Rare disease

Missed perforation presenting as sympathetic ophthalmia in an adolescent Malay girl

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Summary

To describe the rare presentation of sympathetic ophthalmia in a teenage girl with no previous known ocular injury.

BACKGROUND

I hope to share with my colleagues a rare case of sympathetic ophthalmia that can present in adolescence after a missed childhood injury.

CASE PRESENTATION

A teenaged Malay girl from East Malaysia presented with a 6-month history of left eye redness associated with 1-week history of decreased vision and photophobia. Her mother had also noticed recurrent eyelid swelling and a black lesion on the left eye. There was no history of previous ocular trauma or surgery. She had no other medical history of note and no other systemic complaints.

On examination, her visual acuity was 6/12, 6/12⁺, N8 OD and 6/60 OS. The striking abnormality was the presence of a prolapsed iris plugging a defect in the left cornea (figure 1) with bilateral anterior chamber cellular reaction of 2+ OD and 3+ OS with fibrin. The iris was covered by a membrane measuring 2.8×1.5 mm with

limbal vascularisation. The anterior chamber was shallow infero-temporally with mutton fat keratic precipitates. There was no posterior synechiae or cataract and intraocular pressure was normal bilaterally.

Left eye fundal examination was hazy with presence of yellow, subretinal nodules involving the inferior fundus and a hyperaemic optic disc. The right eye fundal view showed a serous detachment involving the right supero-temporal arcade with multiple smaller areas of serous detachment (figure 2).

Fundus fluorescein angiogram showed multiple pinpoint areas of leakage at the posterior pole corresponding to the serous detachments (figure 3).

INVESTIGATIONS

Full blood count, renal profile, random blood sugar, Venereal Disease Research Laboratory, Mantoux test, erythrocyte sedimentation rate, chest x-ray and urinalysis, electrocardiogram.

Figure 1 Anterior segment photograph of the patient's left eye showing prolapsed iris tissue with healed perforation involving the left cornea associated with vascularisation, pupillary distortion and shallow anterior chamber inferotemporally.

Figure 2 Right fundus photograph showing a serous retinal detachment involving the right superotemporal arcade with multiple areas of serous detachment.

Figure 3 Right fundus fluorescein angiogram showing multiple pinpoint areas of leakage at the posterior pole associated with serous detachments and diffuse staining of the optic disc.

DIFFERENTIAL DIAGNOSIS

- ▶ Sympathetic ophthalmia
- ▶ Vogt Koyanagi Harada disease
- ▶ Bilateral granulomatous panuveitis
- ▶ Sarcoidosis
- ▶ Ocular tuberculosis
- ▶ Syphilis

TREATMENT

She was treated with modified pulse intravenous methylprednisolone 125 mg 6 hourly for 3 days with oral cyclosporin A (CSA) 5 mg/kg twice daily. On completion of 3 days of the methylprednisolone, her visual acuity had improved to 6/9⁻³, 6/9⁻¹, N6 OD and 6/12, 6/9⁻³, N8 OS with reduction in size of the Dalen Fuch nodules, reduction in vitreous cells and resolution of the exudative detachments.

Figure 4 Right eye fundus photograph after 6 months of therapy showing complete resolution of the serous detachments and a normal looking fundus.

OUTCOME AND FOLLOW-UP

She remained on a 6-month regime of CSA, low-dose oral and topical steroids with regular monitoring of renal, liver and blood counts. Her visual acuity at 6-month follow-up was 6/9, 6/6⁻³, N6 OD and 6/12, 6/6⁻¹, N6 OS. There was persistent low-grade cellular activity bilaterally but the serous detachments had completely resolved (figure 4). She requested to be monitored in her home state of Sabah, East Malaysia. There was no plan to operate on the left eye as the perforation had sealed spontaneously.

DISCUSSION

Sympathetic ophthalmia (SO) is a T-cell-mediated, bilateral, diffuse granulomatous panuveitis that occurs after penetrating trauma or surgery.¹ The inflammation may commence as early as 3 weeks to 3 months after the injury.¹ The main risk in the UK and Ireland was surgery, in particular retinal surgery, although a history of antecedent trauma is significant.² The incidence of SO after penetrating injury is extremely rare. In fact, a study by Gurdal *et al*³ of 217 patients with repaired penetrating injuries who were followed up to 43 years with a mean of 10.3 years found only one case of uveitis without histological confirmation of SO. However, this case illustrates that it can occur in adolescents following a missed, self-sealing injury and should be suspected in cases of unexplained panuveitis.

The symptoms of SO are blurred vision and photophobia in the sympathising eye as the ciliary body, by virtue of its uveal tissue, is among the first to be affected. Vision and photophobia worsen concurrently in the exciting eye due to a bilateral non-necrotising granulomatous panuveitis. Clues include evidence of ocular trauma in the presence of bilateral retinal pigment epithelium (RPE) disturbance as evidenced by Dalen Fuch nodules which represent collections of epithelioid cells lying between the Bruch membrane and the RPE. Localised exudative detachments tend to overlie these nodules resulting in typical fluorescein angiographic findings of initial hypofluorescence followed by late hyperfluorescence.⁴

SO is a relapsing and remitting condition because once it has developed, enucleation cannot prevent further recurrences in the sympathising eye.¹ It is also such a rare disease that it is impossible to justify enucleation in eyes with remaining useful vision as in this case in order to prevent development of SO.

Intensive systemic and topical corticosteroid therapy has improved the outcome and reduced the relapse rate of SO.^{1 5} It has a generally good prognosis rate with timely treatment and regular follow-up.⁵ The concern in this case is usage of steroids in a growing child. Hence, immunomodulating agents were started right at the start of her therapy for their steroid sparing effects.⁶ The use of immunomodulators in children necessitates monitoring of their bone marrow function and carries a lifelong risk of blood dyscrasias. Alternative therapies include the usage of local therapies such as intravitreal triamcinolone.⁷ However, not only are these more challenging to administer in a child, they also have potential local side effects. Infliximab is another agent which has been shown to have an effect but should be reserved for cases of refractory SO.⁸

Learning points

- ▶ Unilateral ocular trauma may be insidious in children and adolescents.
- ▶ Unilateral ocular trauma leading to perforation predisposes to sympathetic ophthalmitis.
- ▶ Sympathetic ophthalmitis may occur in a growing child.
- ▶ Immunosuppressive treatment for SO in children and teenagers needs to be specially tailored to consider their needs in rapid development.

Competing interests None.

Patient consent Obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Bastion M-LC, Mohamad MH. Missed perforation presenting as sympathetic ophthalmia in an adolescent Malay girl. *BMJ Case Reports* 2012;10.1136/bcr-2012-006525, Published XXX

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