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Rickettsial neuroretinitis: A report of 2 cases

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ABSTRACT

Purpose: The authors present two cases of neuroretinitis caused by *Rickettsia rickettsii* infection.
Observations: Case 1 is a 24-year-old male who presented with 2 months of vision loss. Case 2 is a 38-year-old female who presented with 4 weeks of eye pain and vision loss. Examination of both patients revealed neuroretinitis characterized by optic disc swelling with macular exudates, and subsequent serological analysis was positive for *Rickettsia rickettsii*. Both patients responded favorably to treatment with oral doxycycline and prednisone.
Conclusions and importance: Given the potential for neuroretinitis to cause permanent vision loss, the presence of acute vision loss, optic disc edema, and macular exudates should prompt an evaluation for Rickettsial disease in endemic areas, even in the absence of systemic symptoms or known history of a tick bite.

1. Introduction

Neuroretinitis is an inflammatory disorder of the retina and optic nerve characterized by acute vision loss, optic disc edema, and macular exudates, which occasionally form a macular star. It may be infectious, inflammatory, or idiopathic in etiology.[1,2] The visual prognosis is generally excellent, although permanent vision loss is possible and early treatment with steroids and antibiotics in some cases may limit disease progression and improve long-term outcomes.[3] Neuroretinitis caused by infection with the bacterial species *Rickettsia rickettsii* has rarely been reported in the literature. We report the cases of two patients with significant vision loss from neuroretinitis caused by *Rickettsia rickettsii* infection who had significant recovery of vision after treatment with antibiotics and oral steroids.

2. Case report

2.1. Case 1

A 24-year-old male with no significant past medical history presented with a 2-month history of decreased vision in both eyes. Examination showed best corrected visual acuity (BCVA) of 20/200 in each eye with a normal intraocular pressure (IOP), normal anterior segment exam, and no vitreous cell. Funduscopic examination of both eyes revealed optic disc swelling, disc hemorrhages, peripapillary subretinal fluid and hard exudates in the macula (Fig. 1). Fundus autofluorescence revealed peripapillary hyperautofluorescence in the area of serous retinal detachment. Optical coherence tomography (OCT) revealed disc elevation, peripapillary subretinal fluid and hyper-reflective foci in the retina corresponding to the hard exudates. Upon further questioning, the patient reported discovering a tick bite 2 weeks prior to the onset of his visual symptoms. He denied any rash, fever, or myalgias.

Chest x-ray was negative. Laboratory tests were negative for syphilis (RPR and FTA-ABS), tuberculosis (Quantiferon TB Gold), Lyme, Ehrlichiosis, and Bartonella. However, titers for *Rickettsia rickettsii* IgM and IgG were both elevated. He was treated with a two-week course of doxycycline and a prednisone taper (starting at a dose of 60 mg daily) for Rickettsial neuroretinitis. After 3 months, his visual acuity improved to 20/25 in the right eye (OD) and 20/60 in the left eye (OS). He had been completely tapered off prednisone, and the optic disc swelling and peripapillary subretinal fluid had resolved. There was, however, mild disc pallor evident in both eyes and outer retinal loss on OCT in the nasal macula.

2.2. Case 2

A 38-year-old female with no significant past medical history presented with 4 weeks of eye pain and blurry vision. Her visual acuity was 20/40 OD and 20/400 OS. Examination of her right eye revealed normal IOP, quiet anterior chamber, and 1+ vitreous cells. Examination of her

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left eye revealed normal IOP, 0.5+ cells in the anterior chamber, and 2+ vitreous cells. There was mild optic disc swelling in the right eye and moderate disc swelling with macular star formation in the left eye (Fig. 2). Fluorescein angiography of both eyes revealed hyperfluorescence with leakage at the disc and mild peripheral vascular leakage (Fig. 3). Upon further questioning, she reported a recent history of myalgias and intermittent fevers. She denied any history of a tick bite.

Chest x-ray was negative. Laboratory tests were negative for syphilis (RPR and FTA-ABS), tuberculosis (Quantiferon TB Gold), Lyme, Ehrlichiosis, and Bartonella. *Rickettsia ricketsii* IgG was negative but *R. rickettsii* IgM was positive, indicating a current or recent infection. Treatment was initiated with a two-week course of doxycycline and a prednisone taper starting at 40 mg daily. Four weeks later, repeat testing for *R. ricketsii* IgG was positive. Three months after initiating treatment, her visual acuity had improved to 20/20 OD and 20/25 OS with resolution of anterior chamber cell, vitreous cell, and disc swelling. The patient was completely off of oral steroids at the 3-month point, without any recurrence of inflammation.

