Rickettsial Infections of Retina

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Introduction

Rickettsial infection of the retina is frequent but usually asymptomatic and easily overlooked. It most commonly presents as bilateral or unilateral unifocal or multifocal superficial retinitis associated with mild vitritis. Retinal vascular involvement is common including focal or diffuse vascular sheathing, vascular leakage, retinal hemorrhages, and asymptomatic or rarely symptomatic vaso-occlusive events. Optic nerve involvement also is common including optic disc edema, optic disc staining, optic neuritis, neuroretinitis, and anterior ischemic optic neuropathy.

Case 1: Multifocal Rickettsial Retinitis with Retinal Vascular Occlusion

A 32-year-old-male patient with a 1-week history of febrile illness presented with blurred vision in the left eye (LE). On examination, best-corrected visual acuity (BCVA) was 20/20 in the right eye (RE) and 20/32 in the LE. There were no anterior chamber or vitreous cells on slit lamp biomicroscopy. Dilated fundus examination of the LE revealed multiple white retinal lesions of different sizes adjacent to retinal vessels (Fig. 1a). Fluorescein angiography showed early hypofluorescence and late staining of large lesions and focal retinal vascular leakage (Fig. 1b and c). Optical coherence tomography (OCT) section acquired through the inferotemporal retinal lesion disclosed increased inner retinal reflectivity with posterior shadowing and associated serous retinal detachment (Fig. 1d).

Two days after initial presentation, fundus examination showed a triangular area of ischemic retinal whitening in the macular area (Fig. 2a). Fluorescein angiography confirmed the diagnosis of occlusion of a small macular branch retinal artery (Fig. 2b). OCT section acquired through the superotemporal retinal white lesions showed hyperreflectivity of the inner retinal layers with posterior shadowing corresponding to the area of superficial retinitis and increased reflectivity in the nerve fiber layer zone corresponding to the ischemic retinal whitening area seen clinically (Fig. 2c). The indirect immunofluorescence antibody test was positive for *Rickettsia conorii*, and the patient was treated with doxycycline (100 mg twice a day) for 10 days with subsequent improvement of visual acuity and gradual resolution of abnormal ocular findings (Fig. 3).

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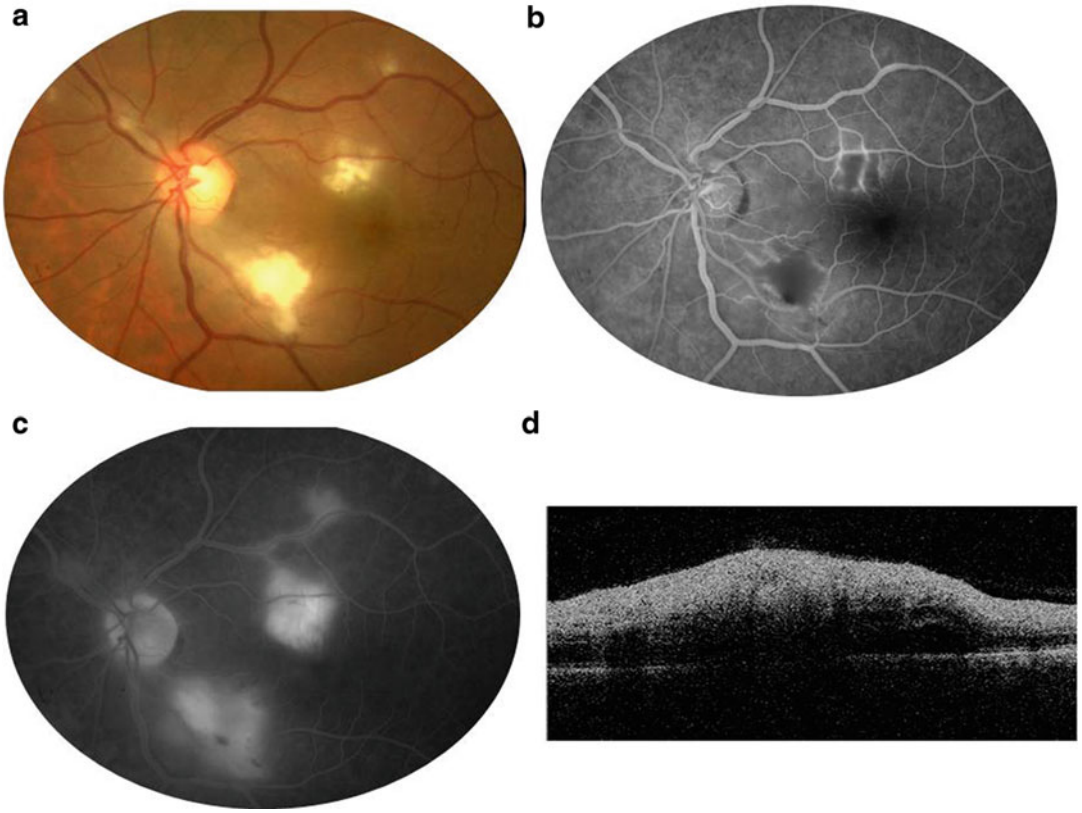

