STEROID PULSE THERAPY IN SEVERE SYMPATHETIC OPHTHALMITIS – A CASE SERIES OF 4 PATIENTS

Ophthalmology

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ABSTRACT

Sympathetic ophthalmitis (SO) is a rare bilateral, granulomatous uveitis that occurs following surgical or accidental trauma to one eye. Injured eye is the Exciting eye and the fellow eye is the Sympathizing eye. Most accepted patho-physiology is immune mediated. In this series, 4 patients with varied presentations were started on steroid pulse therapy (SPT) following routine blood investigation. 1 gram i.v methyl-prednisolone was given on 3 consecutive days in 300 ml Normal Saline under close observation. The patients were then shifted to oral steroid which was gradually tapered over 11 days. The patients were followed up for a minimum period of 14 days and the visual outcome was recorded.

KEYWORDS

Granulomatous uveitis, Dalen-Fuch's nodules, Enucleation, Methylprednisolone, Steroid pulse therapy, Exciting eye, Sympathizing eye.

INTRODUCTION: The term Sympathetic Ophthalmitis (SO) was coined by William Mackenzie (1840), who provided the first full clinical description of the disease. [1]

It can be defined as a rare bilateral, granulomatous uveitis that occurs following surgical or accidental trauma to one eye. [2] Injured eye is the Exciting eye and the fellow eye is the Sympathizing eye. Most accepted patho-physiology is immune mediated.

Epidemiology: It appears to have no predilection toward any particular age, race or gender. Incidence is 0.2% to 0.5% following injury and 0.1% following intraocular injury. [3]

Time of presentation after the inciting injury is variable ranging from 1 week to 66 years, with a majority (90%) occurring within the first year. [4, 5]

Surgical procedures that may lead to sympathetic ophthalmitis include - paracentesis, keratectomy, penetrating keratoplasty, glaucoma drainage device placement, irideocyclectomy, trabeculectomy, trabeculoplasty, iridectomy, cyclodialysis, cataract extraction, retinal detachment repair, laser retinopexy, evisceration, laser cyclocoagulation, vitrectomy, etc. [6, 7, 8, 9, 10]

Clinical Features: The exciting eye is frequently red and irritable. The sympathizing eye develops irritation, pain, blurred vision, photophobia and loss of accommodation.

Uveitis develops in both eyes which may be mild to severe and granulomatous in nature. The degree of severity of inflammation may be markedly asymmetrical in each eye.

On fundus examination, multifocal choroidal infiltrates develop in the mid-periphery with sub-RPE infiltrates corresponding to Dalen-Fuch's nodules. Exudative retinal detachment, vasculitis and optic disc swelling may all manifest. As inflammation subsides chorioretinal scarring may be there.

Investigation: Ultrasound B-scan may show choroidal thickening and exudative retinal detachment.

OCT is useful in quantifying and monitoring the changes.

Fluorescein angiography may show multiple foci of leakage at the level of the RPE with sub-retinal pooling in the presence of exudative retinal detachment.

ICGA may show hypofluorescent spots in active disease, which resolve with treatment.

Management: Enucleation of the severely injured eye within the first week or so of the injury has historically been considered effective in preventing or reducing the severity of SO. But there is some evidence that little useful effect is exerted, particularly with modern standards of surgical repair. Enucleation may be considered in an eye with worse visual prognosis. Evisceration has conventionally been viewed as inadequate, though recent evidence has raised the possibility of a protective effect provided all uveal tissue is removed.

Steroids are the basis of treatment. High dose oral prednisolone is given for several months and gradually tapered according to response. IV methylprednisolone has been tried in severe and refractory cases.

Supplementary topical steroids and cycloplegics are given to target anterior uveitis and peri and intraocular steroids may facilitate to reduce systemic treatment. Also immunosuppressives such as azathioprine, cyclosporine and methotrexate can be used in resistant cases. Biological blockers are sometimes considered.

