Case Report

Surgical Management of Glaucoma in Eyes with Ocular Cicatricial Pemphigoid

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ABSTRACT

Introduction: Glaucoma in ocular cicatricial pemphigoid (OCP) is an important blinding complication. Glaucoma in eyes with OCP is most often managed medically as surgical management is difficult in eyes with cicatrised conjunctiva and often end up with complications or scarring and failure. In some cases where the topical antiglaucoma medications are not tolerated or worsen the OCP and in eyes with medically uncontrolled glaucoma, surgical management would be needed. The glaucoma drainage devices (GDD) are more suitable in these eyes compared to trabeculectomy in these eyes.

Case: Here two cases of a 35-year old and 62-year old males with biopsy proven OCP and steroid-induced medically uncontrolled glaucoma that were successfully managed with Ahmed glaucoma valve are reported.

Observation: Performing an implant in an eye with poor ocular surface is a challenge. Appropriate modifications need to be performed to overcome these challenges. In this report, the surgical challenges and post-operative management have been highlighted.

Conclusion: Despite intraoperative and post-operative challenges of performing glaucoma surgery in eyes with OCP, preoperative planning, meticulous surgery, and post-operative follow-up would go a long way in successfully managing refractory glaucoma in these patients.

Key words: Ahmed glaucoma valve; ocular cicatricial pemphigoid; poor ocular surface; scarred conjunctiva; steroid-induced glaucoma.

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INTRODUCTION

Ocular Cicatricial Pemphigoid (OCP) is a challenging autoimmune disease characterised by progressive conjunctival inflammation leading to cicatricial changes, subepithelial fibrosis, symblepharon formation, and subsequent complications. Glaucoma, a common comorbidity affecting 25-32% of OCP patients (Tauber et al., 1988), poses unique management difficulties due to various factors, including drug-induced toxicity, steroid-related complications, and surgical challenges. Corneal haze limits use of laser trabeculoplasty (Tauber et al., 1988; Tsai et al., 2006; Anwar et al., 2013) and limited success is noted with trabeculectomy surgery (Kaštelan et al., 2013; Tauber et al., 1989). Better success is reported with Ahmed glaucoma valve (AGV) in eyes with multiple ocular cicatrizing diseases including OCP however with higher rate of serious post-operative complications (Almousa et al., 2014). None of the reports discuss details of surgical management and steps taken to minimise or prevent complications in eyes with OCP, which are highlighted in this report.

CASE REPORT 1

A 35-year-old male with biopsy proven OCP was referred for uncontrolled intraocular pressure (IOP) in his right eye (RE) despite earlier treatment with lubricating drops, topical steroids, and amniotic membrane grafts over the past eight years.

Best-corrected visual acuity (BCVA) in RE was 20/20, N06 and for left eye (LE) was 20/30, N18. The IOP in RE was 40 mmHg and in the LE was 12 mmHg on four topical and oral glaucoma medications. Both eyes showed eyelid

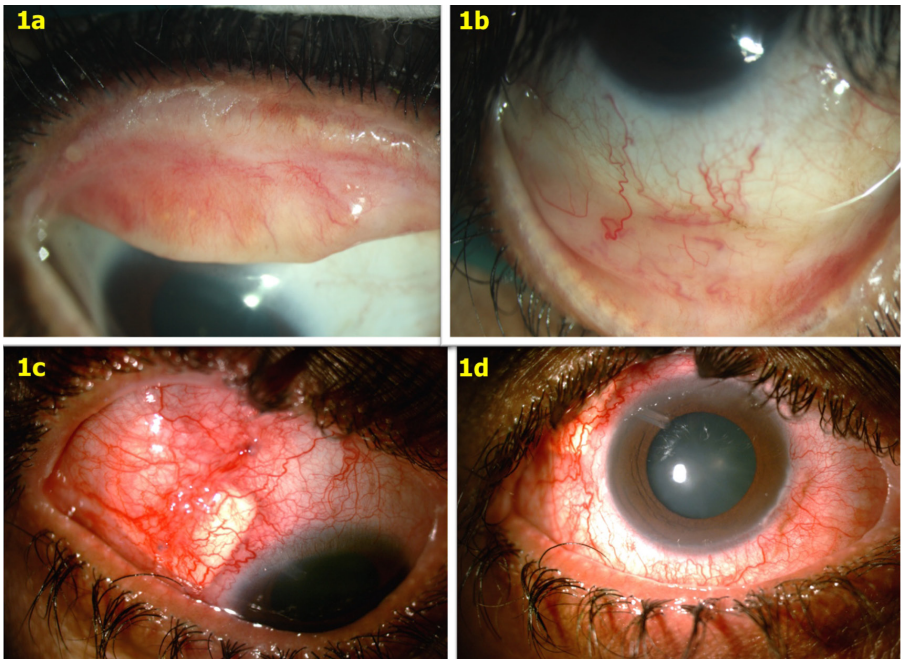

Figure 1: Case 1 preoperative photograph showing upper palpebral and tarsal conjunctiva showing subconjunctival fibrosis and epithelial sloughing; (1a), symblepharon, and forniceal shortening (1b). Post-operative day 1 appearance of bleb showing intact suture line and superior-temporal bleb (1c), well-placed tube in the anterior chamber (1d)

