Microsporidial infection masquerading as graft rejection post-Descemet's stripping automated endothelial keratoplasty

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A 51-year-old immunocompetent male with a history of Fuchs' endothelial dystrophy and immature cataract who underwent Descemet's stripping automated endothelial keratoplasty with intraocular lens implantation in both eyes presented with redness and defective vision of 1-day duration in his left eye. Slit lamp examination revealed coarse superficial punctate lesions with graft edema. He was diagnosed with acute graft rejection and treated with topical steroids. Two days later, symptoms worsened in his left eye with the involvement of his right eye showing a similar clinical picture. An infectious etiology was suspected and *in vivo* confocal microscopy ordered, which revealed hyperreflective dots, highly suggestive of microsporidial spores. The patient was prescribed topical fluconazole 0.3% in both eyes. This unique presentation of bilateral graft edema following microsporidial keratoconjunctivitis in postgraft patients requires a high index of suspicion as it can be easily be mistaken for and mismanaged as acute graft rejection.

Key words: Descemet's stripping automated endothelial keratoplasty, graft rejection, *in vivo* confocal microscopy, microsporidial keratoconjunctivitis

Ocular microsporidial infection has increasingly been reported in the recent past because of increased awareness and emerging trends in its diagnosis and management. The occurrence of microsporidial keratoconjunctivitis in a corneal graft, though relatively rare, has been reported.[1] We present a challenging case of bilateral microsporidial keratoconjunctivitis initially misdiagnosed and managed as graft rejection in a patient who had recently undergone Descemet's stripping automated endothelial keratoplasty (DSAEK) in his left eye and was status post-DSAEK with intraocular lens (IOL) implantation in the right eye since 2012 for Fuchs' endothelial dystrophy.

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Case Report

A 51-year-old immunocompetent male who had undergone DSAEK with IOL implantation in his left eye 7 weeks prior presented with complaints of redness and defective vision in his eye for 1 day. He was on routine postoperative medications with topical prednisolone acetate 1%, and gatifloxacin 0.3% eye drops, each to be administered three times daily. At his previous postoperative visit, 2 weeks earlier, his left eye had been quiet with a clear graft and normal intraocular pressure.

In his right eye, he had undergone DSAEK with IOL implantation in 2012 and was doing well. On examination at this visit, UCVA was 20/40 in the right eye and 20/200 in the left eye. Slit lamp examination of the left eye revealed moderate bulbar and tarsal conjunctival injection with papillae, a few randomly distributed coarse superficial punctate lesions, graft edema with overlying stromal edema, and no keratic precipitates. There was a mild anterior chamber reaction. A presumptive diagnosis of acute graft rejection was made in the left eye. He was admitted and aggressively managed with hourly topical prednisolone acetate 1%. He was also started on systemic steroids injection dexamethasone 2 cc intramuscularly once a day. Next day slit lamp examination of the left eye showed an increase in the corneal edema to the host stroma, involving the peripheral cornea as well. There was moderate anterior chamber reaction but no keratic precipitates. Visual acuity remained at status quo in both eyes. He was continued on the same treatment. On day two of admission, the patient complained of defective vision in his previously operated right eye and on examination, the visual acuity was found to be 20/80. The right eye showed a mild conjunctival injection, diffuse edema of the entire cornea and a few coarse superficial punctate lesions similar to the lesions seen at presentation in the left eye [Fig. 1a]. Left eye examination revealed a large epithelial defect approximately 6 mm × 4 mm in diameter with persistent graft and stromal edema [Fig. 1b]. The typical course superficial punctate lesions at presentation, deteriorating clinical picture, and poor response to topical steroids in the left eye with diffuse corneal edema later in both eyes raised suspicion of an infective pathology and hence *in vivo* confocal microscopy (IVCM) (heidelberg retinal tomography 3-rostock cornea module HRT3-RCM adopting standard techniques) was planned. IVCM in both eyes revealed bright, hyperreflective dots seen from the epithelium to the midstroma, highly

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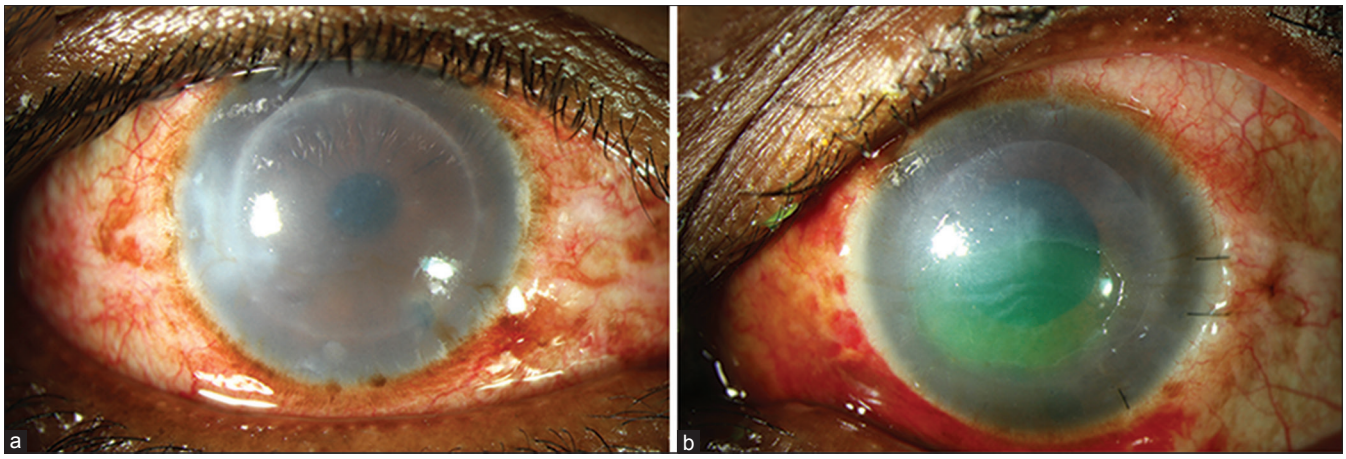

