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Photo Essay

Aggressive Mooren's Ulcer and challenges in its management Tale of three patients in pictures

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A 63 years old male patient from north India presented to the Cornea Services at our center with pain and redness in both eyes, which had gradually worsened over a period of last 6 months. The pain was accompanied with diminution of vision, watering and photophobia in both eyes. There was no history of ocular trauma, intraocular surgery or any systemic illness. Ocular examination revealed uncorrected visual acuity (UCVA) of counting fingers at 1 m (with accurate projection of rays) in the right eye and hand motion close to face (with accurate projection of rays) in the left eye, which did not improve with refraction. Intra ocular pressures could not be measured due to photophobia. Slit lamp examination (Haag-Streit BM 900®) in the right eye showed peripheral descematocele with corneal ulceration extending from 4o'clock to 8o'clock (Fig. 1a). The anterior chamber was quiet and immature senile cataract was noted. Fundal glow was present but the details of posterior segment could not be appreciated. The left eye, showed peripheral descematocele with corneal ulceration extending from 8o'clock to 11o'clock (Fig. 1b). Anterior chamber examination showed mature senile cataract with no view of the posterior segment. Conjunctival injection was present in both eyes and sclera remained uninvolved. The patient was taken up for emergency conjunctival resection and patch graft for both eyes under high dose intravenous steroids (pulsed intravenous methylprednisolone 500 mg daily for 3 days). A free hand fashioned lamellar thickness cresentic customized corneal graft of therapeutic quality was used for both eyes (Fig. 1c and d). The patient was put on hourly topical steroids (1% prednisolone acetate), six hourly antibiotic (0.5% moxifloxacin) and cycloplegics (2% homatropine). The patient maintained stable uncorrected vision of 20/200 in the right eye (Fig. 2a), however the condition of his left eye deteriorated over the next 3 months (Fig. 2b). There was 360° peripheral involvement of the cornea, mature senile cataract and shallow anterior chamber (Fig. 2c). To control the aggressive corneal involvement and to visually rehabilitate the patient, triple procedure (penetrating keratoplasty, cataract extraction and intraocular lens implantation) was planned for the left eye. This was combined with high dose intravenous cyclophosphamide (50 mg/kg) 1 day prior to the surgery and one day after the surgery. A final postoperative uncorrected visual acuity of 20/100 was achieved. The patient did not improve with refraction, and was content with good ambulatory vision (Fig. 2d). He was kept on long-term immunosuppressives, comprising of oral azathioprine 50 mg twice a day.

Our second patient was a 61-year-old male who presented to us with gradually decreasing visual acuity in his right eye over a period of 6 months. The visual loss was accompanied with pain and watering. There was no history of trauma or any significant systemic ailment. The visual acuity in the right eye was finger counting close to face (with accurate projection of rays). Slit lamp examination revealed a large descematocele from 10o'clock till 1o'clock. The anterior segment was normal with immature senile cataract (Fig. 3a and b). Like the previous patient, there was no scleral involvement. The patient was taken up for conjunctival resection with patch graft under intravenous steroids (methylprednisolone 500 mg daily for 3 days). A lamellar thickness cresentric graft was fashioned and grafted using 14 10-0 Nylon suture. At 3 and 9 weeks follow up, the cornea remained stable with acceptable scarring and a patent anterior chamber, with the patient having a vision of 20/200 in the right eye (Fig. 3c and d). Similar to the first patient, this patient was also kept on long-term oral Azathioprine 50 mg twice a day.

Our last patient was a 15-year-old boy who presented with 1-month history of pain, redness, photophobia, and diminution of vision in the right eye. His BCVA in the right eye was finger counting at 1 metre and in left eye was 20/20.

