

The natural history of idiopathic neuroretinitis

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A previously fit 37-year-old woman gave a 5-day history of painless decreased vision in her right eye, perceived as a grey patch over the front of the vision. This had remained unchanged since she had first noticed it. On examination, her right eye visual acuity was 6/36 and she could see 2/11 of the Ishihara plates. There was a right relative afferent pupillary defect. Dilated fundal examination revealed right optic disc swelling and retinal thickening extending to the macula (figure 1A). Optical coherence tomography confirmed the optic disc swelling and retinal thickening but also showed subretinal fluid in the macula (figure 1B).

One week later, she still had optic disc swelling but had developed a classical

macular star (figure 2A). Optical coherence tomography showed the retinal thickening and subretinal fluid had partly resolved but there were new focal areas of hyper-reflectivity in the outer plexiform layer, representing the exudates that made up the macular star (figure 2B). We therefore diagnosed neuroretinitis.

MR imaging of the orbits and brain was normal. Serological testing for syphilis, *Bartonella* and *Borrelia* was positive for *Borrelia*. We gave a 3-week course of intravenous ceftriaxone 2g daily. A subsequent cerebrospinal fluid sample was acellular and her *Borrelia* serology results from a reference laboratory later came back negative.

Her right eye visual acuity improved over 4 weeks to 6/9 and she could see 14/



Figure 1 (A) Optic fundus of the right eye at presentation showing optic disc swelling and macular thickening. (B) Optical coherence tomography showing optic disc swelling, retinal thickening and subretinal fluid (arrowed).

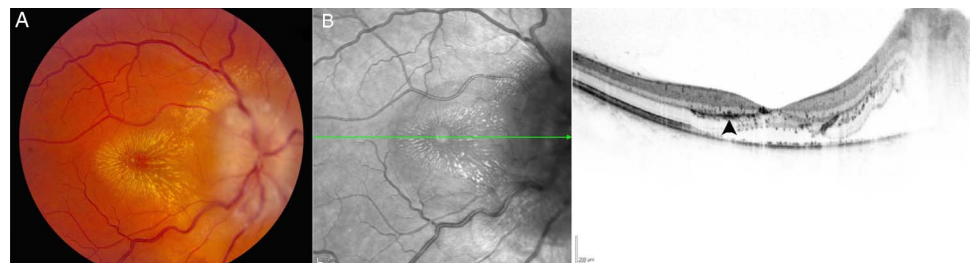


Figure 2 (A) Optic fundus of the right eye 1 week after presentation showing optic disc swelling and a macular star. (B) Optical coherence tomography showing some resolution of the retinal thickening and subretinal fluid, but the accumulation of focal areas of hyper-reflectivity in the outer plexiform layer (arrowhead).



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Figure 3 (A) Optic fundus of the right eye 4 weeks after presentation showing resolution of the optic disc swelling with a persisting macular star. (B) Optical coherence tomography showing that the subretinal fluid has virtually completely resolved.

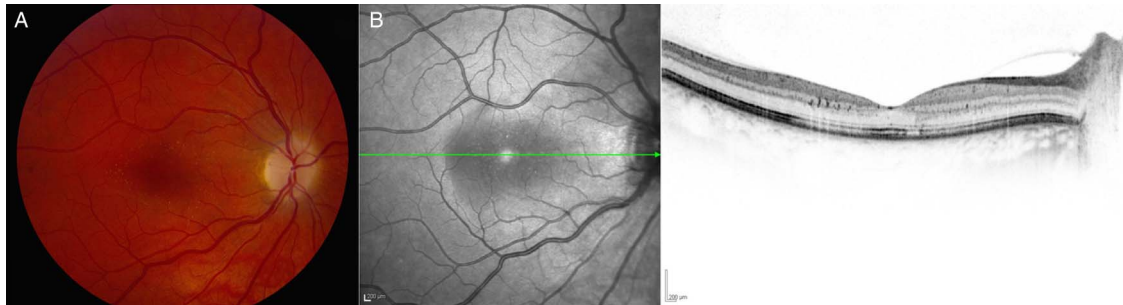


Figure 4 (A) Optic fundus of the right eye 6 months after presentation showing optic disc pallor and resolution of the macular star. (B) Optical coherence tomography showing that the focal areas of hyper-reflectivity in the outer plexiform layer has virtually completely resolved.

17 of the Ishihara plates. The optic disc swelling had lessened and the subretinal fluid had resolved (figure 3).

By 6 months, although her vision had not improved further, the macular star had virtually disappeared and she was left with mild optic disc pallor (figure 4).

Neuroretinitis is an inflammatory condition characterised by optic disc oedema and a macular star.¹ It is probably a form of vasculitis affecting the optic disc with exudation of fluid into the peripapillary retina. Lipid-rich fluid accumulates in the outer nuclear-plexiform space with the aqueous phase passing through the external limiting membrane to accumulate beneath the retina. Due to the loose, radial configuration of the outer plexiform layer, the lipid precipitates in a star-like pattern around the fovea as the serum is resorbed.

The presence of a macular star is not specific to neuroretinitis and can occur in hypertensive retinopathy, papilloedema, diabetic papillopathy and non-arteritic anterior ischaemic optic neuropathy. The other features of the presentation of these conditions help to distinguish these from neuroretinitis.

It is important to recognise neuroretinitis for two main reasons. First, its pathophysiology differs from optic neuritis and there is no link to the subsequent development of multiple sclerosis. Second, although most cases of neuroretinitis are idiopathic and have a good prognosis, it can be associated with a variety of infectious diseases such as cat-scratch disease (*Bartonella*), Lyme disease (*Borrelia*) and syphilis.

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