Fig. 1 (**a**) Color fundus photograph of the LE shows multiple juxtavascular superficial white retinal lesions of variable sizes. (**b**) Early-phase fluorescein angiogram of the same eye shows hypofluorescence of large lesions. (**c**) Late-phase fluorescein angiogram of the same eye shows staining of the retinal lesions and associated focal retinal vascular leakage and mild optic disc hyperfluorescence. (**d**) Horizontal optical coherence tomography section acquired through the inferotemporal retinal lesion shows increased inner retinal reflectivity with posterior shadowing and associated serous retinal detachment

Case 2: Rickettsial Infection Presenting as Neuroretinitis

A 44-year-old-female patient with a 1-month history of febrile illness with skin rash presented with vision loss in the RE. On examination, her BCVA was 20/200 in the RE and 20/20 in the LE. There was a relative afferent pupillary defect in the RE. Slit lamp examination revealed 1+ vitreous cells in the RE. Dilated fundus examination of the RE revealed optic disc swelling with macular star, retinal hemorrhages, and peripapillary serous retinal detachment consistent with a diagnosis of neuroretinitis (Fig. 4a). Fundus examination of the LE showed a white retinal lesion superior to the optic disc (Fig. 4b). Fluorescein angiography showed leakage from the optic disc with no evidence of macular abnormality in the RE (Fig. 5a) and late hyperfluorescence of the white retinal lesion in the LE (Fig. 5b). OCT confirmed the presence of peripapillary serous retinal detachment involving the fovea in the RE (Fig. 6). It also showed increased reflectivity within the inner retinal layers at the superficial retinal lesion in the LE (Fig. 7). Serological testing was negative for *Bartonella henselae* and positive for *Rickettsia conorii*. The patient was treated with doxycycline (100 mg twice a day) for 10 days. One month after initial examination, visual acuity was 20/32 in the RE and 20/20 in the LE. The optic disc edema and serous retinal detachment had completely resolved, with development of peripapillary hard exsudates in the RE (Fig. 8a and b). The white retinal lesion in the LE had almost completely resolved (Fig. 8c).

Key Points

- Systemic rickettsial disease usually presents as a self limited febrile illness with skin rash.
- Ocular involvement is frequent but usually asymptomatic.
- Rickettsial infection of the retina: unilateral or bilateral, focal or multifocal superficial retinitis, mild vitritis, retinal vasculitis, and optic nerve involvement.
- The diagnosis is based on epidemiological data, systemic manifestations, and the pattern of ocular involvement.

