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The Thin Hot Eyes: A Case Series of Corneal Melting

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Abstract: This case series report on three cases of adults presented with corneal melt secondary to multiple aetiologies and challenges in its management. Case 1 and 3 were a sero-positive rheumatoid arthritis (RA) and atypical painless Mooren's ulcer (MU) respectively that presented with corneal perforation, while, case 2 was a unilateral MU patient with aggressive corneal melting. Case 2 and 3 had previous history of ocular surgery. All patients showed progressive corneal melting despite of intensive medical immunosuppressive therapy. Penetrating keratoplasty (PK) was performed for case 1 and lamellar keratoplasty for case 2 and 3. Post operatively, the visual acuity ranged from 1/60 to 6/18. Corneal melting is a vision threatening condition that usually has an underlying systemic aetiology. A systemic workup is crucial in its diagnosis and treatment. The rapid course of the disease and unpredicted response to treatment make every case a challenge in its management.
Keywords: Corneal melting, Peripheral ulcerative keratitis, Rheumatoid arthritis, Mooren's ulcer.

INTRODUCTION

Corneal melting or keratolysis occurs most commonly in association with systemic immune diseases. Collagen vascular diseases are found in 50% of non-infectious peripheral ulcerative keratitis (PUK); RA is the most prevalent [1,2]. It may also be associated with inflammatory causes, infection or occurred following ocular surgery [3].

In contrast, MU have similar appearance and affect the peripheral cornea similar as PUK, however, it is not associated with any systemic immune diseases. In addition, MU never invades adjacent scleral tissue with characteristic features of ulcer with an overhanging inner edge [4].

CASE DESCRIPTION

CASE 1

A 60 years old Indian lady presented with one week history of ocular pain, tearing and progressive deterioration of vision in her right eye. She was diagnosed with sero-positive RA for the past 10 years which was complicated with pulmonary fibrosis. She was on regular follow up under rheumatology team and was previously on oral sulfasalazine 1g twice daily.

Her visual acuity at presentation was counting finger at 1 metre OD and 6/12 OS. The slit lamp examination of the right eye revealed central corneal perforation measuring 2.1mm x 2.1mm in dimension with plugged iris and surrounding stromal infiltrates. The anterior chamber was shallow with irido-corneal touched (Figure 1). The left eye showed multiple punctate epithelial erosion with early nuclear sclerosis cataract and the fundus examination was unremarkable.

She underwent an immediate right cyanoacrylate corneal patch graft upon admission followed by PK which was performed within two days of admission. She was initiated on oral prednisolone 40mg once daily, oral cyclosporine 50mg bidaily, topical Minims® Dexamethsone 0.1% every four hourly for the right eye and her oral sulfasalazine was continued. Prophylactic antibiotic along with oral doxycycline 100mg twice daily and vitamin C 1g once daily were also started. Post-operatively, she developed persistent corneal melt at the cornea host junction tissue on day 5 and 6 weeks post-PK. Pulsed intravenous (IV) methylprednisolone 500mg bidaily was commenced for 3 consecutive days and was then transitioned to oral prednisolone of 1mg/kg/day with weekly tapering dose. She responded well with the intensive immunosuppressive therapy with no other episode of cornea melt. However, she developed acute renal impairment and acquired pneumonia along with fungal keratitis secondary to oral cyclosporine usage. Her renal function and infection improved within two weeks of cyclosporine cessation. The oral cyclosporine was replaced with oral azathioprine 50mg bidaily and

topical cyclosporine 0.05% four times daily on both eyes. At one year post PK, her best corrected visual acuity (BCVA) of the right eye was 6/36 with no signs of cornea melt (Figure 2).

