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Macular Star and Central Visual Loss: Two Pediatric Cases

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Case 1

A 14-year-old girl with a past medical history of left eye amblyopia, presented to an ophthalmologist with 4 days of worsening central vision in her right eye. She reported exposure to 6 household cats, but denied any recent trauma, including scratches. She had no fever, and no eye pain. Best corrected visual acuity was “counting fingers” at a distance of 4 feet with her right eye, and 20/30 in her left eye. Her right eye revealed 2+ optic disc edema and a macular star (Figure 1A). She had no palpable preauricular or submandibular lymph nodes.

Antibody titers against *Bartonella quintana* and *Borrelia burgdorferi* were negative. Pertinent negative laboratory evaluations from blood included polymerase chain reaction (PCR) assays for herpes simplex virus (HSV), varicella zoster virus (VZV), Epstein-Barr virus (EBV), toxoplasmosis, and cytomegalovirus (CMV). She also had a negative tuberculin skin test, a nonreactive venereal disease research laboratory test (VDRL), and a normal serum angiotensin-converting enzyme (ACE) level. Laboratory evaluation revealed an elevated *Bartonella henselae* immunoglobulin M (IgM) antibody titer of 1:32 (normal <1:16), and a positive immunoglobulin G (IgG) titer of 1:512 (normal <1:64).

Hospital Course

The patient was treated with 4 weeks of twice daily doxycycline (100 mg) and rifampin (300 mg). One month after treatment, she had significant improvement in the visual acuity of her right eye, from “counting fingers” on presentation to 20/60 at first follow-up. Six months after the initial presentation, she demonstrated continued improvement with a right eye visual acuity of 20/30, and further resolution of the macular star (Figure 1B).

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Author Contributions

TEN and CRO wrote the first draft of the manuscript. RLL and WAA contributed to acquisition and interpretation of the data. All authors critically revised and gave final approval of the manuscript.

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Case 2

A 17-year-old girl presented with a complaint of an enlarging gray spot in her right central visual field of 2 weeks' duration. A few days prior to the change in her vision, she had 2 days of gradually resolving generalized muscle pains and headaches, followed by 3 days of pain with right eye movement. She also reported 2 weeks of intermittent fever and chills. Although she had a kitten with fleas, she could not remember being scratched or bitten.

Eye examination by an ophthalmologist revealed that her visual acuity was 6/200 in the right eye and 20/20 in the left with a right afferent pupillary defect. There was diffuse temporal visual field loss in the right eye. Dilated fundus examination photograph showed 4+ disc edema with inferotemporal hemorrhage and a combination of exudative and vascular changes. She had a macula star with 3+ macular edema and tortuous vessels (Figure 2A). The left eye examination was normal. Pertinent negative laboratory test results included an antibody screen for Lyme disease, and a PCR of blood for VZV. Laboratory evaluation revealed an elevated IgG antibody titer against *Bartonella henselae* of 1:256 (normal <1:64).

Hospital Course

She was treated with twice daily doxycycline (100 mg) and rifampin (300 mg) for 2 weeks. At a one-month follow-up visit, her vision had improved to 20/100 in her right eye; the disc edema had decreased dramatically, and the visual field had improved. When she was seen 9 months after the onset of disease, her visual acuity had improved to 20/40. Although an afferent pupillary defect remained, the fundoscopic examination showed improved results (Figure 2B).

Final Diagnosis

Neuroretinitis secondary to cat scratch disease.

