Bilateral necrotizing sclero uveitis in a Nigerian: Management and outcome

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ABSTRACT

Introduction: The clinical entity of necrotizing sclerouveitis, though not previously reported in Nigerians and the West African region has been well documented in the literature. The condition is potentially visually debilitating. Case Report: A 83-year-old male who presented to the Eye Foundation Hospital after several years of monitoring of his bilateral advanced glaucoma and having had bilateral cataract surgery, with progressive bilateral temporal sclera ulceration and active ocular inflammation. He was diagnosed as a case of anterior necrotizing sclerouveitis. He was co-managed with a rheumatologist and had steroids and immunosuppressive, Mycophenolate Mofetil with resultant remarkable improvement. Conclusion: This case is presented to heighten the index of suspicion among ophthalmologists practicing in the region and to provide a guide to management.

Keywords: Glaucoma, Necrotizing, Scleritis, Sclerouveitis, Uveitis

INTRODUCTION

Necrotizing scleritis can be associated with uveitis resulting in devastating ocular complications and vision loss. The clinical entity of necrotizing sclerouveitis has been well documented in literature [1]. It requires prompt diagnosis and management to preserve vision and the globe, especially when bilateral. Search done using Medline, Google scholar, and African Journal Online (AJOL) did not reveal any report of a case of necrotizing sclerouveitis occurring in a Nigerian. A case of surgically induced necrotizing scleritis has been reported previously [2]. The acuteness and progressive nature of the condition, low index of suspicion and uncertainty of the management, poses a threat of blindness, especially if as in this case presented, the disease is bilateral.

We present a rare case of acute bilateral idiopathic necrotizing scleritis complicated by uveitis in an elderly male Nigerian who had bilateral progressive temporal scleral ulceration. In this patient, a decision on treatment was taken by a team of ophthalmologist and rheumatologist, which when instituted resulted in immediate improvement in the patients clinical condition with a resolution of the ocular inflammation, cessation of further scleral thinning and return of vision. This case is being reported as a guide to other ophthalmologists who may have such a rare case present to them.
CASE REPORT

A 83-year-old, known glaucoma patient had undergone bilateral phacoemulsification of cataract and posterior chamber intraocular lens insertion four years ago and had attended his routine follow up visits at the Eye Foundation Hospital. His intraocular pressure had been controlled in the lower teens with a combination therapy of: Gutt Alphagan (Brimonidine) 12 hourly; Gutt Duotrav (Travoprost + Timolol) nocte; Gutt Azopt (Brinzolamide) 12 hourly applied to both eyes.

His routine clinic visits had been uneventful until July 2016 when he presented with complaints of severe bilateral eye pain, redness, worsening vision and mild discharge. He was not a known diabetic or hypertensive and did not report any injury to the eyes.

Physical examination of the eyes showed a visual acuity of counting fingers (previously 6/36) in the right eye. Vision in the left eye had always been poor, at light perception only, due to optic nerve glaucomatous damage (Figure 1A and B).

Both eyes were injected as they showed prominent superficial vessels. He also had mucopurulent discharge in both eyes. The anterior chamber and posterior segment of both eyes were normal as there was no evidence of intraocular inflammation. Intra-ocular pressure was 12mmHg in both eyes. Systemic review revealed no significant finding.

Our presumptive diagnosis at this time was bilateral infective conjunctivitis and episcleritis. He was commenced on ciprofloxacin ophthalmic solution applied every eight hourly to both eyes, topical flurbiprofen was also applied eight hourly. His anti-glaucoma medications were also maintained.

During the follow up visit a week later, his vision had deteriorated further in the right eye to barely hand motion and he had difficulty in navigating his way. Pain in the eyes had increased considerably and he had severe conjunctiva and deeper vessel injection. He had also developed bilateral saucer like ulceration of the conjunctiva and sclera in the temporal bulbar region of both eyes, worse on the right (Figure 2A and B).

This ulcer measured 3mm in its widest diameter in the right eye, but was less than this in the left eye. Despite the significant injection of the deeper scleral vessels, the ulcer appeared whitish. There were coarse keratic precipitates in both eyes and 3+ cells and flare in both anterior chambers.

