Very early OCT response to intravenous pulse steroid in Vogt-Koyanagi-Harada disease

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DESCRIPTION

A Caucasian woman in her 40s presented with blurry vision in both eyes for 3 days. Best corrected visual acuity was 20/32 and 20/25 in her right eye and left eye, respectively. Intraocular pressure was 12 mmHg in both eyes, and anterior segment evaluation was unremarkable. Fundus examination revealed multiple neurosensory retinal detachments in the posterior pole and optic disc hyperemia bilaterally. Enhanced depth imaging–optical coherence tomography (EDI-OCT) showed large areas of subretinal fluid (SRF), bacillary layer detachment (BALAD) and markedly thickened choroidal layers in both eyes (figures 1A and 2A). There were no integumentary, neurological or auditory complaints. A diagnosis of presumptive Vogt-Koyanagi-Harada (VKH) was made and the patient was started on intravenous methylprednisolone 1 g/day for 3 days and mycophenolate mofetil. Approximately 20 min after the first pulse of methylprednisolone, there was remarkable improvement of SRF and BALAD already visible on EDI-OCT (figures 1B and 2B), which went on to resolve completely by week 1. In the following 8 months, there was a decrease in choroidal thickness and also progressive and continued reconstitution of the ellipsoid zone on EDI-OCT (figure 3). Mycophenolate was increased to 2 g/day over 8 weeks, and oral steroids were tapered gradually to 7.5 mg/day over 7 months. At 8 months' follow-up, patient remains stable with 20/20 vision in both eyes and sustained remission clinically and on optical coherence tomography (OCT). VKH disease is a primary stromal choroiditis, coursing with exudative retinal detachments and usually mild anterior and vitreous inflammation. Inflammation is thought to originate primarily from the choroid, affecting nearby structures in a spill-over fashion.[1 2] Progression to chronic recurrent granulomatous anterior uveitis and sunset glow fundus with chorioretinal atrophy is likely, unless early and aggressive immunosuppression is implemented.[3] It has been previously suggested that there may be a window of therapeutic opportunity, between 2 and 4 weeks of disease onset, where starting aggressive immunosuppression as soon as possible may modify the disease course,[3 4] allowing for progression to sunset glow fundus to be altogether avoided. To the best of our knowledge, this is the first report wherein objective and remarkable improvement just minutes after a first pulse of intravenous steroid was documented, contributing to the existing body of evidence on the benefit of this approach. Steroid-sparing agents should also be introduced within the first few days to help prevent disease recurrence[3] and provide the best chances for true disease course modification.[5] The ongoing improvement of the outer retinal layers on OCT through to last follow-up at 8 months brings further ultrastructural detail in support of this strategy.

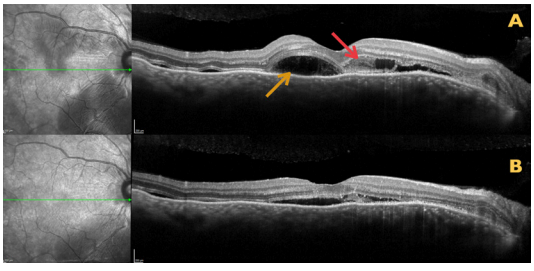

Figure 1 Right eye: (A) Multiple, macular SRF pockets (yellow arrow) and BALAD (red arrow) nasal to the fovea at baseline. (B) Improvement of SRF pockets and BALAD 20 min after the first pulse of intravenous methylprednisolone. BALAD, bacillary layer detachment; SRF, subretinal fluid.

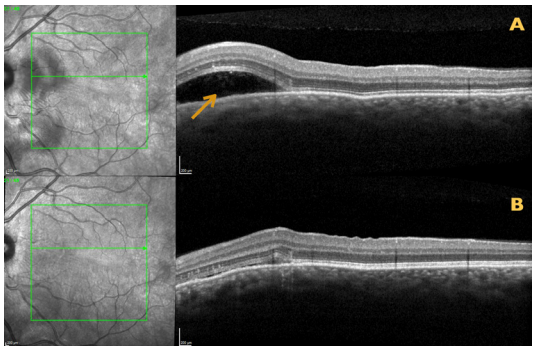

Figure 2 Left eye: (A) Single, peripapillary SRF pocket (yellow arrow). (B) Almost complete reabsorption of peripapillary SRF pocket 20 min after the first pulse of intravenous methylprednisolone. SRF, subretinal fluid.

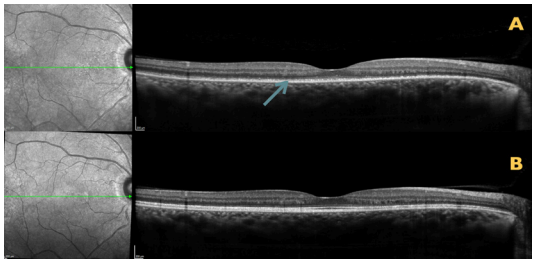

Figure 3 Right eye: (A) at 1 month, disruption of the EZ (blue arrow); (B) at 8 months, complete reconstitution of the EZ without any residual structural abnormalities. EZ, ellipsoid zone.

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Learning points

- Vogt-Koyanagi-Harada disease is a primary stromal choroiditis that can progress to chronic illness with a poor visual prognosis if not treated promptly.
- On the first 2–4 weeks of presentation, treatment with systemic steroids and early immunosuppression may cure the disease.
- Response to pulsed intravenous methylprednisolone may be extremely fast, starting within minutes.
- Early pulsed intravenous methylprednisolone and/or early immunosuppression may promote continued restoration of outer retinal layers even after 6 months.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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