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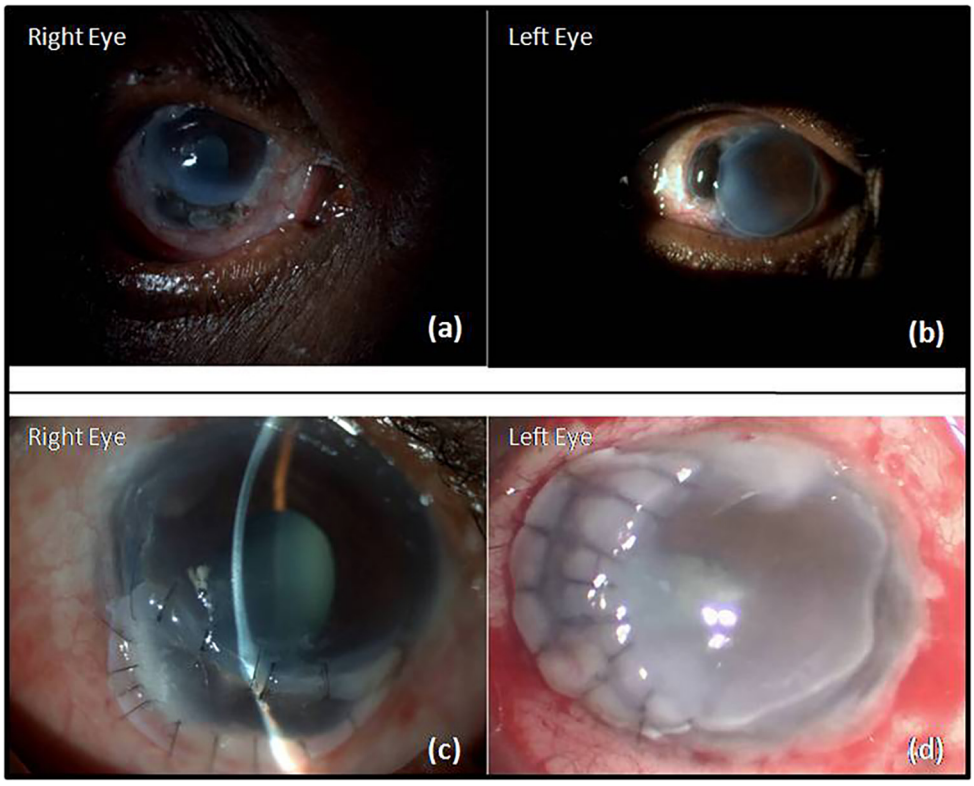

Fig. 1. (a) Slit Lamp photo of the right eye at presentation showing peripheral descematocele with corneal ulceration extending from 4o'clock to 8o'clock. (b) Slit Lamp photo of the left eye at presentation showing peripheral descematocele with corneal ulceration extending from 8o'clock to 11o'clock. (c and d) Slit lamp photo of both eyes after patch graft.

