

Sympathetic Ophthalmitis in Nigerians - Case Reports

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SUMMARY:

We describe two cases of sympathetic uveitis in two Nigerian patients. The first case occurred after a penetrating injury while the second was after surgery in a painful blind eye. The aetiopathogenesis as well as the management of the condition in Nigeria are discussed.

KEY WORDS: *Sympathetic Ophthalmitis (Uveitis), Management, Nigeria*

INTRODUCTION

Sympathetic Ophthalmitis (uveitis) is a rare bilateral diffuse granulomatous T Cell mediated uveitis that occurs from about two weeks to many years after penetrating ocular injury¹. It is associated with traumatic uveal incarceration and even more rarely following intraocular surgery^{2,3}.

The injured eye is referred to as the exciting eye, while the second eye is the sympathising eye. Removal of the injured eye before the disease occurs usually protects completely against inflammation developing in the non-injured eye. Once the inflammation starts however, removal of the exciting eye probably has little effect on the course of the disease especially after three to six months. The disease occurs in both eyes (that in the sympathising eye usually begins insidiously) and it presents with blurred vision, photophobia and redness. Some patients may present with accommodation difficulties, and also develop cutaneous and neurological features such as alopecia, vitiligo, vertigo, tinnitus etc. the clinical findings are those of granulomatous panuveitis and include the following: mutton fat keratic precipitates, cells and flare in the anterior chamber.

There are usually inflammatory cells in the vitreous as well and patches of yellowish choroidal infiltrates. Small, discrete, yellowish infiltrates in the retinal pigment epithelium called Dalen-Fuchs nodules (histologically found to lie between the Bruchs membrane and the retinal pigment epithelium) are also visible and may indicate severe dis-

ease. Sometimes, there may be only mild uveitis which could be non granulomatous and it is known that besides the uvea and the retina other ocular structures can be involved. The complications of severe uveitis such as synechiae, rubeosis, glaucoma, blindness, phthisis etc occur in the end stage disease.

Sympathetic Ophthalmitis appears to be delayed type of hypersensitivity reaction of the uvea to antigens located on the retinal pigment epithelium. The antigen however remains unknown, as recent studies by de Smet M.D et al⁴ appears to exclude the S-Antigen (a member of the arrestin family of proteins). Arkhipova L.T.⁵ in his studies and then review of the literature has proposed that Sympathetic Ophthalmitis can be regarded as an autoimmune disease.

The lymphocytes which infiltrate the affected tissues in the disease process are mostly the T type - the suppressor/cytotoxic subset (OKT8). A few are however B lymphocytes.

The Dalen - Fuchs nodules which although not pathognomonic, are characteristic of the disease are composed of a mixture of histiocytes, depigmented retinal pigment epithelia cells and scattered lymphocytes⁶ Sympathetic uveitis is a clinico-pathologic diagnosis. However, in the two cases presented, enucleation for histological studies was not done, so the diagnosis was based on the clinical features.

The two cases are now presented.

CASE I

A healthy seventeen years old Nigerian student presented to the eye department with a history of trauma to the right eye. He had sustained a ruptured globe with uveal tissue prolapse during a fight and presented 4 days after the injury. There was no past medical history of significance. His corrected vision was no perception of light (NPL) in the right eye and 6/6 in the left eye. He had extensive corneoscleral laceration with uveal prolapse in the right eye. There was also complete disruption of the anatomy of the globe with adhesions. The left eye was essentially normal and so was the intraocular pressure. Systemic examination was carried out and patient admitted to the eye ward.

The condition was thoroughly explained to both patient and relatives including all the potential risks, then enucleation of the blind eye offered as the treatment of choice. Unfortunately this was unacceptable to the patient who rather wanted conservative approach. He was then placed on systemic and topical antibiotics and given a booster dose of tetanus toxoid and baseline investigations include fasting blood sugar, urea and electrolytes, full blood count and genotype, erythrocyte sedimentation rates carried out.

The patient then disappeared from the ward and only surfaced two weeks later with a history of rapidly diminishing vision in his good eye.

