Case Report

Management of Perforated Mooren's Ulcer with a Rotational Scleral Autograft in Abuja

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Abstract

Mooren's ulcer is a fairly common peripheral corneal melting disease in Nigeria. Cases presenting late with perforation and uveal prolapse are a serious challenge to management, particularly in a setting where corneal grafting is not routinely practiced. To describe the management of perforated Mooren's ulcer using scleral rotational autograft. A case of bilateral Mooren's ulcer with corneal melting and uveal prolapse in the left eye (LE) involving 1½ h was managed with a scleral rotational autograft at the Rachel Eye Center, Abuja. The patient was a 75-year-old female retiree with concomitant rheumatoid arthritis involving the knee and metacarpophalangeal joints. Following a 360° peritomy and cryotherapy in both eyes, a partial thickness scleral hinged autograft was developed and rotated over the uveal prolapse and corneal defect whereupon, it was fastened with interrupted 10/0 nylon sutures and secured with a bandage soft contact lens. The integrity of the globe was maintained for over 6 months with the scleral flap which became largely transparent from the 13th day after the surgery. The anterior chamber went flat temporarily, but this was reversed with firm pad over a contact lens. This technique is recommended as at least a stopgap before the availability of corneal grafts.

Key words: Corneal melting, cryotherapy, Mooren's ulcer, peritomy, rotational flap, scleral autograft

Introduction

Mooren's ulcer is a rare, painful marginal melting ulcer of the cornea, which was first described by a German Ophthalmologist Albert Mooren (1829-1899) in the year 1867.[1]

It presents as a painful keratoconjunctivitis with typical guttering of varying segments of the peripheral cornea.[2] It has been reported in Nigeria by various authors.[3,4]

From his observations, Kietzman[3] surmised that there were possibly two distinct epidemiological populations: An older age group affected by a mild unilateral form of the disease which often responded to therapy, and a younger age group between 20 and 30 years of age, presenting with aggressive form of the disease, unresponsive to therapy. However, Lewallen and Courtright in a meta-analysis[5] are of the opinion that currently available data does not support this generalization.

The natural history of Mooren's ulcer is that it may heal spontaneously or it may progress centrally but may sometimes perforate often with consecutive endophthalmitis and phthisis bulbi.[6]

Access this article online

Quick Response Code:

Website: www.nigerianjournalofophthalmology.com

DOI: 10.4103/0189-9171.195203

The pathogenesis is thought to be related to the production of collagenases by the subjacent conjunctival tissue, with autoimmune underpinnings such as deficiency in circulating suppressor T-cell populations.[7,8] The production of autoimmune antibodies to corneal stroma (so-called corneal associated antigen or Co-Ag) with cross reactions to hepatitis C virus had been demonstrated by Gottsch *et al*.[9]

The management of unperforated ulcer has often been along the lines of topical steroids, antibiotics, analgesics, and antimetabolites such as methotrexate[3] and cyclosporine.[10]

The use of combined cryotherapy and peritomy was advocated by Aviel in Blantyre, Malawi, in 1972.[11]

The management of perforated ulcer is more problematic and challenging. Small perforations have been treated with tissue

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How to cite this article: Babalola OE. Management of perforated mooren's ulcer with a rotational scleral autograft in Abuja. Niger J Ophthalmol 2016;24:80-3.

adhesives, whereas larger perforations have been treated with crescent- shaped corneal graft, tectonic grafts, and free lamellar scleral patch grafts particularly in perforated measles ulcers.[12-15]

We carried out a scleral rotation flap in a case of bilateral Mooren's ulcer with perforation in one of the eyes. This procedure was necessitated by the need to save an eye at risk of loss to endophthalmitis. We hereby report on our experience with this procedure.

Case Report

The patient OO is a 71-year-old female retiree, who presented to us on August 5, 2015. She had seen a "growth" in the LE for 2 weeks. There was no history of trauma.

The patient suffers from painful multijoint rheumatoid arthritis particularly involving both knee joints as well as the joints of both hands. She is as a result wheelchair bound.

On general examination, the patient appeared distressed about her situation. She had swollen knees and metacarpophalangeal joints bilaterally [Figure 1].

On ocular examination, her visual acuities right/left were 6/12+ and 1/60.

In the right eye (RE), some thinning and guttering were visible in the peripheral cornea between 2 and 5 o'clock position with a descemetocele in the 2.30 position. A pterygium was apparent nasally amidst moderate congestion of the eye [Figure 2].

In the LE, the congestion was more apparent. A perforation of the cornea at the 6–7.30 o'clock position, about 1½, with uveal prolapse was evident [Figure 3]. There was additional guttering between 2 and 6 o'clock temporally and between 11 and 7.30 o' clock nasally.