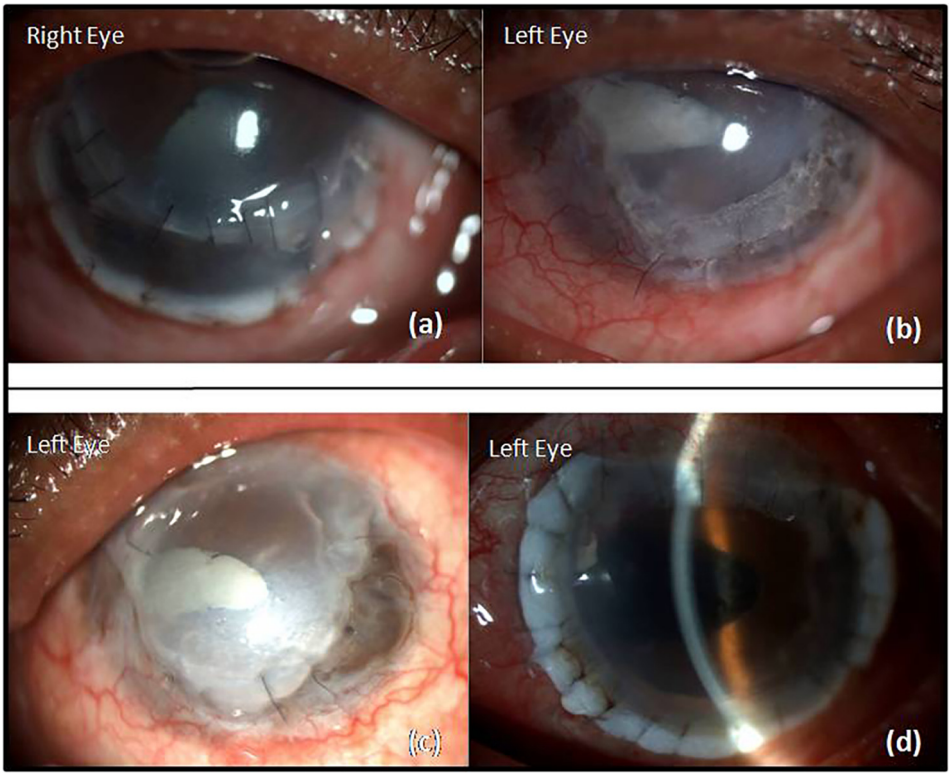

Fig. 2. (a) Slit lamp photo of the right eye 3 months post patch graft. (b) Slit lamp photo of the left eye 3 months post patch graft. (c) Slit lamp photo of the left eye showing 360° peripheral involvement of the cornea, mature senile cataract and shallow anterior chamber. (d) Status post triple procedure (penetrating keratoplasty, cataract extraction and intraocular lens implantation) in the left eye.

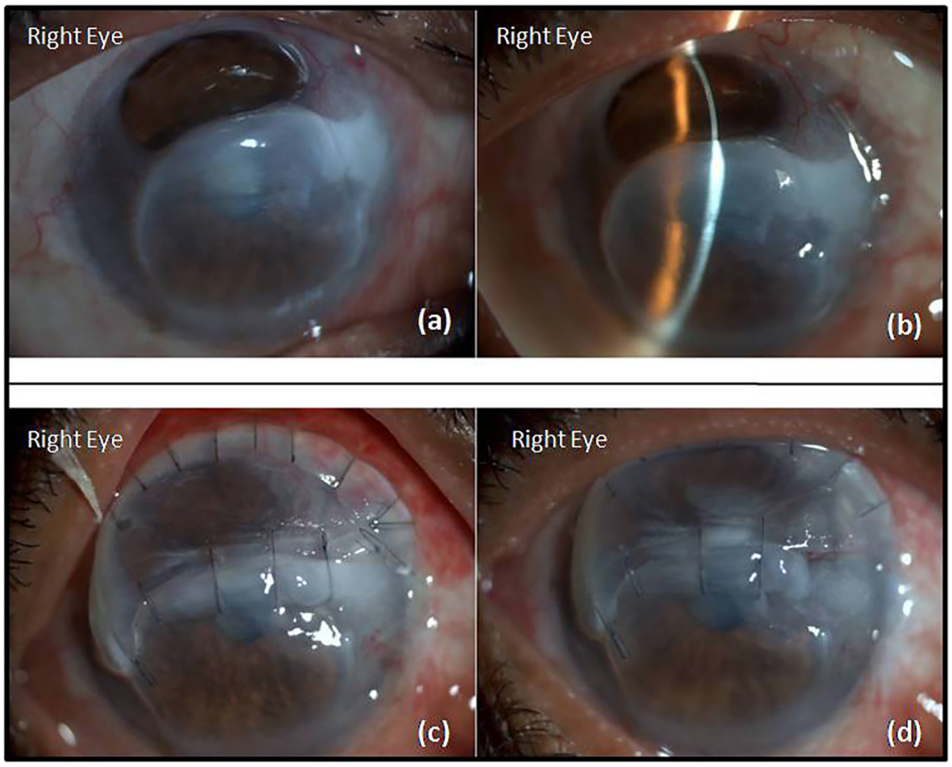

Fig. 3. (a and b) Slit lamp photo of the right eye showing large descematocele from 10o'clock till 1o'clock. (c and d) 3 and 9 weeks photo of the right eye post cresentric patch graft.