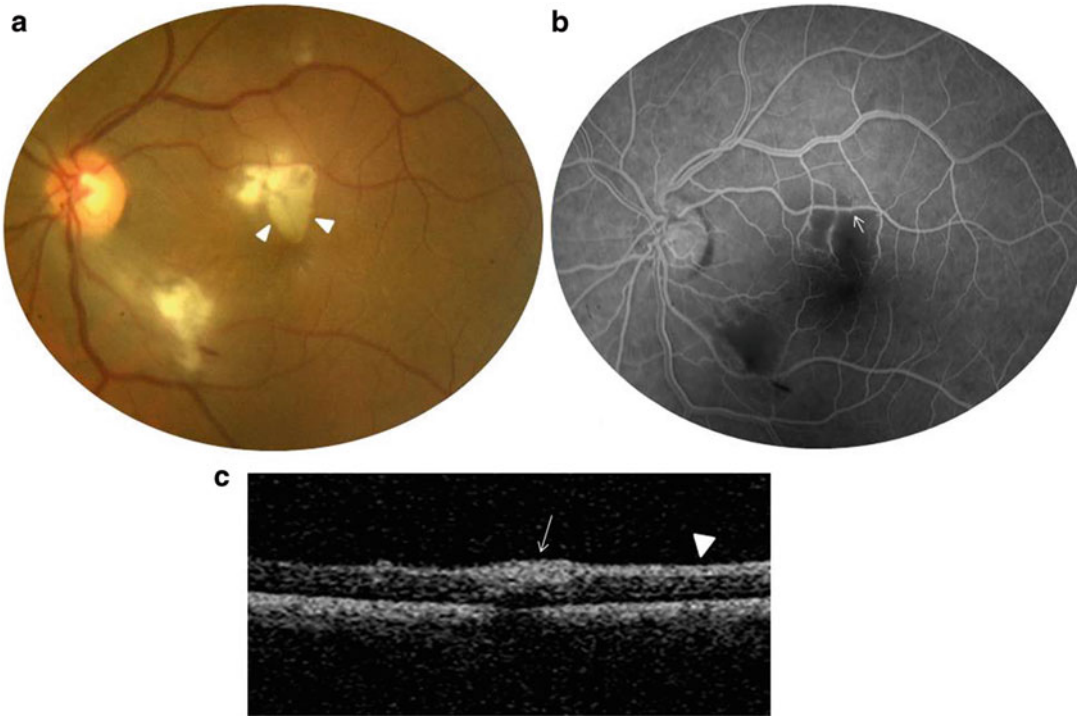

Fig. 2 (**a**) Color fundus photograph of the same eye, two days later, shows the presence of a triangular area of ischemic retinal whitening adjacent to inflammatory lesion (*arrowhead*). (**b**) Early-phase fluorescein angiogram confirms the diagnosis of occlusion of a small macular branch retinal artery (*white arrow*). (**c**) OCT section of the same eye taken through the superotemporal white lesions reveals infiltration of the inner retinal layers with posterior shadowing corresponding to the area of superficial retinitis (*white arrow*) and increased reflectivity in the nerve fiber layer zone consistent with the diagnosis of branch retinal arterial occlusion (*white arrowhead*)

Fig. 3 Color fundus photograph of the same eye, taken two weeks after initial presentation, shows an almost complete resolution of the white retinal areas. Note the occurrence of a partial macular star

Confirmation of diagnosis relies on positive serology and PCR in selected cases.

- Management: Doxycycline. Other antibiotics: macrolides and fluoroquinolones.
- Prognosis of ocular disease: usually self-limiting and rarely persistent visual loss.
- Prevention: Personal protection against tick bites and improvement of sanitary conditions.

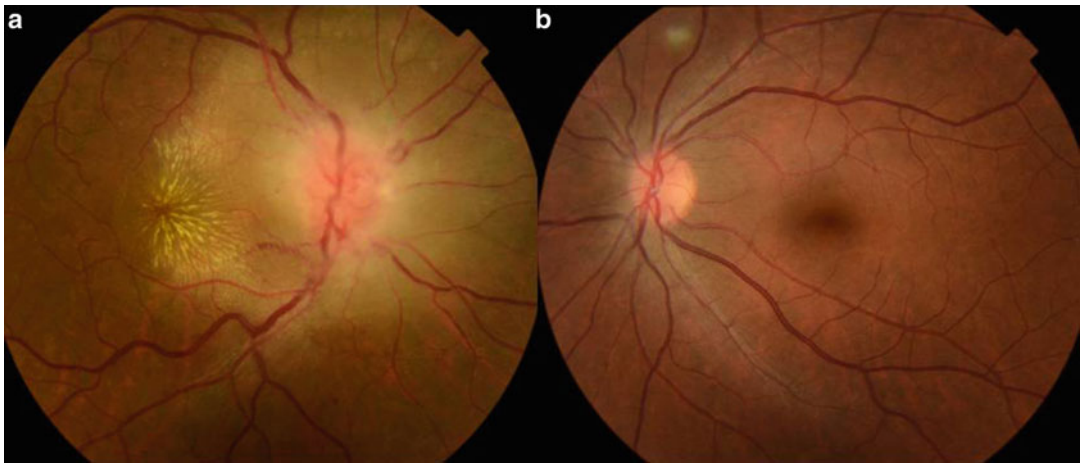

Fig. 4 (**a**) Color fundus photograph of the RE shows a marked optic disc edema, retinal hemorrhages, and a macular star consistent with a diagnosis of neuroretinitis. (**b**) Color fundus photograph of the LE shows a white retinal lesion superior to the optic disc