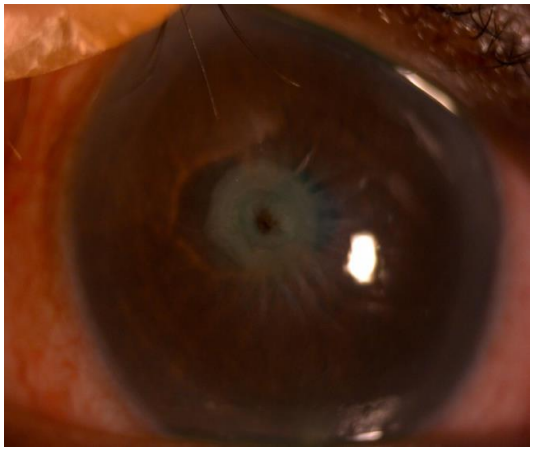

Fig-1: Right central corneal perforation with shallow anterior chamber

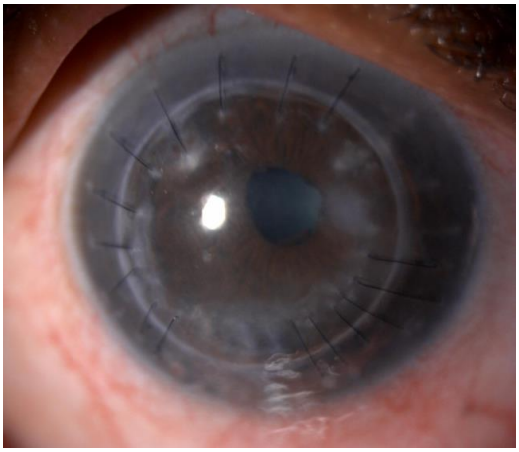

Fig-2: One year post right penetrating keratoplasty

CASE 2

A 44 years old Malay gentleman with no known medical illness presented with history of progressive visual loss in the left eye associated with ocular pain and redness over a year. He started to develop the symptoms six months following pterygium excision done at another local hospital and was unsure of mitomycin C usage during procedure. He denied any history of ocular trauma, symptoms of autoimmune diseases or other associated diseases.

His BCVA on presentation was hand motion OD and 6/6 OS. Anterior segment examination of the left eye revealed a crescent shaped 270 degrees peripheral corneal thinning and melting extending from 4 to 1 o'clock (Figure 3). It was positive for fluorescein stain superonasally indicative of the overlying epithelial defect. The overhanging edge and remaining cornea was oedematous. The adjacent conjunctiva was also injected. Examination of the right eye revealed normal ocular findings.

He was investigated to rule out systemic diseases causing peripheral ulcerative keratitis (PUK). All his relevant investigations were negative including anti cytoplasmic antibody (ANCA), rheumatoid factor (RF), antinuclear antibody (ANA) and hepatitis C screening. Other haematological parameters such as full blood count, renal profile and liver function test were also within normal limit. The inflammatory markers were elevated with erythrocyte sedimentation rate (ESR) of 22mm/ hour, while his was C-reactive protein (CRP) 0.63g/dl. Based on the clinical findings and workups, a diagnosis of MU was made.

Systemic immunosuppressive therapy was initiated with IV methypredisolone 500mg once daily for three days followed by oral azathioprine 50mg TDS and prednisolone of 1mg/kg/day with weekly tapering dose. He was also started on topical prednisolone acetate 1% four times daily, topical moxifloxacin every 4 hourly, oral doxycycline 100mg twice daily and vitamin C 1g once daily. However at three months after the initial presentation, the cornea melt showed progression to 360 degree peripheral involvement with

Central island of oedematous cornea and extension of the epithelial defect. Conjunctival resection was then performed. Despite of these, his cornea continued to progressively melt. He underwent tectonic lamellar keratoplasty (LKP) (corneo-scleral graft) under general anaesthesia to prevent further progression. Four month post-operatively, his BVCA ranged from 1/60 to counting fingers and showed stable disease activity (Figure 4).

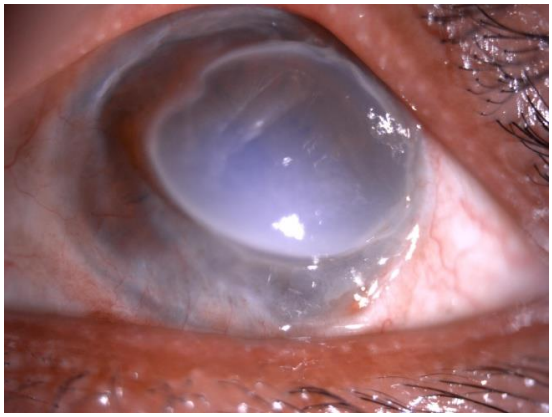

Fig-3: Left crescent shaped peripheral corneal thinning and melting with overhanging edge

