

The Early Signs of Sympathetic Ophthalmia

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ABSTRACT

Sympathetic Ophthalmia is a rare form of form of bilateral, non-necrotizing granulomatous uveitis that may develop after ocular injury. This is a case of sympathetic ophthalmia in a 31 year old man presenting five months after a penetrating globe injury to his left eye. Vision loss in the right eye was unexpected and the patient's uveitis became refractory to prednisone treatment. The patient was successfully treated and his uveitis remains quiet with combined methotrexate and prednisone therapy. This case was well documented with fluorescein angiography and fundus photography throughout the treatment course and highlights early findings of the disease for clinicians and learners.

RÉSUMÉ

L'ophtalmie sympathique est une forme rare d'uvéite granulomateuse bilatérale non nécrosante qui peut se manifester après une lésion oculaire. Le cas d'ophtalmie sympathique présenté s'est manifesté chez un homme de 31 ans, cinq mois après qu'il ait subi une blessure pénétrante du globe oculaire. La perte de la vision dans l'œil droit était imprévue et l'uvéite du patient est devenue réfractaire au traitement avec de la prednisone. Le traitement combiné avec du méthotrexate et de la prednisone a été un succès et l'uvéite du patient reste silencieuse. Ce cas a été bien documenté à l'aide de l'angiofluorographie et de la photographie du fond de l'œil tout au long du traitement. Il illustre bien les premières constatations de la maladie pour les cliniciens et les apprenants.

INTRODUCTION

Sympathetic ophthalmia (SO) is a rare form of bilateral, non-necrotizing granulomatous uveitis that is associated with ocular injury to the "exciting eye", and the contralateral eye, known as the "sympathizing eye", developing posterior inflammation. [1,2]. In severe cases this inflammation can progress to optic nerve swelling and exudative retinal detachment, resulting in legal blindness [1,2]. SO has an incidence of 0.03 per 100,000 in the general population and 0.5% after a penetrating ocular injury [2]. First-line medical treatment for SO involves high dose systemic steroids tapered over 2-3 months to a maintenance dose [3,4]. If a patient demonstrates a lack of response to treatment, steroid-sparing immunosuppressive medications, such as Cyclosporine, Cyclophosphamide, and Azathioprine, can be used [3,4].

CASE PRESENTATION

A 31 year old man presented with decreased vision OD five months following a traumatic globe rupture OS. Despite a primary globe rupture repair and later a pars plana vitrectomy with silicone oil and laser retinopexy, the eye had no light perception OS (Figure 1). He described an insidious onset of blurred vision OD with photophobia and orbital pain at the site of trauma. The best-corrected visual acuity (BCVA) was 20/25 OD, there were 2+ cells and 2+ flare in the anterior chamber (AC), and vitreous OD

had no cells and no flare.

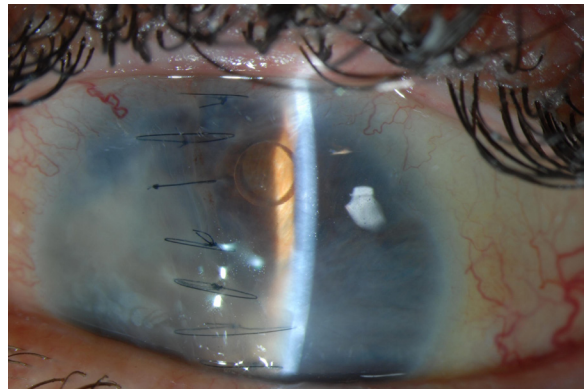


Figure 1: Left eye of a patient following primary closure and a vitrectomy with silicone oil infusion.

Multiple subretinal lesions were noted on funduscopy (figure 2) (Topcon TRC-50DX). Fluorescein angiography (IVFA) (Topcon TRC-50DX) showed discrete subretinal lesions, primarily in the macula and inferior retina, with early hypofluorescence, late hyperfluorescence, and leakage from the optic disc. The left eye could not be assessed because of the prior traumatic injury, dense cataract, and corneal sutures. SO was suspected because of panuveitis in the right eye with anterior segment inflammation and mild vitritis, with a history of penetrating trauma to the left eye. Prednisone 1 mg/kg/d was started with a tapering dose over

Keywords: sympathetic ophthalmia, Methotrexate, uveitis

Case Report



Figure 2: Colour fundus montage of the right, sympathizing eye showing early choroidal inflammatory lesions throughout the fundus, suggestive of Dalen Fuchs nodules.

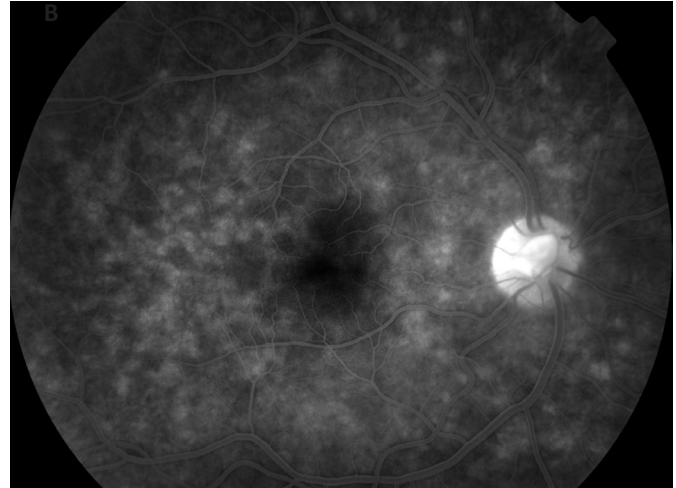


Figure 3: Fluorescein angiography, venous phase, right eye, with systemic prednisone showing choroidal hyperfluorescence and late optic disc leakage as angiographic signs of ocular inflammation (panuveitis).

two months, in addition to prednisolone every 2 hours OU. Seven months following globe rupture, the patient had decreased vision with BCVA of 20/30 and 0.5+ cells in the AC OD. A repeat IVFA showed a similar pattern of choroidal lesions and optic disc leakage (Figure 3) (Topcon TRC-50DX; 35°).