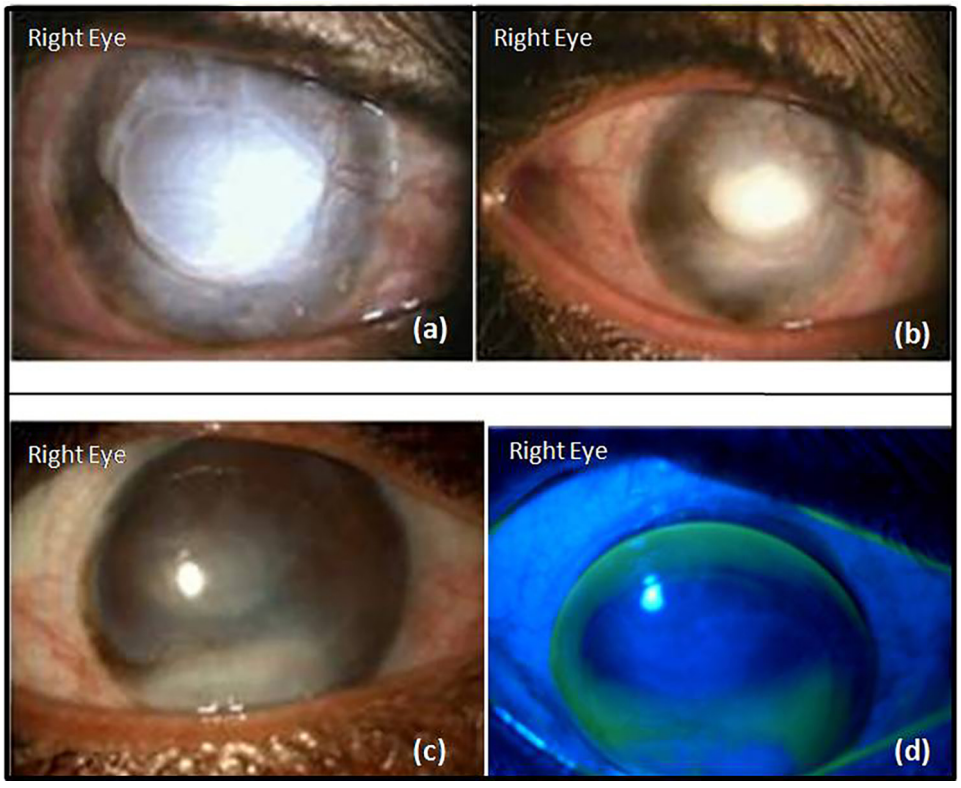

Fig. 4. (a) Acute corneal hydrops (Grade 2) in resolving Mooren's Ulcer. (b and c) 1 week and 2-month post C3F8 injection, resolving corneal edema is seen. (d) Rose-K IC (Irregular Cornea) contact lens in the right eye.

Slit-lamp examination of the right eye showed peripheral corneal ulceration extending from 2o'clock to 10o'clock position with cellular infiltration and a central overhanging edge. Iris incarceration with pseudocornea formation was noted inferiorly in an area of 2o'clock hours. Fundus was not visible in the right eye. Conjunctival resection along with tissue adhesive (TISSEEL Kit from Baxter AG, Vienna, Austria) and bandage contact lens (Purecon™, size 18 mm) placement was performed. He was treated with systemic as well as hourly topical steroids. One month later, he presented with acute onset diminution of vision in the right eye. The BCVA dropped to perception of light. On slit-lamp examination, there was mild conjunctival congestion with marked corneal edema (central 5 mm) and peripheral cornea scarring as previously. A diagnosis of acute corneal hydrops (Grade 2) in a resolving Mooren's ulcer was made (Fig. 4a). Intracameral injection of 0.1 ml of nonexpansile perflouropropane (14% C3F8) gas was done through a 30-gauze needle under topical anesthesia. 75% fill of the anterior chamber was achieved. On postoperative examination, the central corneal edema started resolving after 1 week (Fig. 4b). Two months later, the edema completely resolved and there was central corneal scarring of macular grade (Fig. 4c). He was fitted with Rose-K IC (Irregular Cornea) contact lens in the right eye and BCVA of 20/80 was achieved (Fig. 4d).

Comment

Mooren's ulcer is a rapidly progressive, painful, ulcerative keratitis which initially affects the peripheral cornea and may spread circumferentially and then centrally.[1] It occurs completely in absence of any diagnosable systemic disorder that could be responsible for the progressive destruction of the cornea and the aetiology remains uncertain.[2] It has been related to trauma, corneal foreign bodies, acid burns, metabolic disturbances, vitamin deficiency, fifth nerve changes, periarteritis nodosa, Wegener's granulomatosis, local nutritional disturbances, hookworm infestation, syphillis, tuberculosis and virus.[3–10] Through this series, we would like to highlight the various ways in which mooren's ulcer can present and various options of managing these patients. However, all the three patients had few features in common-presence of pain, absence of scleral involvement, normal systemic workup (Complete blood counts, Rheumatoid Factor, Anti Nuclear Antibodies, C Reactive Proteins, Erythrocyte Sedimentation Rate) and male preponderance. Also, all the patients responded well to treatment, highlighting the importance of quick, aggressive and decisive treatment strategies that need to be undertaken.

Conflict of interest

The authors declared that there is no conflict of interest.

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