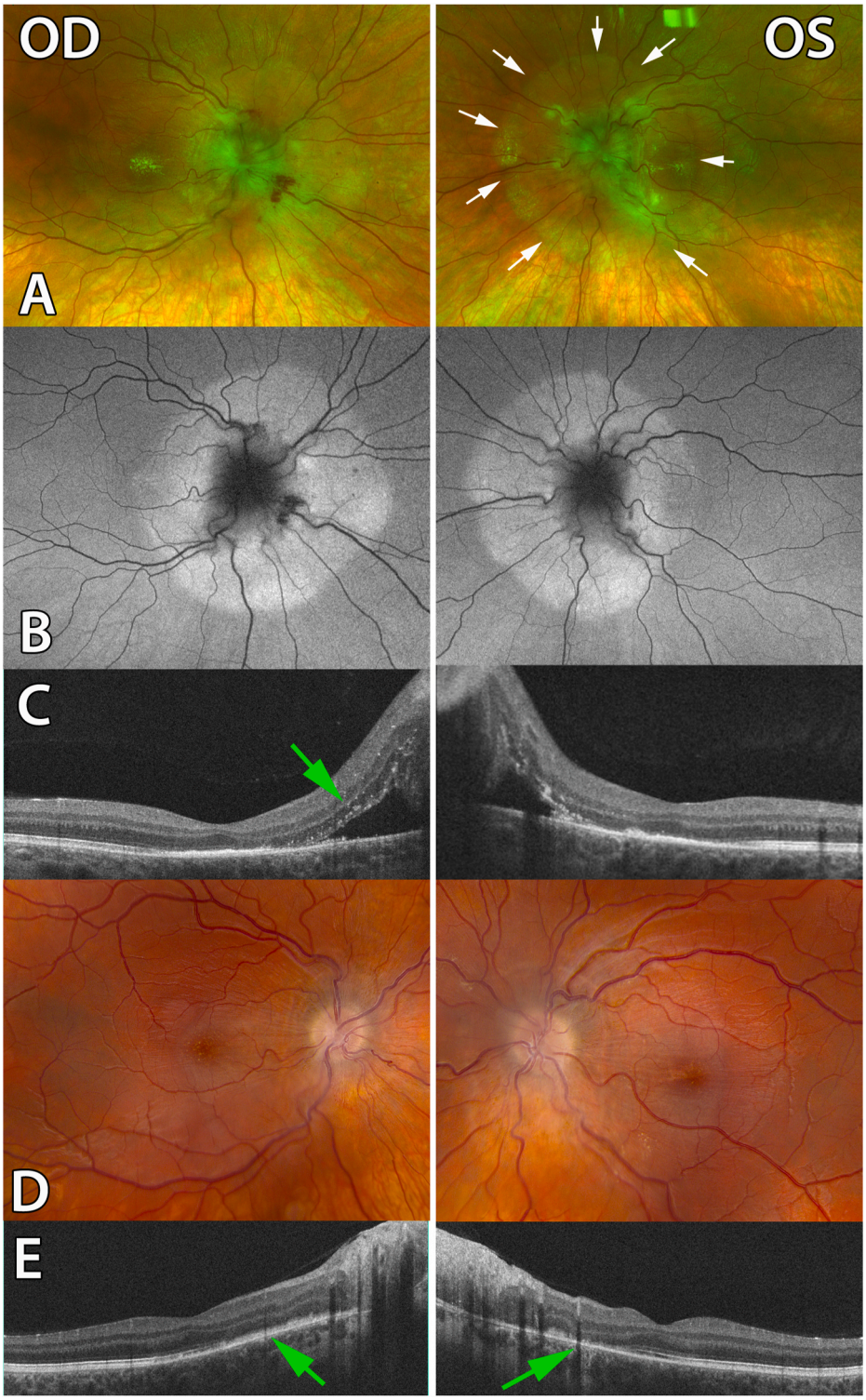

Fig. 1. Multimodal imaging of case 1. (A) Baseline fundus photos of the right (OD) and left (OS) eyes showing disc swelling, peripapillary subretinal fluid and macular exudates in both eyes (OU). The margins of area of exudative retinal detachment are evident (white arrows). Disc hemorrhages are seen OD. (B) Baseline fundus autofluorescence revealed peripapillary hyperautofluorescence in the area of exudative retinal detachment OU. (C) Baseline macular optical coherence tomography (OCT) shows nasal subretinal fluid, outer retinal irregularity and hyper-reflective foci (green arrow). (D) Fundus photos 3 months after presentation shows resolution of the disc swelling but interval development of mild disc pallor and macular retinal pigment epithelial changes. (E) Macular OCT 3 months after presentation shows interval resolution of subretinal fluid but persistent nasal outer retinal loss (green arrows). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