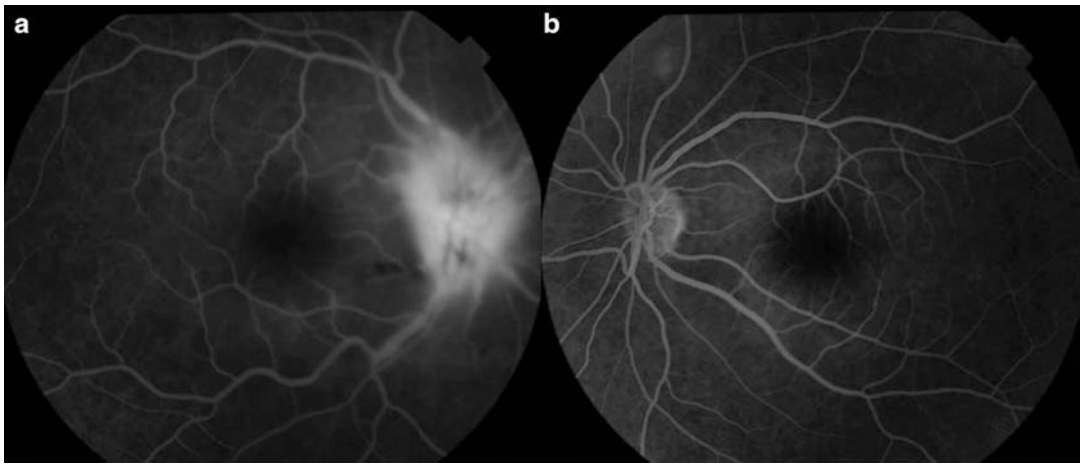

Fig. 5 Late-phase fluorescein angiograms of the same patient shows leakage from the optic disc with no evidence of macular abnormality in the RE (**a**) and late hyperfluorescence of the white retinal lesion in the LE (**b**)

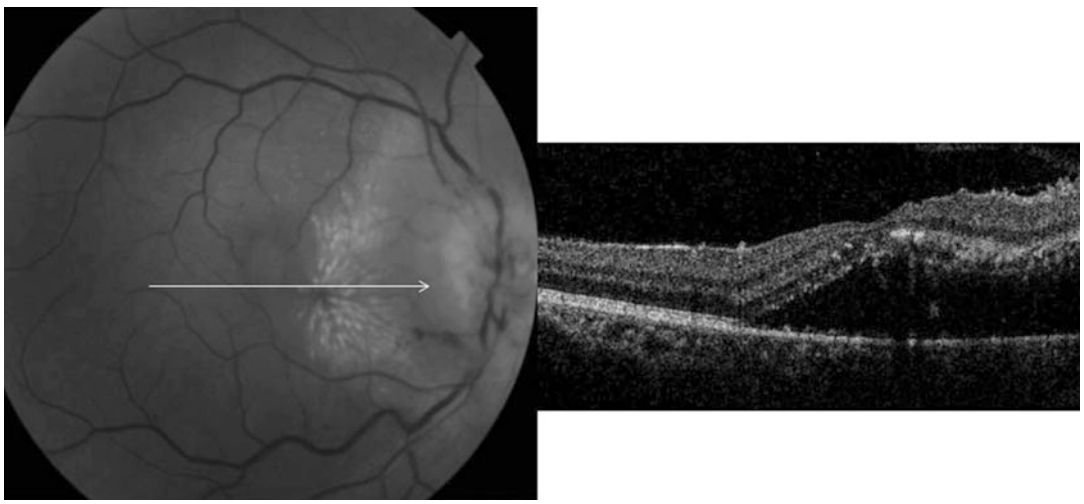

Fig. 6 Horizontal OCT section of the RE shows peripapillary serous retinal detachment involving the fovea

