CASE REPORT

Sympathetic ophthalmitis with predominant posterior segment involvement

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SUMMARY

We report a case of a 22-year-old obese man, intellectually disabled since birth, who developed sympathetic ophthalmitis in his right eye (RE) 2 months after a penetrating injury to his left eye; this despite primary repair went into phthisis with no perception of light. Initially the patient presented with features of anterior uveitis in the RE, for which oral and topical steroids were prescribed. The patient failed to keep regular follow-up appointments, and returned 2 months later with exudative retinal detachment, and was placed on steroids and immunosuppressives. The visual acuity of the RE on discharge was 6/60 with -0.5×180 .

BACKGROUND

Ocular trauma, once described as the 'neglected disorder', has been highlighted as a major cause of visual morbidity. Sympathetic ophthalmitis (SO) commonly occurs after accidental non-surgical ocular trauma. In the modern era, SO is rare, possibly on account of the tendency to rapidly institute steroid therapy. Predominant posterior segment involvement is not a regular feature of this disease. Exudative retinal detachment has been reported in only a few cases from India. This case is of interest since possibly on account of intellectual disability, treatment and follow-up were neglected, resulting in inadequate management, leading to an exudative retinal detachment in the sympathising eye: an occurrence which would be rare if timely therapy had been instituted.

CASE PRESENTATION

A 22-year-old man reported within 6 h after sustaining a penetrating accidental injury to his left eye (LE) with a wooden stick. On general examination, the patient was a young man, obese (weight 102 kg, height 172 cm, body mass index 34.4 kg/m^2), with a pulse rate of 78/min, blood pressure 114/68 mm Hg, respiratory rate 16/min, and was afebrile. The patient was intellectually disabled since childhood. He was youngest of the nine siblings. On examination, globe perforation with uveal tissue prolapse revealed visual acuity (VA) of the LE with no perception of light (NPL) and 6/6 in the right eye (RE). The patient underwent corneoscleral suturing and was discharged a week later on a combination of ofloxacin (0.3%) and dexamethasone (1%) eye drops along with oral prednisolone (60 mg once daily, with taper over 3 weeks), and advised regular weekly follow-ups.

The patient reported back to us 2 months later with marked diminution of vision in the RE

associated with pain, redness and photophobia for 1 week. Vision was HM in the RE; the LE was phthisical. Biomicroscopy revealed a hazy cornea, with cells 2+ and flare 2+ in the anterior chamber. The pupil was fixed and non-reactive, with posterior synechiae present in the superotemporal and superonasal quadrants. The lens was clear, with cells 3+ in the retrolenticular space; vitreous was hazy, although fundus revealed no abnormality, and retinal vessels could be traced to the second order branching. The pressure (non-contact tonometer) in the RE was 9 mm Hg. There was no history of any cutaneous changes (such as vitiligo, alopecia, poliosis), joint pain or joint stiffness, oral or genital ulcers, abdominal pain, altered bowel habits or blood in stools. With a provisional diagnosis of SO, indoor treatment was instituted. The patient instilled a combination of ofloxacin (0.3%) and dexamethasone (1%) eye drops six times a day, along with tropicamide (0.8%) and phenylephrine (0.5%) eye drops once at night, in addition to oral prednisolone (60 mg once daily, with taper). A posterior subtenon injection of triamcinolone (40 mg in 1 ml) was given in the RE on the third day of admission, considering that this was the only functional eye. The patient improved symptomatically and was discharged 4 days later with a VA of 6/18 in the RE, with instructions to keep a weekly follow-up.

The patient failed to keep the follow-up appointments, and reported 2 months after discharge, with sudden, painless and marked diminution of vision. He had stopped oral prednisolone after 3 weeks but continued to use the topical medication. VA of the RE was HM, with inaccurate projection in the inferior quadrant. Biomicroscopy showed mild, diffuse superficial conjunctival congestion; a nebular corneal opacity was present at 11 o'clock position (0.5 mm in diameter) about 1 mm from the limbus. The anterior chamber was of normal depth with reaction present (cells 2+, flare 2+). The pupil was fixed and dilated, irregular in shape with peaking at 11 o'clock ([figure 1](#)). Multiple iris pigment clumps ([figure 1](#)) were seen on the anterior capsule of the lens, and a retrolental flare was observed (vitreous cells and haze grade 1).