Discussion

Cat scratch disease (CSD), first described by Parinaud in 1889, usually presents with an influenza-like illness and lymphadenopathy.¹ CSD is caused by the Gram-negative bacillus, *Bartonella henselae*, and is thought to be the result of bacterial inoculation from cat scratches and bites, or bites from cat fleas.² Neuroretinitis, a condition characterized by abrupt visual loss, disc edema, and a macular star was first attributed to *B henselae* infection by Sweeney in 1970.^{3,4} Neuroretinitis and other optic neuropathies may occur in a small percentage of patients with CSD,⁵ a condition that presents a significant risk to vision. The largest study to date found that only 55% (29/53) of eyes improved by at least 3 lines on the Snellen chart at the time of final follow-up.³ This study also revealed that only 68% (36/53) of eyes achieved a final visual acuity of 20/40 or better, and that 6% (3/53) of the eyes had acuities of 20/200 or worse by the end of follow-up.³ The pathogenesis of the disease is not completely understood, but it is thought to arise from focally increased vascular permeability. The classical macular star of CSD occurs in the context of improving macular edema; as leaked serum is resorbed, lipid exudates precipitate in a stellate pattern.⁶ Nevertheless, decreasing disc edema is not always followed by macular star formation.

The diagnosis of neuroretinitis is initially based on clinical findings, including optic disc swelling and either a macular star or widespread retinal exudates.⁷ CSD is likely responsible for a majority of cases of neuroretinitis, especially in the context of cat exposure and supporting associated signs and symptoms.¹ However, a subset of patients with acute ocular complaints fail to provide a history suggestive of cat exposure, yet have strong serologic evidence of *B henselae* infection.³ One of the 2 patients presented in this report had neuroretinitis without the classic lymphadenopathy. Lymphadenopathy was once considered to be 1 of 4 diagnostic criteria for CSD,⁷ yet recent data suggest that CSD, particularly neuroretinitis, should be suspected even in the absence of lymphadenopathy. Serologic testing should be done to increase diagnostic accuracy. The sensitivity of immunofluorescence assays for antibody detection is roughly 88%.⁸ The serologic test is considered positive if, in the appropriate clinical setting, seroconversion occurs or if either IgM or IgG antibody titers against *B henselae* are significantly elevated. Sometimes an elevated IgG antibody titer alone can be used as a marker of acute *B henselae* infection because the presence of IgG antibody tends to be transient. Roughly 75% of IgG-seropositive patients will be seronegative 1 year after disease onset.⁸ Documentation of seroconversion is not always possible in patients with *B henselae* neuroretinitis, because most patients already have IgG antibodies when visual symptoms begin.

To date there has been no consensus on an effective treatment regimen for *B henselae* neuroretinitis. There have been no randomized controlled trials comparing different antibiotic regimens, and there is no certainty as to which patients, if any, are likely to benefit from such therapy. Although *B henselae* infection is commonly self-limited and the majority of patients have positive outcomes even in the absence of treatment, antibiotics are generally recommended for severe systemic and intraocular infections.^{1,2,7,9} Recommended therapy varies from 2 to 4 weeks of doxycycline or erythromycin, with or without adjuvant rifampin.^{3,7} These recommendations are based on the effectiveness of these antibiotics as demonstrated in immunocompromised HIV-positive individuals with *B henselae* infection.⁷ While one of our patients received 4 weeks of treatment with doxycycline and rifampin, and the other received only 2 weeks of the same antibiotic regimen, both had favorable outcomes, with near complete resolution of their visual deficits.

Conclusion

These cases illustrate the clinical presentation, diagnosis, and treatment of this unusual infectious disease. Additionally, it highlights the need for further research regarding antibiotic therapy for CSD neuroretinitis, especially in children.

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Figure 1.

Case 1: *Bartonella henselae* neuroretinitis. Fundal photographs of right eye. (A) Acute presentation demonstrating disc edema (red arrow) and macular star (blue arrow). (B) Photograph 5 months after presentation, demonstrating significantly reduced exudates in the macula but with disc pallor.

Figure 2.

Case 2: Bartonella henselae neuroretinitis. Fundal photographs of right eye. (A) Acute presentation demonstrating disc edema and macular star (blue arrow). (B) Nine months following presentation, demonstrating significantly decreased disc edema and minimal macular exudates.