margin thickening, subconjunctival fibrosis and lid margin ulceration and keratinisation (Figure 1a), stenotic puncta, trichiasis, conjunctival fibrosis, forniceal shortening (Figure 1b), and superficial punctate keratopathy. The angles were open, RE had advanced glaucomatous disc damage and LE had posterior subcapsular cataract and early disc damage. The OCP was medically managed with tab cyclophosphamide 50 mg twice daily, tab prednisolone 60 mg once a day, topical cyclosporine, mild steroid and lubricants.

Due to uncontrolled IOP in RE, AGV implantation was performed. Limbus-based conjunctival flap was made supero-temporally and FP7 AGV implant was placed in the conjunctival pocket and was secured 8 mm posterior to the limbus with two 9-0 polypropylene sutures. The tube was inserted into the anterior chamber (AC) through a 4-5 mm scleral tunnel and was secured with 10-0 nylon, the entire length of tube was covered with a scleral patch graft. Conjunctiva was closed with 8-0 polyglactin continuous mattress suture.

Day-1 post-operatively, the RE IOP was 06 mmHg, conjunctiva was well apposed (Figure 1c), with well positioned tube (Figure 1d). Tablet prednisolone 60 mg once daily, tab doxycycline 100 mg twice daily for three weeks, topical preservative-free methylprednisolone eye drops (the intravenous injection form of 500 mg of methylprednisolone was reconstituted with sterile nonpreserved normal saline to achieve a final concentration of 1% and dispensed in a lubricating drop bottle, the patients were instructed to keep them refrigerated), moxifloxacin eye drops, tacrolimus eye ointment and lubricating eye drops were prescribed. In second week, topical aqueous suppressants, long-acting preservative-free formulation of timolol 0.5% once daily was started, tab prednisolone, and methylprednisolone eye drops were tapered.

At six-months post-operatively, the visual acuity (VA) was 20/20, N06, IOP was 15 mmHg on topical preservative-free formulation of timolol 0.5% with a good bleb. He was continued on a maintenance dose of oral Prednisolone 10 mg and low dose topical steroid.

CASE REPORT 2

A 62-year-old gentleman with biopsy proven OCP had steroid induced glaucoma (betamethasone eye drop use for 15 years). His BCVA was 20/20, N06 in RE and 20/25, N06 in LE. IOP was 17 mmHg in RE and 38 mmHg in LE on 4 topical and oral glaucoma medications. Both eyes showed lid margin thickening, distichiasis, blocked puncta, symblepharon in the lower nasal fornix (Figure 2a) with congested conjunctiva, and superficial punctate keratopathy. Both eyes were pseudophakic, open angles and LE had advanced glaucomatous damage.

OCP was treated with tab Prednisolone 60 mg once a day, Cyclosporine 0.05% eye drops, Loteprednol etabonate 0.5%, Tacrolimus 0.03% eye ointment and topical lubricants.

In view of medically uncontrolled IOP, GDD was planned. Because of severe conjunctival fibrosis and forniceal shortening, an FP8 AGV implant was chosen and the rest of the procedure was similar to case 1. On the post-operative day one, the LE VA was 20/125 and IOP was 09 mmHg, the conjunctival wound