Ophthalmoscopy revealed an epiretinal membrane with macular oedema ([figure 2](#)), with an exudative retinal detachment almost covering the inferior quadrant just reaching up to the disc. A pigmented (black) scar, around half-disc dioptre long was present one disc dioptre nasally to the disc. Ocular movements were full and free. The non-contact intraocular pressure was 9 mm Hg.

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Figure 1 Slit lamp photograph of the right eye showing dilated, fixed pupil, irregular in shape with peaking present at 11 o'clock position. Multiple iris pigment clumps are seen on the anterior capsule of the lens.

After admission, the patient was given intramuscular dexamethasone (32 mg twice daily, for 7 days). When the patient showed no signs of improvement, oral azathioprine was added (100 mg twice daily). Over the next 7 days the VA improved to 6/60 (with -0.5×180). The patient was discharged with advice of the medication and instructions to keep a weekly follow-up.

INVESTIGATIONS

As the patient was intellectually disabled, it was difficult to get the appropriate co-operation for good-quality fundus photographs. For the same reason, optical coherence tomography and fundus fluorescein angiography were not performed. B-scan of the RE showed retinal detachment and choroidal thickening, while the vitreous showed scattered haemorrhages with normal optic nerve (figure 3). Blood sugar levels, renal function tests, liver function tests and complete blood counts were all within normal limits.

Figure 2 Fundus photograph of the right eye showing hazy media and epiretinal membrane with macular oedema.

Figure 3 B-scan of the right eye showing retinal detachment and choroidal thickening, while the vitreous showing scattered haemorrhages with normal optic nerve.

OUTCOME AND FOLLOW-UP

The VA of the RE on discharge was 6/60 with -0.5×180 and NPL in the LE.

DISCUSSION

SO is a bilateral diffuse granulomatous uveitis that occurs after either surgery or penetrating trauma to one eye, which is then referred to as the exciting eye, while the non-injured eye is called the sympathising eye. The time between the inciting injury and SO may range from a few days to decades, with 80% of the cases occurring within 3 months, and 90% within 1 year.^{1 2} SO occurs most commonly after accidental non-surgical ocular trauma.^{3 4} It has also been reported after different intraocular procedures such as cataract extraction, trans-scleral neodymium:YAG cyclodestruction, evisceration, paracentesis, iridectomy, pars plana vitrectomy and retinal detachment repair.^{5 6} Incidence following non-surgical penetrating wounds is 0.19%, after intraocular surgeries is 0.007% and after pars plana vitrectomy is 0.01%.^{3 7} Patients usually present with blurry vision, pain, epiphora and photophobia in the sympathising eye. The inflammatory response seen in the anterior segment is a granulomatous iridocyclitis in the classic case: with mutton fat keratic precipitate and varying grade of cells and flare in the anterior chamber. In the posterior segment, the extent of inflammation can vary: vitritis, retinal vasculitis, chorioiditis or papillitis. At times the inflammation may lead to a serous retinal detachment and optic nerve swelling. The sequelae of inflammation vary, depending on the severity of the ocular inflammation and therapy. Secondary glaucoma as well as cataracts can occur. Systemic corticosteroids are the corner stone of therapy. In cases who respond poorly to steroids, or have clinically significant side effects, cyclosporine, azathioprine or other immunosuppressives can be used for long-term

immunomodulatory therapy. Exudative retinal detachment has been reported in only a few cases from India.⁸ This case is of interest since possibly on account of intellectual disability, treatment and follow-up were neglected, resulting in inadequate management, leading to an exudative retinal detachment in the sympathising eye: an occurrence which would be rare if timely therapy had been instituted.

Learning points

- ▶ Sympathetic ophthalmitis is a rare and visually devastating panuveitis.
- ▶ Thorough examination of anterior and posterior segment (B-scan and optical coherence tomography, if needed) of the normal eye in cases of unilateral ocular trauma should be conducted.
- ▶ Prompt and effective management with systemic immunosuppressive agents may control the disease and help in retention of good visual acuity.
- ▶ Steroid therapy should be slowly tapered over 6 months; an aspect which was neglected on account of the intellectually disabled nature of the patient.
- ▶ Controversy still exists over enucleation of the exciting eye after development of sympathetic ophthalmitis.

Contributors GSG made substantial contributions to the intellectual content. He was involved in conception, design of the manuscript. GSG was the resident incharge responsible for patient care. AW diagnosed the posterior segment pathology and advised on treatment. He participated in drafting of the manuscript and critical revision of the manuscript for important intellectual content. SMZ and NA gave valuable insight on management of the case, acquisition of the data and the final drafting of the manuscript.

Competing interests None.

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