figure 1B) The left eye demonstrated late leakage at the optic nerve without vascular pathology. OCT imaging of the lesion initially demonstrated inner retinal thickening (Figure 2A); this same area demonstrated retinal atrophy on repeat OCT six months later. (Figure 2B.) The patient was treated with oral doxycycline 100 mg twice daily for 4 weeks. At follow up exams, the patient maintained his baseline vision with stable central scotoma, without recurrence of new episodes of arterial occlusion. There was no evidence of macular scar on follow up.

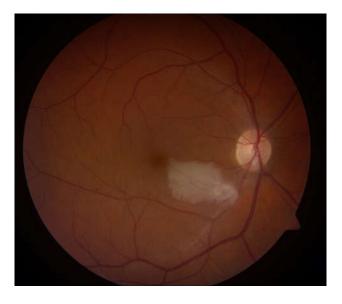


Figure 1A. Fundus photograph demonstrating retinal whitening and edema in inferior macula

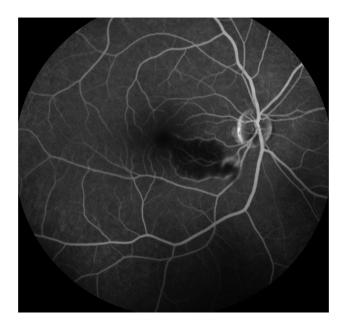


Figure1B. Mid phase fluorescein angiogram demonstrating branch retinal arterial occlusion and edema

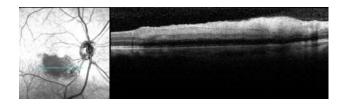


Figure 2A. SD-OCT through the macular lesion showing inner retinal thickening in the acute phase

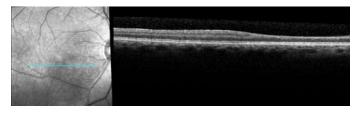


Figure 2B. SD-OCT through the same lesion 6 months after onset showing retinal atrophy

DISCUSSION

Retinal artery occlusion (RAO) in elderly patients is most commonly caused by embolic events; however, in young adults, it is extremely uncommon and usually has a multifactorial etiology including autoimmune diseases, infectious etiologies, cardiovascular disorders or coagulopathies. The patient had undergone a recent evaluation by his primary care provider and his routine test results were reported as normal. Several options for proceeding could be argued, including observation, systemic evaluation for risk factors such as coagulopathies and embolic sources or vascular imaging such as CTA head and neck. Based on the acute nature of his visual complaints, recent history of flu like symptoms including headache, fevers, chills and myalgia in addition to his exposure to cats and recent cat scratch, it was decided to initiate a systemic work up for inflammatory and infectious causes of RAO. Extensive systemic work up was positive only for IgM and IgG titers for Bartonella henselae, hence the retinal findings were attributed to the patient's systemic Bartonella infection. An inflammatory work up including ESR, CRP, ANA, ANCA, double stranded DNA, JO-1, SSA/B, Smith antibodies, SCL-70, RNP antibodies and histone antibodies did reveal mildly elevated ESR of 26 consistent with systemic inflammation, however specific autoimmune testing was largely negative. Given the lack of a past rheumatologic personal and family history, this finding was considered non-significant in the present case. In similar cases where there is a questionable personal or family history of coagulopathy or concern for cardiac source of embolus, a more in-depth investigation into these matters may be appropriate. Measures such as echocardiogram for valvular vegetations and comprehensive hematologic panel for coagulopathy could be pursued. Due to the history and physical exam findings in this case, only a brief hematologic work up including cryoglobulin and anti-thrombin III was performed.

Conclusion

Bartonella henselae is a gram-negative bacterium and is the cause of cat scratch disease. The classic presentation of cat scratch disease involves either Parinauds Ocular-glandular Syndrome, which consists of lymphadenopathy and follicular conjunctivitis, or neuroretinitis, consisting of lymphadenopathy and granulomatous inflammation of the retina and optic nerve.^[1,2] Classic fundus findings in Bartonella neuroretinitis include optic nerve edema, macular star (indicative of neuroretinitis) and possibly signs of vasculitis.^[3] Bartonella can invade vascular endothelial cells and lead to retinal vascular thrombosis and occlusion [4] or even vasoproliferation.^[5] Cat scratch disease is a self-limiting entity and systemic treatment has not shown to change the course of the disease.^[6] However, in immunocompromised patients, in children, and in cases with severe ocular manifestations, medical intervention is recommended.^[6] In patients with RAO, treatment may minimize the chance of further vascular occlusive episodes. Doxycycline is the treatment of choice since it has good ocular availability. Healthy individuals need 2 to 4 weeks and immunodeficient patients need 4 months of antibiotic therapy^{[7].} In contrast to older patients, RAO in young and mostly healthy adults is a rare entity and has a broad differential diagnosis. Relevant history taking and having a high index of suspicion for the infectious causes of RAO is essential in successful diagnosis and management.

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Glossary of Abbreviations

CTA: computed tomography angiogram OCT: Optical coherence tomography OD: ocular dexter (right eye) OS: ocular sinister (left eye) RAO: retinal arterial occlusion SD-OCT: spectral domain optical coherence tomography

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