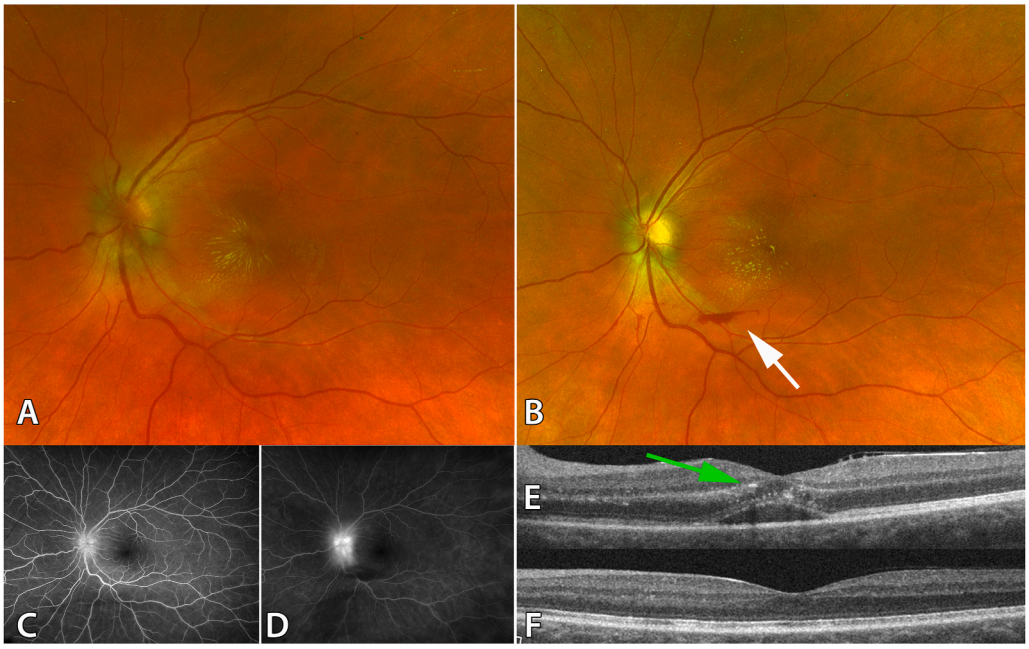

Fig. 2. Multimodal imaging of the left eye of case 2. Baseline fundus photo (A) showing disc swelling, vascular congestions and macular exudates forming a macular star. With 2 months of treatment, the disc swelling and vascular congestion have resolved (B) and the macular exudates have reduced. There is a small preretinal hemorrhage (white arrow) which was first noted 1 week following treatment initiation. Early (C) and late (D) fluorescein angiographic images showing hyperfluorescence and leakage of the optic disc. Baseline macular optical coherence tomography (E) revealed macular thickening, subretinal fluid and hyper-reflective foci corresponding to the hard exudates. Six months later (F) the macular morphology has normalized.