On examination, the vision in the eye was down to counting fingers. There was severe panuveitis with mutton fat keratic precipitates on the cornea endothelium. The anterior chamber showed 4+ of inflammatory cells. The iris was oedematous with some posterior synechie. There were also inflammatory cells in the vitreous. It was not possible to examine the posterior segment due to the hazy media and the traumatised eye (right) was now phthisical. A diagnosis of sympathetic ophthalmitis was made on the history and the clinical findings. The patient then had repeat baseline investigations and he was placed on:-

- IV Hydrocortisone 100mg stat
- followed by tablets prednisolone 100mg daily
- subconjunctival Depomedrol 40mg stat (both eyes)
- Topical dexamethasone 2 hourly (both eyes)
- Topical Atropine 1% bd (both eyes)

The inflammation in the left eye reduced considerably after the first week and the visual acuity improved to 6/12 unaided.

The patient was then discharged on systemic and topical steroids and given a follow up appointment. When he came four weeks later, the vision in the good eye (left) had dropped to Non Perception of Light (NPL). At this stage not much could be offered the patient apart from rehabilitation and counselling.

It is relevant to note again that although the patient was a seventeen years old, he had an educated adult relative with him at each visit and the case and consequences of refusing treatment was explained to both of them at each stage of management.

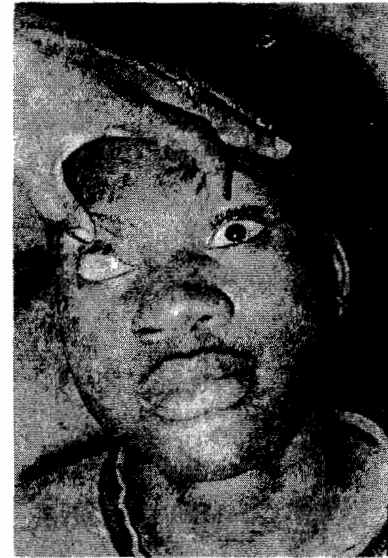


Figure 1: Case of Sympathetic Ophthalmitis in a 17 year old

CASE II

A 60 years old lady presented with a left painful blind eye. She presented with severe pain and left sided headache which had lasted for 2 weeks. There was no history of allergy or other serious illnesses.

On examination, the corrected vision in the left eye was non perception of light while that in the right was 6/6. She had a hazy cornea, shallow anterior chamber with an intumescent cataract in the left eye. The intraocular pressure was elevated at 58mmHg. The right eye was normal with intraocular pressure of 12mmHg. Systemic examination was essentially normal except for mildly elevated blood pressure (160/85mmHg).

After baseline investigations she was placed on:-

- IV Diamox injection 500mg stat followed by
- diamox tablets 250mg twice daily,
- Slow k 1 tablet daily,
- G. Timoptol 0.5% twice daily left eye.

When the intraocular pressure had settled to 24mmHg after 48 hours she had an uncomplicated intracapsular cataract extraction after the visual prognosis had been fully explained to her and the relations.

Post operative recovery was uneventful and the patient was discharged on the 3rd day with a left comfortable eye and an intraocular pressure of 10mmHg. She was reviewed in the clinic a week after surgery, when she was found to be making satisfactory recovery. Her next follow up appointment was fixed at six weeks post surgery, but she failed to keep this. She however presented 8 weeks later

with a history of photophobia in the right eye with a lot of particles which were preventing her from seeing. This had come on about 7 weeks post operatively and she had waited at home another week before presenting. On examination, the vision in the right eye had deteriorated to counting fingers. There were mutton fat keratic precipitates on the corneal endothelium with cells and fundal details could not be visualised in both eyes. A clinical diagnosis of sympathetic ophthalmitis was made. She had a systemic review and routine examination including Hb, FBC, ESR and urea and Electrolytes. Chest X-ray, fasting blood sugar and blood pressure check. The findings were all within normal limits and the blood pressure was 160/85.

She was commenced on:-

- IV hydrocortisone 100mg stat
- Prednisolone 100mg daily
- subconjunctival Depomedrol 40mg in the right and left eye stat
- Topical dexamethasone 2 hourly in the right and left eye for 1 week then 4 hourly for another 2 weeks
- G. atropine 1% R & L eye bd.

The vision improved from counting finger to 6/60 and the patient was able to navigate unaided. The medication was reviewed after 3 weeks and she was placed on prednisolone tabs 40mg daily with weekly monitoring of her vital signs including blood sugar and blood pressure. The frequency of the topical steroid was also reduced to four times daily.