PATIENTS AND METHODS: 4 patients with severe SO presenting with varied features were included in this study. After thorough examination of the patients and preliminary investigations, they were started on pulsed steroid therapy.

Pulse Steroid protocol:

IV Methylprednisolone: 20 mg/kg body weight/day in 300 ml NS X 3 days

Followed by Oral Prednisolone acetate: 1 mg/kg body wt. /day X 11 days

The patients were followed up for a minimum period of 14 days and the visual outcome was recorded.

CASE 1: A 24 yr old male presented with diminution of vision in R/E (4 days), pain and redness in B/E (2 days). Presenting vision was 6/60 in R/E and no light perception in L/E. He had h/o penetrating injury to his L/E about 45 days back for which he underwent sclero-corneal repair immediately. On examination under slit lamp, cells and flare 2+ in B/E with fine KPs were seen. In the L/E, the repaired sclero-corneal scar was clearly visible.

On funduscopy, in L/E fundal glow was absent. In R/E, disc edema, macular edema and choroidal folds were present. The patient was started on SPT immediately after routine blood investigations turned out to be normal. After 14 days of follow up, on fundus photograph it was observed that the disc edema and macular edema subsided considerably and vision in R/E (unaided) was 6/12.

R/E

L/E

R/E Fundus before Treatment

R/E Fundus after Treatment

CASE 2: A 36 yr old male presented with diminution of vision in L/E (30 days) and pain and redness in L/E (3 days). R/E was prosthetic. He gave history of penetrating injury in his R/E about 1 yr back which was initially repaired. But the pain in the R/E was so severe that the patient ultimately underwent enucleation of the eye after about 2 and half months of injury.

The presenting vision in L/E was FC at 2 m. Pupil was irregular and sluggishly reacting. Cells and flare 2+ and mutton fat KPs present. Fundus was normal. SPT was given along with topical drops. After 14 days of follow-up the vision improved to 6/60.

L/E

CASE 3: 28 yr old male with photophobia and DOV in OS (4 days). In R/E, vision was no light perception with h/o penetrating injury 4 months back which was repaired. In L/E, vision was FC at 1m with Cells and flare 2+ and exudative membrane in pupillary area. SPT was started. After 14 days follow up, vision was 6/60.

R/E

L/E

CASE 4: A 44 yr old male presented with redness, photophobia and diminution of vision L/E (3 days). Presenting vision was HM +ve in R/E and 6/24 in L/E. He had h/o penetrating injury to his R/E about 7 months back which was repaired immediately. On examination, cells and flare 2+ and KPs were present. Steroid pulse therapy was given and the final visual outcome after 2 weeks was 6/12 in L/E.

R/E

L/E

DISCUSSION: A number of theories have been put forward to understand the etiology of sympathetic ophthalmitis. But still it is poorly understood owing to the disease being rare.

Pathophysiology is believed to be auto-immune reaction against the

exposed ocular antigens from the inciting eye which may be found in the uveal tissue, retina or choroid.

In a study done by Lubin et. al. [11] on 105 eyes with sympathetic ophthalmitis, he found that histologic sections of all the cases showed evidence of penetrating injury similar to our study where history of penetrating injury was present in all the cases. Also in his study Lubin found frequent involvement of optic nerve in the form of neuritis, papillitis, optic atrophy and even meningitis.

In his study done in 1975, Schlaegel [12] mentions papillitis to be a prominent feature of sympathetic ophthalmitis and can be used to follow the clinical course of the disease. In our study, disc edema was present in one case which resolved after treatment with pulse steroid.

Dalen-Fuchs nodules are said to be typical but not pathognomic of Sympathetic ophthalmitis. But Lupin [11], in his study found Dalen Fuchs nodules to be present in all his cases on histological evaluation. In our study histological examination could not be done. Yet the fundus pictures of the cases were not suggestive of such nodules.