Prednisone was started in combination with methotrexate (MTX) and folic acid supplementation. One month later, the patient's BCVA improved to 20/20 OD and there was no inflammation. IVFA showed resolution of hypo- and hyperfluorescent lesions OD (Figure 4) (Topcon TRC-50DX; 35°). Six months later, the patient was able to taper off of the corticosteroid and his ocular inflammation remains in remission, with no chorioretinal lesions after four months of follow-up on MTX alone (Figure 5) (Topcon TRC-50DX; 35°).

DISCUSSION

We present a case of a patient who developed SO and

decreased vision five months following a penetrating globe injury OS. The recent globe injury OS and the discovery of choroidal lesions, which were likely early Dalen-Fuchs nodules, resulted in a high clinical suspicion of SO and early implementation of therapy. After initial remission of the panuveitis, the recurrence of inflammation was treated with prednisone and a steroid-sparing agent, MTX. The use of long-term immunomodulatory therapy, such as Cyclosporine, Azathioprine, Mycophenolate, Chlorambucil, Cyclophosphamide or MTX, in addition to corticosteroids, has been reported in cases where uveitis did not respond to corticosteroids alone [5]. Intravitreal steroid injections of triamcinolone acetonide, fluocinolone acetonide implants, and infliximab have also been used to control uveitis in cases of SO [1].

MTX hinders the production of thymidylate via inhibition of dihydrofolate reductase, and it has a disproportionate effect on rapidly dividing cells, such as leukocytes [4]. Dihydrofolate reductase is essential for the production of purines

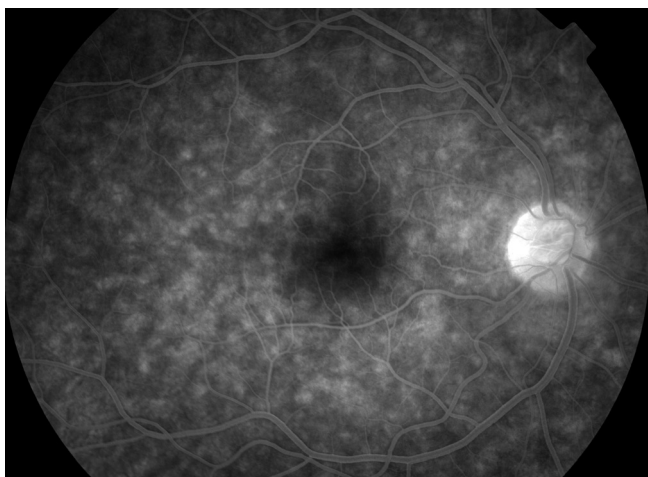


Figure 4: Fluorescein angiography of the right eye, venous phase, after one month of combined MTX-prednisone therapy showing early resolution of chorioretinal lesions and a reduction of optic disc edema.



Figure 5: Colour fundus photograph of the patient's right eye after methotrexate therapy showing clinical resolution of inflammatory chorioretinal lesions.

and thus its inhibition can affect DNA replication. Treatment of non-infectious ocular inflammation is a common off-label use of MTX [1,4,5]. This patient's uveitis relapsed following tapering of oral corticosteroid and was then controlled with oral MTX. The dose of MTX was within the range previously reported and was increased from an initial dose of 10 mg to the therapeutic dose of 15 mg, with ongoing therapy. [4] Remission of inflammation and a resolution of chorioretinal lesions were noted after four weeks of combined therapy.

CONCLUSION

SO is a vision-threatening disease that must be identified and treated promptly to improve prognosis and prevent blindness. This case highlights and documents early findings in SO, and the importance of an early diagnosis and treatment in preventing vision loss. Dalen-Fuchs nodules are characteristic feature of SO, but appear only in 25% to 35% of cases [6]. These nodules begin as a deposit of macrophages in the early phase, but later are primarily composed of lymphocytes and non-functional retinal pigment epithelium [7, 8]. MTX was originally developed as chemotherapy for and remains a mainstay of the treatment of cancer [1,4,5]. The increased susceptibility of fast-dividing cells, such as lymphocytes, to the inhibition of dihydrofolate reductase, makes MTX a treatment option for non-infectious ocular inflammation [4]. Severe uveitis with pain and photophobia following a history significant for ocular surgery or trauma, should raise the suspicion for SO.

LIST OF ABBREVIATIONS

SO - Sympathetic ophthalmia
OU - Both eyes (oculus uterque)
OS - Left eye (oculus sinister)
OD - right eye (oculus dexter)
BCVA - Best Corrected Visual Acuity
IVFA - Fluorescein angiography
MTX - Methotrexate

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