Case Report

Superior Forniceal Advancement Conjunctival Pedicle (SFACP) in the management of corneal perforations and impending perforations due to rheumatoid arthritis related autoimmune corneal melts

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Abstract
Peripheral ulcerative keratitis (PUK) is a disorder consisting of a crescent-shaped destructive inflammation of the perilimbal corneal stroma. PUK may occur in variety of systemic diseases including collagen vascular disease. We describe the outcome of superior forniceal advancement conjunctival pedicle (SFACP) in two patients with corneal perforation and impending perforation due to rheumatoid arthritis (RA) related autoimmune corneal melt that has not previously been reported. Both patients had good recovery with restoration of vision when SFACP was performed as an adjunct to systemic immunosuppressive agents. SFACP is a valuable surgical option which may have an important role in reducing progression of corneal melt and underlying disease when used along with adequate systemic immunosuppressive agents.

Introduction
Peripheral ulcerative keratitis (PUK) may occur in variety of systemic diseases including collagen vascular disease. It remains a major diagnostic and therapeutic challenge and may result in corneal perforation that leads to sight threatening sequelae.1 Restoration of the integrity of the globe and ocular rigidity is necessary to preserve the visual potential. Surgical options to manage non traumatic corneal perforations are emergency tectonic graft, tissue gluing, conjunctival flap, and patching with exogenous materials.2-4 We describe the role of superior forniceal advancement conjunctival pedicle (SFACP) in the management of rheumatoid arthritis (RA) related corneal perforation or impending perforation that to our knowledge, has not previously been reported.

Case Report
An interventional case series is presented, which included the patients with corneal perforation or impending perforation secondary to RA related corneal melt that had SFCAP performed. SFACP is a technique for corneal perforations and impending perforations.5 A prominent blood vessel was identified in superior conjunctiva and conjunctival pedicle was formed with an intact blood supply and underlying Tenon’s capsule about 14 - 15 mm away from limbus. The advancing edge of the pedicle was placed on the area of perforation or impending perforation and sutured with 10/0 nylon. Postoperative medication consisted of G chloramphenicol and dexamethasone 0.1% four times daily for 1 month. The corneal sutures were removed after 2 to 3 weeks.

Case 1:
A 66-year-old Caucasian female presented with right painful red eye for 1 week. The diagnosis of right impending corneal perforation due to RA related PUK was made and VA was decreased to 6/60. She was a known patient of RA for 30 years and was using non steroidal anti inflammatory drugs (NSAID). Right SFACP was performed and immunosuppression regime was oral prednisolone 30 mg OD and cyclophosphamide 100
mg BD. After 1 week PUK was healing with no impending corneal perforation and oral prednisolone was tapered off gradually to 10mg. The conjunctival pedicle retracted itself after 4 weeks with complete resolution of ulcer. After 2 months cyclophosphamide was reduced to 50mg BD and oral prednisolone to the maintenance dose of 2.5mg. Six months later her right VA was 6/9 and there was no other episode of PUK or corneal melt (Figure-1). Cyclophosphamide was stopped and oral prednisolone 1mg daily was continued.

**Case 2:**

A 50-year-old Caucasian male presented with pain, redness and photophobia in the right eye for 3 weeks. There was RA related right corneal melt leading to a small corneal perforation at 3 o’clock position which was plugged with iris and VA was reduced to 6/60. Past medical history revealed RA and the patient was on oral sulphasalazine 75 mg daily. An emergency SFACP was performed next day and his immunosuppression treatment consisted of oral cyclophosphamide 100mg BD and oral prednisolone 30mg OD. The corneal perforation sealed with no leak on day 1. The conjunctival pedicle retracted completely after 3 weeks with healing of corneal perforation and associated melt. There was no further recurrence of the PUK or corneal melt. The oral prednisolone was reduced by 5mg every 2 weeks to the maintainance dose of 5mg. Two months later cyclophosphamide was stopped and oral prednisolone was tapered off slowly within 6 months. After 6 months VA was 6/9 and there were no signs of corneal melt (Figure-2).

**Discussion**

PUK is a devastating complication of RA that can lead to rapid corneal melt and perforation. The peripheral cornea has discrete morphologic and immunologic characteristics that predispose it to inflammatory reactions. Involvement of limbal vasculature can result in inflammatory cell recruitment and peripheral corneal necrosis and ulceration secondary to the liberation of the collagenolytic and proteolytic enzymes from these cells.6 The primary therapy for severe PUK associated with RA is aimed at aggressively controlling inflammation with systemic steroids combined with immunosuppressive agents like cyclophosphamide, cyclosporine A or azathioprine.7-9 An application of corneal glue and bandage contact lens has its role in corneal perforation or impending perforation. Although, it provides good tectonic support but does not contribute to the healing of the corneal ulcer and perforation. Tectonic corneal graft is an effective procedure in corneal perforation which requires donor tissue whereas SFACP is an alternative treatment option which does not require donor tissue and has shown encouraging results. SFACP is formed from peripheral non inflamed conjunctiva that promotes healing of non healing corneal ulcers due to its high vascularity. It not only helps to seal the perforation or prevents impending perforation but also rapidly eliminates the necrotic tissue and therefore is the stimulus for production of vasogenic substances.6 The corneal graft failure rate is relatively high in the RA patients although systemic immunosuppression increase the chance of anatomical success.10 The adjunct use of SFACP along with systemic immunosuppressive agents may have an important role in reducing progression of corneal melt and underlying disease. Both patients were managed successfully with SFACP and adequate immunosuppression; and achieved good visual rehabilitation.

**Conclusion**
Although it is a small number of cases but it describes SFCAP as an effective surgical procedure which has an adjunct part along with immunosuppression in corneal perforation or impending perforation secondary to auto immune related corneal melt.

References