The inflammation in the anterior chamber of the left eye settled down but vision remained poor due to the density of the inflammatory cells in the vitreous. Six months after the initial presentation, the patient had retained a vision of 6/60 in that right eye. She was then lost to follow up.

DISCUSSION

It has long been established that sympathetic uveitis develops in patient who have penetrating wound in the eye as this provides access for intraocular antigens to reach regional lymph node and trigger the cascade which eventually leads to the autoimmune disease^{7,8,9} Thus, procedures such as laser photocoagulation are not associated with the disease despite serious disturbance to the interior of the globe and uveal tissue. Using histological and immunological studies, it has been suggested that sympathetic ophthalmia represents a T-cell mediated cytotoxic reaction toward antigen in or around the uveal melanocytes, retinal pigment epithelium or retina^{10,11}

Marie Comer et al¹² have proposed that patients with HLA-A2, HLA-B5, and HLA-DR 4 are more at risk of developing sympathetic ophthalmia after an ocular injury.

Other melanocytes bearing tissue in the body could be affected because of their common embryological origin in the neural crest.

The management strategy for the disease is in two folds. The first is preventing it from occurring, while the second is aggressive immunosuppression in established disease. Severely traumatised eye that is blind should be enucleated within two weeks of the trauma. This is known to prevent disease occurring in most cases. Enucleation is preferred to evisceration because of the potential risk of antigenic tissue retention which could still activate the inflammatory process in the opposite eye.

Immunosuppression is the mainstay of treatment of Sympathetic Ophthalmitis. The first line is high dose systemic and topical steroids. Periocular steroid injection could also be used as in our cases. Once under control, the steroid is gradually tapered and could take up to a year before complete withdrawal. If adequately instituted, this regime usually leads to a favourable visual outcome^{3,11}. In patients intolerant to steroids or if found inadequate, other immunosuppressives such as Cyclophosphamide, Azathioprine, Cyclosporine etc are used. These drugs it must be remembered have haematological side effects besides their nephrotoxicity and hepatotoxicity and as such complete blood count, liver function test and urea and electrolytes should be done before commencement of treatment, and then repeated periodically in the course of the treatment. The two patients described had penetrating ocular wounds, with the second case occurring post operatively. Post operative sympathetic uveitis is the less common form of presentation. It is however the more commonly reported form in recent literature^{11,12,13}. The excitement we believe has to do with the state of the eyes before the primary surgical procedure. The presence of intumescent cataract with the secondary angle closure glaucoma resulting in unbearable pain warranted the cataract surgery despite the eye being blind in our case. Daxecker F and Kieselbeach G¹⁴ described a case where the disease occurred after a patient has had over six operations in an eye which had primarily developed secondary angle closure glaucoma from central retinal vein occlusion. The importance of appreciating this clinical entity cannot be over emphasised. Although it is rare, when it occurs, uveal inflammatory reaction tends to be more vigorous in blacks than white patients¹⁵. It is important that when handling trauma cases with incarceration of uveal tissue that the patients understand the possibility of sympathetic uveitis occurring if the eye is not enucleated and this is best done within the first two weeks of injury. It is not always possible to have consent to enucleate especially in young adults who find it more difficult to come to terms with losing an eye. In our case even the adult who accompanied the patient could not prevail on him to have an enucleation

and sadly he is now blind, though continuing with his education is a school for the blind.

With our second case with the painful blind eye, it should be noted that alternative lines of management should be considered when faced with similar cases where possible. This will include retrobulbar alcohol and cyclodestructive procedures to relieve pain.

Our patient had come from over 600km away in the rural area hence she was offered surgery as first line of management. The non availability and prohibitive cost of immunosuppressive drugs like cyclosporine should make us wary of operating on a painful blind eye. And it is also very important that patients are fully aware of the risk of surgery before operating on such eyes.

Sympathetic Ophthalmia remains a preventable disease in most cases if only it is kept at the back of the Doctor's mind when managing eyes at risk. We owe it a duty to explain to patients in the language they will understand when suspicious cases are encountered. This will enable us give the appropriate first line treatment and help prevent the disastrous consequences of loosing vision in both eyes as a result of problems arising from one blind eye.

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