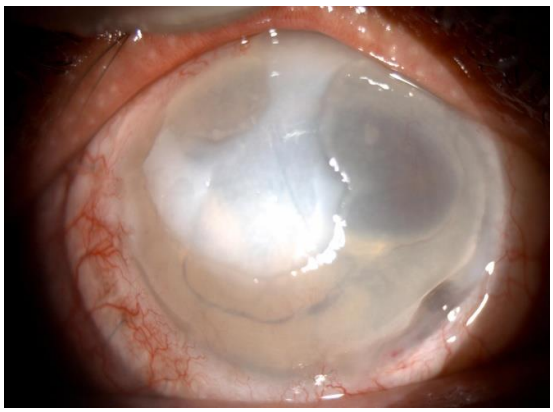

Fig-4: Post four months left lamellar keratoplasty (corneoscleral graft)

CASE 3

A 40 years old, Malay gentleman with underlying diabetes mellitus, hypertension and history of resolved Steven Johnson Syndrome (SJS) 10 years ago, presented with left eye painless progressive deterioration of vision associated with redness for one year duration. He had bought over the counter topical steroid drop regularly to relieve the symptoms without ophthalmologist consultation. He had history of bilateral myopic laser refractive surgery done three years prior to presentation and his vision was initially good postoperatively.

Visual acuity at presentation was 6/24 OD and counting finger OS. Anterior segment examination showed a crescent-shaped peripheral cornea thinning nasally with undermined edges extending from 1 to 8 o'clock in the right eye and 6 to 12 o'clock with limbal involvement in the left eye but no associated scleritis. There was a slow leak over the edge of peripheral cornea thinning on the left eye and the anterior chamber was deep (Figure 5). There were no signs of secondary infection. Other ocular and systemic examination was unremarkable.

All relevant investigations to rule out causes of PUK were negative except for his inflammatory marker which was moderately elevated with ESR of 46mm/ hour and CRP of 1.24mg/dl. Diagnosis of MU was made based on the clinical findings and negative workup for underlying systemic diseases.

He was initiated on systemic immunosuppression of IV methylprednisolone 500mg once daily for three days along with oral cyclosporine 500mg bidaily. The methylprednisolone was transitioned to oral prednisolone of 1mg/kg/ day with weekly tapering dose. He was also started with topical prednisolone acetate 1% four times daily and topical moxifloxacin every 4 hourly on both eyes along with oral ciprofloxacin 500mg twice daily and oral doxycycline 100mg twice daily. He underwent left peripheral corneal banana patch graft to seal the linear perforation along the edge of the cornea thinning. Post-operatively, his visual acuity of the left eye improved to

6/18. However, he developed acute renal impairment and hepatitis secondary to cyclosporine usage, which was then changed to oral mycofenolatemofetil 500mg bidaily. His renal and liver function improved within one month of cyclosporine cessation. One year post-operatively, he BCVA remains stable with no signs of recurrence (Figure 6).

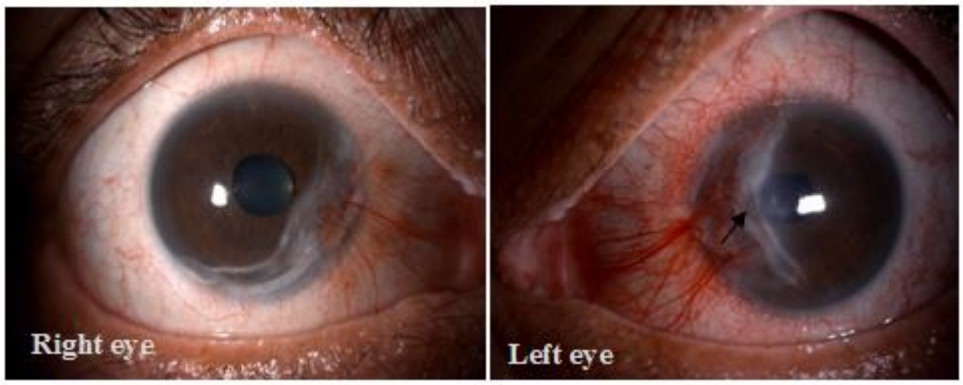

Fig-5: Bilateral crescent-shaped peripheral cornea thinning nasally with undermined edges with perforation on the left eye (arrow)