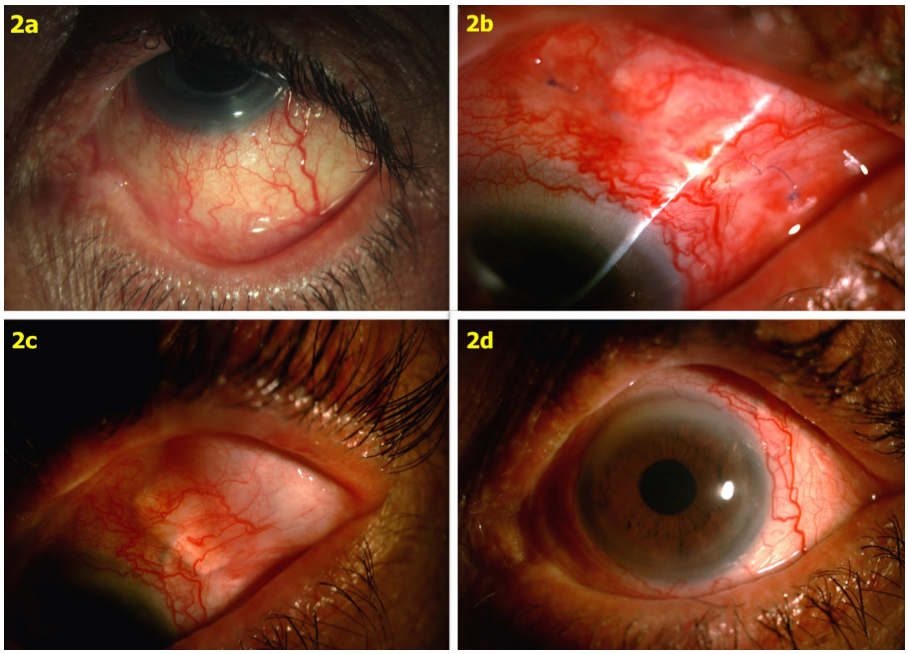

Figure 2: Case 2 showing symblepharon and forniceal shortening (2a), post-operative day 1 picture showing intact suture line and minimally ischemic conjunctiva close to the suture line (2b), 6-month post-operative photograph with good bleb (2c) and quiet anterior segment (2d)

was well apposed (Figure 2b), and the tube was in position. Tablet cyclophosphamide 50 mg, tab doxycycline 100 mg, Dexafree eye drop (dexamethasone phosphate 1% preservative-free), moxifloxacin 0.5% and lubricant eye drops were prescribed.

On day five, a small conjunctival wound gape was noted, IOP was 9 mmHg and there was no leak. The anterior chamber was shallow, there was peripheral choroidal detachment. A large diameter (18 mm) bandage contact lens was placed covering the gape (to avoid mechanical trauma to the defective area) and tablet prednisone 60 mg was added to the previous prescription.

By second week, the conjunctival gape was well apposed, the bleb was elevated and tense and IOP was 25 mmHg and choroidal detachment resolved. Topical preservative-free timolol and brimonidine combination was added, dexamethasone eye drop was tapered and stopped, and tab prednisolone was tapered until 5 mg per day maintenance dose. At six months, the BCVA in the LE was 20/30p, N6, IOP was 13 mmHg with a good bleb (Figure 2c, 2d).

DISCUSSION

OCP is an autoimmune disease characterised by chronic inflammation and progressive scarring of the conjunctiva associated with several ocular complications (Tsai et al., 2006; Tauber et al., 1989; Szabó et al., 2016). Glaucoma is an important sight-threatening complication in these patients. Anticipating excess scarring and wound-related complications, most of us defer any incisional surgery more so glaucoma surgery in these eyes. However, in medically

uncontrolled glaucoma, we should choose an appropriate procedure that would be safe and effective. The authors of this report, could not find any published literature to guide us in surgically managing glaucoma in eyes with OCP. Almousa et al., (2014) showed the success of AGV for IOP control in eyes with ocular surface disease of various causes, however, complications were noted in a quarter of these eyes. Authors reported repeated tube exposure in one patient which required multiple repairs and ultimately needed explantation in that eye with OCP. Other complications reported were hypotony, flat anterior chamber, choroidal effusion, and hyphema. They did not specify the success of AGV and complications, particularly in OCP.

In both of the currently reported cases, there was a significant reduction of the IOP post-AGV implant. In one case, choroidal effusion, shallow AC, and a small conjunctival wound gape was noted. Both the choroidal effusion and the conjunctival wound gape were managed conservatively which resolved in one week. Intraoperative surgical modifications and post-operative use of preservative-free medications and oral immunosuppression possibly contributed to the successful outcome. Wound modulating agent tab doxycycline may help by promoting wound healing and decrease the fibrotic response post-operatively (Dubey et., 2013; Gupta et al., 2017; Kiranmaye et al., 2014). Using an appropriately sized drainage implant is important to prevent serious intraoperative and post-operative complications like implant exposure and extrusion in the presence of fibrosis and shortened conjunctiva. Despite a long scleral tunnel, using a partial thickness scleral patch graft and meticulous conjunctival closure may help to prevent tube exposure. Gonioscopy assisted transluminal trabeculotomy (GATT) can be an option in cases with open angles. However, steroid response post GATT can be a significant problem in these cases due to the need for usage of long-term steroids in these cases.

CONCLUSION

To conclude, despite intraoperative and post-operative challenges of performing glaucoma surgery in eyes with OCP, preoperative planning, meticulous surgery, and post-operative follow-up would go a long way in successfully managing refractory glaucoma in these patients.

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