A diagnosis of necrotizing sclero uveitis (possibly auto-immune) was entertained. Patient was referred to a rheumatologist to exclude connective tissue disease.

Laboratory work up showed hematocrit 41%; erythrocyte sedimentation rate – 25 mm/hour; normal white blood cell count; rheumatoid factor – negative; anti-nuclear antibody - negative; C-reactive Protein (CRP) – normal; SGOT, SGPT, Cholesterol, Fasting Blood Sugar, Cholesterol, Triglycerides all within normal limits.

The patient was started by the rheumatologist on pulse Methylprednisolone, Mycophenolate Mofetil – 500 mg bd and Prednisolone tablet – 10mg daily. He was reviewed a week later and reported significant improvement in pain level, redness and vision. Visual acuity in the right eye reverted to previous – 6/36. He has been followed up for 18 months now and his improvement has been sustained. He is now completely independent.

At this point, one week after commencing systemic therapy with a favorable response, it was considered safe to introduce topical steroids in the form of topical dexamethasone 1% ophthalmic solution. This was initiated at a cautious eight hourly dose, while still on his topical ocular hypotensive. Note that topical steroids had not been commenced at the beginning of the treatment for concerns that this may promote further scleral thinning.

In the course of time, he has regained vision in the right eye, which now sees 6/36 as before the disease onset, the scleral ulceration almost immediately ceased to progress and started healing with vascularization of the ulcer bed noted early (Figure 3).

Resolution of the anterior chamber cells was also noted early; within two months the cornea was free of keratic precipitates (KP) and a detailed view of the fundus, which was initially absent because of the KPs and intraocular inflammation, became present again.

Figure 1 (A and B): Fundus picture of right and left eye showing bilateral advanced glaucomatous optic nerve changes.

Figure 2 (A and B): Note the injected eyes and focal areas of sclera ulceration in both the right and let eyes (worse in the Figure 3).

DISCUSSION

Uveitis as a cause of vision loss and blindness had previously been reported in Nigerians [2–4]. Of these three, one case was attributed to a surgically induced necrotizing scleritis following pterygium excision.
and adjuvant use of Mitomycin. In the later patient, investigations did not reveal any systemic disease. Though reported rarely, anterior necrotizing sclero-uveitis, as seen in this patient, may be sight threatening and urgent intervention with immunosuppressive is critical [1, 5, 6]. Necrotizing scleritis can present with an accompanying uveitis and may be bilateral [1]. This condition may be rather severe and is recognized as a devastating form of anterior scleritis for which prompt treatment is critical. In the severest form of the disease, about 21% of patients with this condition may actually die [1]. The presence of scleritis may thus portend serious systemic disease [1, 6].

The reported changes of ectasia and destruction of sclera tissue occurs only in the necrotizing form of this condition [1]. This is mostly in the form of an avascular area appearing in the center of an area of scleritis, which then gets broken down to form a slough eventually leaving an area of thinning. Possible misdiagnosis as bilateral conjunctivitis with eye discharge may be made as in this case. The discharge may have arisen from the slough from the area of tissue thinning. This is a possible error during the early stages of the disease and should be avoided. Episcleritis is another possible differential diagnosis. This patient was also misdiagnosed as a case of episcleritis at presentation.

There is a strong association with immunological diseases in this condition though our patient did not have markers of such. Rheumatoid factor and Anti-Nuclear Antibody are recognized markers of immunological diseases, but both were negative in our patient. Despite this timely intervention with systemic therapy rapidly resulted in the cessation of ocular pain.

Severe ocular pain unresponsive to painkillers have been associated with scleritis [5], our patient confirmed this. Also, cessation of ocular pain is an indication of improvement as seen in our patient. Being pain free is a positive consequence of treatment.

CONCLUSION

Anterior necrotizing sclero-uveitis may present to an ophthalmologist practicing in our region. Early recognition, investigations and management with immunosuppressive and steroids may be life and vision sparing. Topical steroids can be delayed if there are concerns regarding its use earlier on in the disease; but can be introduced as the systemic situation comes under control. Outcome of treatment can be rewarding, with recovery of vision and a cessation in the progression of scleral thinning.

REFERENCE


Author Contributions

Ogugua Ndubuisi Okonkwo – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

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Conflict of Interest
Authors declare no conflict of interest.

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