Figure 1: (a and b) Slit lamp photograph depicting right and left cornea post-Descemet's stripping automated endothelial keratoplasty (a) right eye few coarse superficial punctate lesions with graft edema and overlying host stromal edema (b) left eye-6*4 epithelial defect along with diffuse corneal edema of graft and entire host cornea

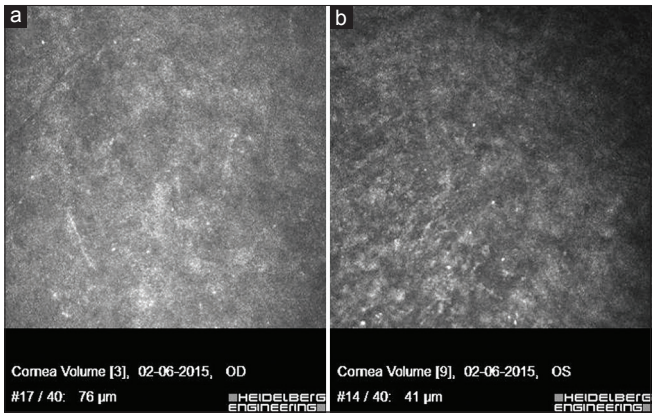

Figure 2: (a and b) Confocal microscopic images of corneal stroma of right and left eye at level of 76 µm and 41 µm, respectively hyperreflective dots interspersed around the keratocytes suggestive of microsporidial spores

suggestive of microsporidial spores [Fig. 2a and b].The topical and systemic steroids were discontinued, and the patient was prescribed topical fluconazole 0.3% 6 times a day and oral acetazolamide 250 mg twice a day. Ocular examination showed improvement, and he was discharged from hospital. He was advised to continue topical fluconazole while oral acetazolamide was stopped. The patient was called for a weekly review and on the third scheduled visit showed improvement in visual acuity with steadily resolving graft and overlying stromal edema. The UCVA had improved to 20/40 in the right eye and 20/60 in the left. A repeat confocal microscopy at 3 weeks revealed the complete disappearance of the hyperreflective dots in both eyes. The epithelial defect in the left eye had healed completely. Topical fluconazole was continued at a lower frequency, thrice a day for another week.

Discussion

Microsporidial keratoconjunctivitis has been reported as early as 2003 in immunocompetent individuals with an exponential increase in reported cases in the past 5 years or so. The first report of its occurrence in a corneal graft was in 2005.[1] Recently, microsporidial stromal keratitis masquerading as graft rejection has been reported by Pradhan *et al*.[2] but to the best of our knowledge, microsporidial keratoconjunctivitis presenting as bilateral graft edema has not been described so far in the literature. The initial presentation of conjunctival injection along with defective vision and graft edema led to a misdiagnosis of graft rejection with less attention being paid to the overt superficial punctate keratitis. The postoperative use of topical steroids in the left eye could have led to a local immunosuppressive effect which triggered the onset of keratoconjunctivitis and masked the severity of the signs and symptoms causing a diagnostic dilemma. The involvement of the other eye and suboptimal response to steroids was a wake-up call, raising suspicion of an infectious rather than an immune pathology, which was firmly established by confocal microscopy. The IVCM findings were consistent with microsporidial spores as reported in the literature.[3] As a tertiary eye care hospital, we see 2–3 cases of microsporidial keratoconjunctivitis per week; we have observed a good therapeutic response to topical fluconazole 0.3%, and therefore, it is our preferred choice of medication once the diagnosis is established by smears. There are however reports of other topical medications being used with varying degrees of success. Since there is no optimal or established treatment regimen for postgraft microsporidial keratoconjunctivitis, we started the patient on topical fluconazole 0.3% in both eyes. Bilateral involvement and the presenting clinical picture ruled out donor-related infection. Scraping for smears and culture was not initially attempted with the apprehension of a negative result due to the scanty material.

Conclusion

In reporting this unique case of bilateral graft edema after microsporidial keratoconjunctivitis, we have attempted to highlight the need for a high index of suspicion to rule out infectious causes when dealing with postoperative graft edema and to share our experience in managing the case. IVCM is a good tool for diagnosis of acanthoemeba and fungi, but for bacteria and microsporidia, there have been reports of false positive results. IVCM can prove invaluable when dealing with deeper corneal pathology.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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