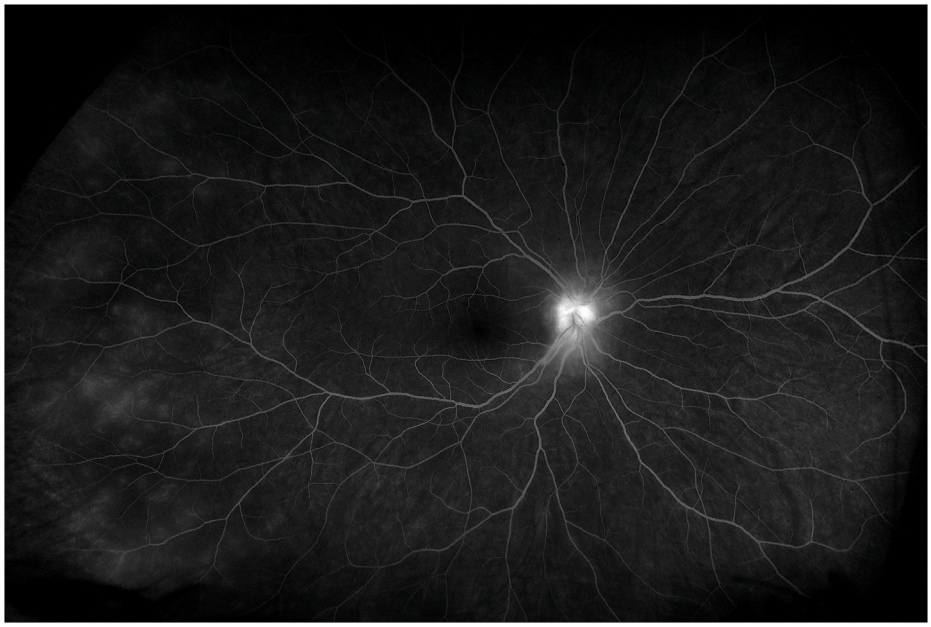

Fig. 3. Late fluorescein angiographic image of the right eye of case 2 at presentation showing hyperfluorescence of the disc and mild peripheral vascular leakage.

3. Discussion

Rickettsia ricketsii, the tick-borne pathogen responsible for Rocky Mountain Spotted Fever (RMSF), has been rarely reported to cause neuroretinitis. Systemic infection with *R. ricketsii* classically presents with the triad of fever, myalgias and arthralgias, and petechial rash. Ocular manifestations of Rickettsial disease include conjunctivitis, keratitis, anterior uveitis, panuveitis, retinitis, retinal vascular changes, and optic nerve involvement.[4] In our review of the literature, we found only one reported case of macular star in association with *R. rickettsii* infection.[5]

The literature suggests that the near-complete recovery of vision is typical in patients with neuroretinitis of various etiologies.[6] Treatment of infectious neuroretinitis cases varies with specific etiology, with the role antibiotic therapy being questionable in some cases such as Bartonella neuroretinitis.[3,7] Although both of our patients showed marked improvement of disc swelling and macular subretinal fluid within the first week of treatment with doxycycline and steroids, our first patient did not experience the same degree of vision improvement, with a final BCVA of 20/60 OS. It is possible that the longer time to initiating treatment (2 months after the onset of his symptoms), and the more severe disc swelling played a role in his diminished response to therapy. In the one previously reported case of *R. ricketsii* neuroretinitis with macular star formation, the patient was treated after seven weeks of vision loss with oral doxycycline alone for 14 days, with a last reported BCVA of 20/30 OD and 20/100 OS. Given the paucity of literature on neuroretinitis in Rickettsial disease, it is thus possible that visual recovery in some such cases is not as good as in other causes of

neuroretinitis. It is additionally unclear whether antibiotics, oral steroids or a combination produce superior visual outcomes in cases of Rickettsial neuroretinitis.

Familiarity with the epidemiology of infectious causes of neuroretinitis will help ophthalmologists make the appropriate diagnosis and initiate early treatment.[8] RMSF is caused by the transmission of *Rickettsia rickettsii* by certain tick species, most commonly the wood tick (*Dermacentor andersoni*) and the American dog tick (*Dermacentor variabilis*).[5] Most cases are reported in May through August, coinciding with when the ticks are most active. Although RMSF was first identified in the Rocky Mountains, over 50% of cases are now reported in one of five states: North Carolina, Tennessee, Oklahoma, Arkansas, and Missouri, due to the geographic distribution of tick species.[9] Despite these trends, cases have been reported throughout the contiguous United States and can occur during any month of the year. Given the potential for neuroretinitis to cause permanent vision loss, the presence of acute vision loss, optic disc edema, and macular exudates should prompt an evaluation for Rickettsial disease in endemic areas, even in the absence of systemic symptoms or known history of a tick bite.

Patient Consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

No conflicting relationship exists for any author.

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