Based on the characteristic peripheral guttering and thinning of both corneas, a working diagnosis of Mooren's ulcer was entertained.

The differentials include Terrien's marginal ulcer, peripheral ulcerative keratitis, and rheumatoid corneal melting disease.

A decision was made to carry out a 360° peritomy and cryoapplication in both eyes. The procedure was carried out by the author. The peritomy preceded the cryoapplication. The integrity of the globe in the LE was secured by carrying out a hinged rotational scleral flap consisting of a 5 mm by 5 mm half thickness flap of sclera subjacent to the corneal defect.

The procedure was carried out under local anesthesia (retrobulbar injection of 2% lidocaine with adrenaline plus sub-Tenon's infiltrations). The flap was secured with interrupted 10/0 nylon sutures. Eight sutures were applied in all. Six to the leading edge of the rotated flap and two to repair an inadvertent tear in the flap [Figures 4 and 5]. A bandage soft contact lens was placed in both eyes.

Patient was on admission for the next 8 days placed on guttae dexagent, ciproxin, and tropicamide. In addition, tablets ascorbic acid 2 daily, diamo × 250 mg b.d., and tylenol 2 t.i.d. were prescribed. The lacrimation and discomfort reduced markedly over the period of admission.

The patient became more comfortable over the next few weeks and the eyes quietened down. As shown in Figure 6, on the August 18, 2015 (i.e., 13 days after the autograft), the transposed scleral became transparent enough to show the uveal tissue. Gutt nepatop (nepafenac 0.1%) and tears naturale were added to the treatment.

On the 26th of September, significant vascularization was noticed over the graft of the LE and near to the descemetocele in the RE. On the 3rd of October, intracameral Avastin injection was administered in both eyes and the sutures were removed. Visual acuity was 6/6 RE corrected, and 6/60 LE corrected. The situation remained stable for 14 weeks.

However, on the clinic visit of November 21, 2015, 14 weeks after surgery, the AC went flat in the LE. There were no clear antecedents before the flattening of the anterior chamber but

Figure 1: Rheumatoid arthritis affecting the metacarpophalangeal joints of both hands and the knee joints of both legs.

Figure 2: Right eye of the patient with evident guttering of the peripheral cornea. There is an inflamed pterygium partially obscuring the guttering.

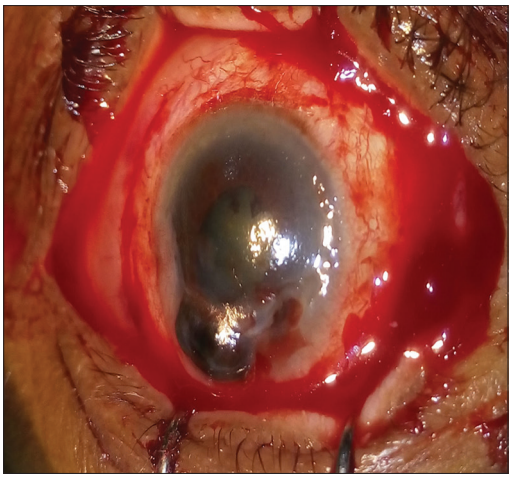

Figure 3: Left eye of the patient with prolapse of the uvea in associated with guttering and melting of the peripheral cornea. This picture was taken during the surgery after the 360° peritomy and before the cryotherapy and scleral flap was raised.

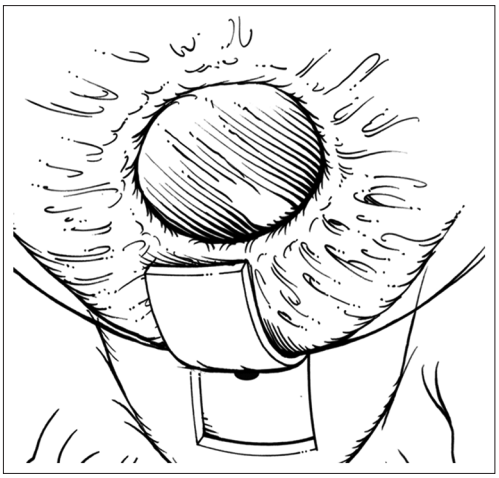

Figure 4: Figure indicating how hinged half thickness scleral flap is raised to cover the area of prolapse.

Figure 5: The scleral flap secured in place and covered with a contact lens (August 5, 2015).