Early excision of the injured eye is thought to be the best prophylaxis when there is no chance of saving useful vision. [6]

Treatment of Sympathetic ophthalmitis is based on prompt and aggressive anti-inflammatory therapy. Corticosteroids form the mainstay. In a case study done in 1997 by Hebestreit et al. [13], he stated a case of recalcitrant sympathetic ophthalmitis that was effectively treated with pulsed steroid therapy with methylprednisolone 20mg/kg body weight. Following this, many other studies were undertaken to see the efficacy of pulse steroid in sympathetic ophthalmitis. In our case study, all the cases responded well to pulse methylprednisolone.

STERIOD PULSE THERAPY: It has been effective in several types of immunologic diseases. Onset of action is rapid and rigorous and short time side effects are mostly absent. Therapeutic profile is far superior and side effects are negligible.

CONCLUSION: Sympathetic ophthalmitis is a rare disease with variable presentations and extremely variable time of onset from the actual time of injury.

Prompt and aggressive anti-inflammatory therapy restores vision.

Steroid pulse therapy in Sympathetic Ophthalmitis meticulously works with negligible side effects compared with long term oral steroid or other cytotoxic drugs. So, it should be considered as a better treatment option as compared to the long term oral treatment.

Further studies with long term follow up of the cases are required to determine the long term prognosis of these eyes following the treatment.

REFERENCES:

1. Mackenzie W, Hewson A, Jones TW. A Practical Treatise on the Diseases of the Eye. Blanchard and Lea; 1855.
2. Duke-Elder S (1966) Sympathetic ophthalmitis. In: Duke-Elder S (ed) Diseases of the uveal tract. Mosby, St. Louis, pp 558-593.
3. Makley TA Jr, Azar A (1978) Sympathetic ophthalmia. A long-term follow-up. Arch Ophthalmol 96(2):257-262.
4. Goto H, Rao NA (1990) Sympathetic ophthalmia and Vogt-Koyanagi-Harada syndrome. Int Ophthalmol Clin 30(4):279-285.
5. Singh TG, Devi KS, Pradipkumar L, Singh NR. Sympathetic ophthalmia in a case of penetrating ocular injury associated with brain injury: A case report. J Med Soc 2015;29:116-8.
6. Rao Rajji V, Rao NA. Sympathetic ophthalmitis. In Yanoff M, Duker JS. Ophthalmology. 4th ed. China: Elsevier Saunders; 2014. p. 767-9.
7. Castiblanco CP, Adelman RA. Sympathetic ophthalmia. Graefes Arch Clin Exp Ophthalmol.2009;247:289-302.
8. Fuchs E. Concerning sympathetic ophthalmia inflammation (including remarks without traumatic serous iritis). Albrecht van Cnefea Arch Ophthalmol 1905;61:365-456.
9. Ganesh SK, Sundaram PM, Biswas J, et al. Cataract surgery in sympathetic ophthalmia. J Cataract Refract Surg 2004;30:2371-6.
10. Gasch AT, Foster CS, Grosskreutz CL, et al. Postoperative sympathetic ophthalmia. Int Ophthalmol Clin 2000;40:69-84.
11. LUBIN JR, ALBERT DM, WEINSTEIN M. Sixty-Five Years of Sympathetic Ophthalmia A Clinicopathologic Review of 105 Cases (1913-1978). OPHTHALMOLOGY. 1980 February; 87: p. 109-121.
12. Schreck E. Weitere veruche zum nachweis eines spezifischen erregers bei sympathetischer ophthalmie. Albrecht v Graefes Arch Klin exp Ophthalmol 1975; 193:229-43.
13. Hebestreit H, Huppertz HI, Sold JE, Dammrich J. Steroid-pulse Therapy May Suppress Inflammation in Severe Sympathetic Ophthalmia. Journal of Pediatric Ophthalmology and Strabismus. 1997 March; 34(2): p. 124-6.