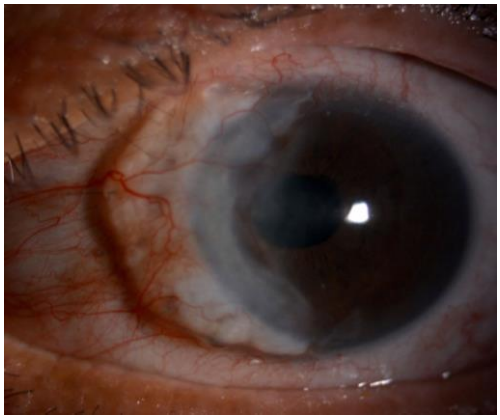

Fig-6: Post one year left peripheral corneal banana patch graft

DISCUSSION

Corneal melting is a common prelude to the development of corneal perforation. This case series illustrate the different aetiologies of immune-mediated corneal melting and the challenges in its management to halt the disease progression and prevent its recurrence.

Clinical presentation of PUK in RA is variables. Corneal melting in RA occurs late in the disease course with a mean age of onset 19.6 years after diagnosis [5]. In our first case, the patient presented with PUK after ten years of diagnosis. The serious ocular complications in case of RA with PUK presents challenges to the ophthalmologist at various stages in the management. The primary treatment for acute disease control is systemic corticosteroid and patient with imminent visual loss, pulsed methylprednisolone may be initiated [6, 7]. Corticosteroid alone unable to halt the disease activity and immunomodulatory agent should be started concurrently. Furthermore, these agents have shown to prolong corneal graft survival rate [7, 8]. Surgical intervention is warranted in case of corneal perforation to preserve structural integrity of the globe. Nobe *et al*. reported PK in patient with PUK has shown to have high failure rate, most commonly due graft melt from recurrence of PUK [8]. Studies have shown that many patients require multiple grafts with 20 to 40% graft survival at 6 months [8, 9]. In relation to our first case, patient underwent PK and showed recurrence of corneal melting post operatively which responded to the intensive immunosuppression therapy. She was not subjected for methothrexate, although it is the first line of immunomodulatory agent in RA associated PUK [2], in view of its side effect causing pulmonary toxicity which may complicate her underlying pulmonary fibrosis.

MU is an idiopathic, progressive and painful ulcerative keratitis that occurs in the absence of any systemic diseases. The exact aetiology of MU is unkown, however, there are evidences suggesting that it is an autoimmune basis. Zegans *et al*. found that previous history of corneal surgery, trauma or infection was reported in 68% of patient with MU [11]. Both of our patients in second and third case had history of ocular surgery that predisposed them to development of

MU. In addition, history of SJS in our third case may result in conjunctival cicatrisation which creates a hostile ocular surface environment which may be responsible for triggering the autoimmune mediated keratolysis.

The main approach and goal in the management of MU is to arrest the destructive immune process and promote healing of the corneal surface. Most clinicians advocate on the 'step ladder approach' in the treatment of MU. Initial treatment may include local and pulsed systemic corticosteroid therapy during the acute phase. However, if corticosteroid therapy fails to control the disease progression, conjunctival resection should be performed. The use of second line immunosuppressive agents is reserve in cases of bilateral or resistant MU to halt the progressive immune mediated keratolysis [12]. Madino et al reported that the progression of MU in 4 of 13 patients who did not respond to topical corticosteroids or conjunctival resection was halted after second line immunosuppressive therapy initiated [13]. Both of our patients (case 2 and 3) with MU, however, failed to response to the aggressive immunosuppressive therapy initiated. The cornea in both cases was progressively melting which warrant for surgical intervention. A tectonic LKP (corneo-scleral graft) was performed for the patient in the second case due to presence of extensive peripheral corneal melting with limbal involvement. As for the third case, a peripheral corneal banana patch graft was done to seal the linear perforation along the edge of the cornea thinning. LKP offers several advantages over PK in PUK patients with perforation or impending perforation. The risk of rejection is greatly reduced for lamellar graft. It also permits the preservation of maximum amount of host tissue and adds on to the cornea thickness. This reduces the risk of recurrent perforation if the corneal melting process reactivates [14].

CONCLUSION

Corneal melting is a vision threatening condition that usually has an underlying systemic aetiology. A systemic workup is crucial in its diagnosis and treatment. The rapid course of the disease and unpredicted response to treatment make every case a challenge in its management.

Competing interest

The authors declare that they have no competing interests.

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