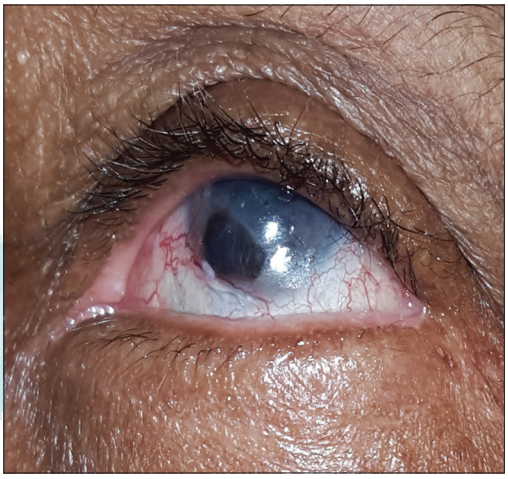

Figure 6: Thirteen days postoperative, the scleral transplant has become transparent, and the uveal prolapse is secured behind the flap and contact lens. Visual acuity is 6/60 uncorrected.

possibly the contact lens fit was not tight enough. On close examination, it was clear that the transposed sclera was still largely intact, but there appeared to be a small breach in its integrity at about 4.30 position 3 mm from the limbus. This situation was remedied by fitting a tighter contact lens and applying firm pad and plaster. The AC reformed after 2 weeks. The patient had a recurrent mucopurulent discharge, possibly a reaction to the contact lens, which was managed with topical antibiotics and nonsteroidal anti-inflammatory drops such as diclofenac. The patient is currently stable.

Discussion

Mooren's ulcer is rare but not altogether uncommon. In our practice, where we see about 2000 new patients every year, we tend to see two to three cases every year. This gives a hospital yearly period prevalence of 1/1000 in the Abuja Area. What is unique about this case however was the age of the patient (71), bilateral nature of the disease, and the perforation at presentation. According to the observations of Kietzman,[3] bilateral severe disease was more common among the young male population in Nigeria. This case was therefore clearly an exception to that rule.

As was evident from the photographs, the patient was wheelchair-bound by advanced and chronic rheumatoid arthritis for which she was on occasional nonsteroidal anti-inflammatory drugs only. This involved both the large joints of the knee and the small joints of the hands, specifically the metacarpophalangeal joints. There was a striking symmetry to the joint involvement which argued more in favor of rheumatoid rather than osteoarthritis.[16] Furthermore, the association between rheumatoid arthritis and Mooren's ulcer is well established.[17] This patient was managed with 360° peritomy and cryoapplication.[16,17] Other workers have utilized peritomy restricted to the extent of the ulcer, but in this case, we carried out a 360° peritomy because we felt this

would better preempt the spread of the ulcerative lesions. This has been the practice with other authors, some of who have also used amniotic membrane and 20% autologous serum in refractory cases.[18]

Several authors have utilized scleral patch grafts to salvage corneal perforation as we did in this case.[19,20] However, most authors have not used a hinged flap. Possibly, the hinged nature of the flap ensures continuous blood supply to the flap and may help to ensure viability.

In Nigeria, cadaver corneal donations are not readily available and the practice is just beginning to be encouraged. Therefore, corneal grafts may need to be imported from overseas though donations from philanthropic organizations. The process of importation of the tissues can be time-consuming. When these donations are available, they tend to be very expensive and may not be affordable to most patients in the country. Therefore, it may be necessary to take remedial measures at least on the short-term.

The use of rotational scleral flaps in the management of perforated corneal ulcers has been described by other Nigerian authors,[14] particularly in the management of perforated Measles ulcers. Measles is however not as common as it was in the past, largely due to continuing though not universally successful immunization programs embarked upon by the government.[15]

Our procedure was envisaged as a temporary measure pending the time when the patient would be able to afford to pay for a proper corneal graft. However, this has not eventuated.

Several workers[21-26] have reported on the fate of sclera transplanted into cornea, particularly on the transition into transparency. This occurs with both autografts and allografts. This transition into transparency has been experimentally observed in rabbits to take place slowly over several months.[26] Electron microscopic observation indicated that the collagen fibrils of the sclera broke down by unraveling into smaller fibrils similar in diameter to those of the cornea. Both active macrophages and fibroblasts could be found in the transitional area. This process can be described as an induction of transparency by contiguity to clear cornea.

In the case under review, however, it was observed that the transplant cleared up much faster than reported elsewhere, i.e., by 13 days after surgery.

In summary, the patient under review is a 70-year-old woman with bilateral Mooren's ulcer, perforated on the right, and treated with 360° peritomy and cryotherapy in both eyes. In addition, a rotation scleral autograft was carried out in the RE. The eyes have held for 6 months of observation; so far and we have been surprised by the rapidity of transparency in the grafted tissue. The method of hinged rotation scleral graft is recommended for perforated ulcers in which corneal grafts are not readily available.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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