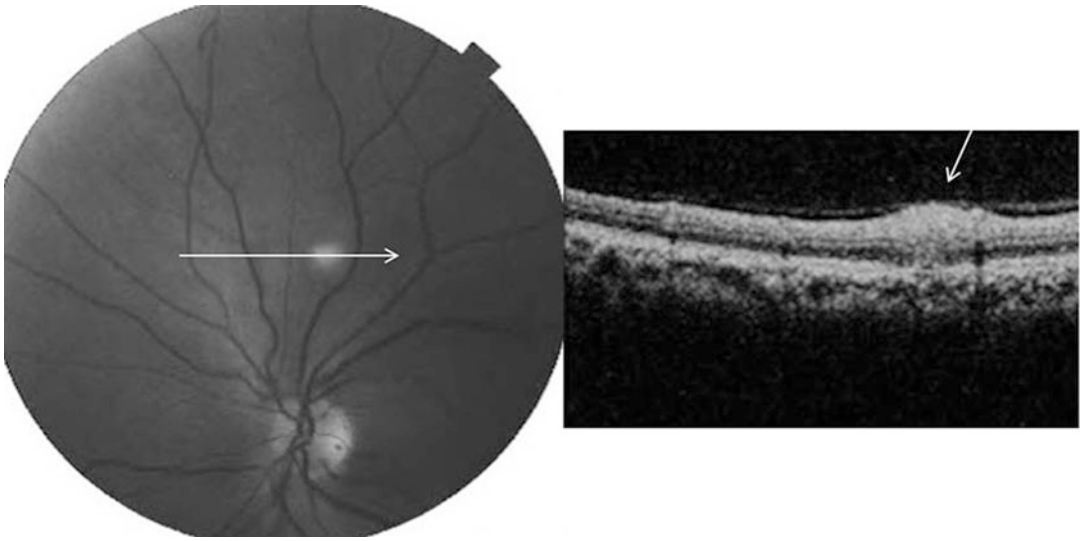

Fig. 7 Horizontal OCT section through the white retinal lesion of the LE shows increased reflectivity of the inner retinal layers (*white arrow*) corresponding to the area of superficial retinitis

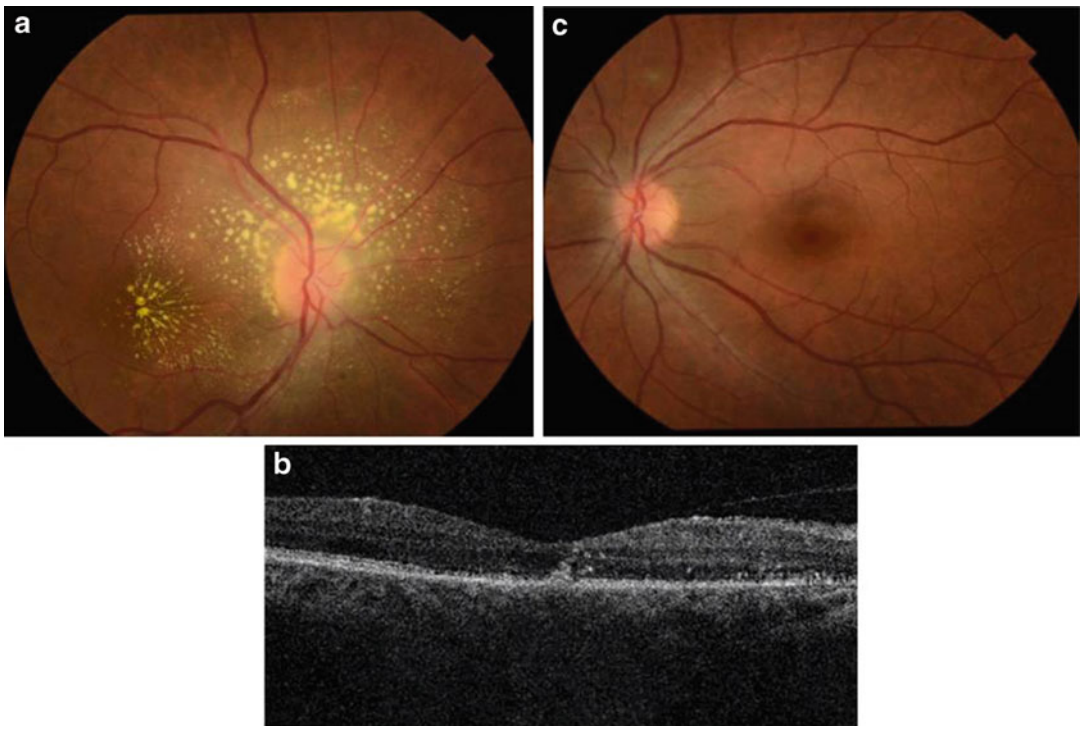

Fig. 8 (**a**) Color fundus photograph and (**b**) OCT of the RE in the same patient taken one month after initial presentation show a complete resolution of the optic disc edema and serous retinal detachment with development of peripapillary hard exsudates. (**c**) Color fundus photograph of the LE shows an almost complete resolution of the white retinal lesion with no obvious chorioretinal scarring

Suggested Reading

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