Management of phacoemulsification surgery in immune-mediated progressive cicatrising conjunctivitis (PCC)

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Introduction

- Progressive Cicatrising Conjunctivitis (PCC) describes a diverse group of immune disorders which lead to persistent inflammation and chronic conjunctival scarring. [1] Thirty underlying conditions are reported with mucous membrane pemphigoid (MMP) (prevalence=1:45,000) being the most common aetiology in Europe. [2] Causes can be local or systemic and include Stevens-Johnson Syndrome/Toxic Epidermal Necrolysis (SJS/TEN),Sjogren's disease, Atopic keratoconjunctivitis (AKC), Acne Rosacea and Drug Induced Cicatrising Conjunctivitis (DICC). [2]
- PCC causes severe morbidity due to conjunctival shrinkage and compromised tear film. Visual loss occurs secondary to ocular surface failure and secondary keratopathy. [1] Phacoemulsification is challenging due to difficult access, poor anterior chamber visualisation and risk of post-operative surface epithelial breakdown, reactivation and/or progression of disease. [3] We report outcomes of a management protocol that minimises post-operative complications in PCC.

Methods

- Medical records of 130 consecutive cataract surgeries over 14 years (2005-2019) performed on 88 patients (aged (50-87) years, 49 (56%) Female) with established ocular surface disease presenting to a regional tertiary referral centre. (**Figure 1**)
- Patients were inflammation-free for a minimum of 3 months before surgery with 49(56 %) requiring systemic immunosuppression**.** (**Figure 2**) Six-month outcomes are reported.
- All patients underwent the same ocular surface optimisation protocol.
- **Ocular surface pre-surgery pre-conditioning protocol:** preconditioning topical non-preserved dexamethasone 6x/day for 2 weeks pre-operatively, intravenous methylprednisolone 500mg-1g on the day of surgery, intra-operative hydroxypropyl-methylcellulose corneal protection, amniotic membrane to the corneal surface, and post-operative topical non-preserved dexamethasone, chloramphenicol and lubricants.

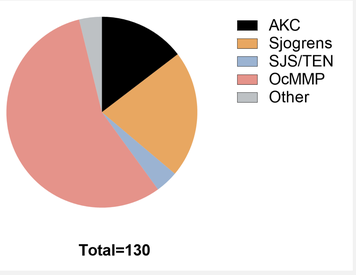


Figure 1: PCC causes . OcMMP (n=73); Sjogren's (n=28); AKC(n=19); SJS/TEN(n=5); Other causes: Rosacea (n=3); DICC (n=1); neoplasia (n=1).

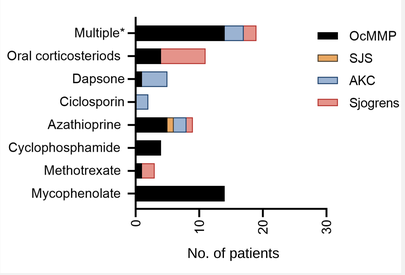
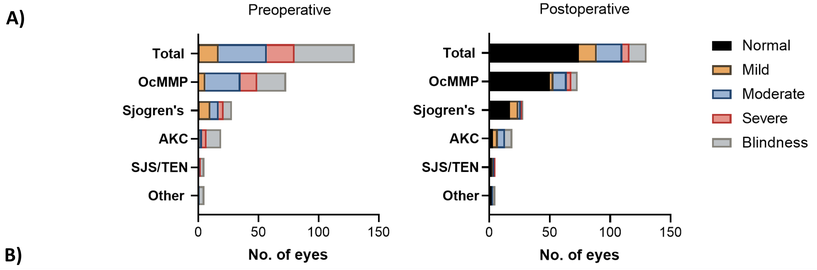


Figure 2: Systemic immunosuppression by disease type and agent. *Patient was on more than one immunosuppressive agent before surgery.

Results

- 54 eyes (42%) had >50% forniceal-foreshortening prior to surgery with 37 eyes (28 %) requiring intraoperative lid-sutures.
- **Postoperative vision:** significant improvement at 6-months for the three largest groups (OcMMP: P=0.000; Sjogren's: P=0.0001 , AKC: P= 0.001). There was significant improvement in vision for the complete dataset at 6-month follow-up (P= 0.001) with an average decrease of 0.86 logMar. **(Figure 3)** 7 patients had a decrease in vision: Two patients due to probable disease progression, two due to severe dry eye disease, one due to advanced glaucomatous field loss, one due to retinal detachment and one with cystoid macular edema.

A)



B)

Snellen visual acuity	LogMAR visual acuity	WHO visual impairment category
> 6/12	*< 0.30*	*None*
6/12-6/18	*0.30 – 0.50*	*Mild impairment*
6/18-6/60	*0.50 – 1.00*	*Moderate impairment*
6/60-3/60	*1.00 – 2.00*	*Severe impairment*
< 3/60	*> 2.00*	*Blindness*

Figure 3: Pre- and postoperative visual acuity. A) Best-corrected visual acuity B) WHO visual impairment scale.(6)

- **Postoperative complications:** whilst there was no worsening of surface staining, 4 (3%) eyes developed persistent epithelial defects that resolved with increased lubrication. The most common complication was filamentary keratitis. **(Table 1)** Two patients demonstrated disease progression and one had corneal decompensation. There were no complications recorded for DICC, neoplasia or rosacea patients. No patients developed corneal ulceration, perforation or endophthalmitis. There is no significant relation between preoperative systemic immunosuppression and postoperative complications. (P= 0.6722)

	Postoperative uveitis	Severe dry eye /Filamentary keratitis	Microbial keratitis	Exacerbation/ progression	Persistent epithelial defects	Cystoid macular edema
OcMMP	*1*	*32*	*0*	*1*	*3*	*1*
Sjogren's	*0*	*2*	*0*	*1*	*0*	*0*
AKC	*1*	*1*	*0*	*0*	*0*	*2*
SJS/TEN	*0*	*1*	*1*	*0*	*1*	*0*

Table 1: Postoperative complications.

Discussion

- Previous studies reporting cataract surgery outcomes in cicatrising conjunctivitis found significant improvements in visual acuity ranging from 0.2 to 1.2 logMAR. **(Table 2**) The improvement however was temporary and low vision was associated with epithelial defects and corneal complications such as corneal decompensation, keratinisation and vascularisation.
- In our study, there was reduced rates of disease progression compared to published reports of cataract surgery in patients with ocular surfaces disease.
- Presence of perioperative inflammation and dryness with poorer cataract outcomes and disease progression have been reported. In our cohort, filamentary keratitis associated with dry eye was the most common post-surgical feature despite copious intraoperative HPMC and amnion overlay with therapeutic contact lens protection for three weeks post surgery. Vigilant postoperative monitoring and timely implementation of rescue therapy is crucial to optimise patient clinical outcomes.

	VA improvement (LogMar, Snellen lines)	Disease progression	Keratopathies
Audit	*0.9, 9*	*1.5 %*	*FK (25%);*
Maza et al. (1988)[7]	*0.3, 3.5*	*0 %*	*Corneal ulcers (11.5%)*
Geerling et al. (2000)[5]	*0.2, 2*	*40 %*	*PED (6.7%), perforation(13%)*
Donthineni et al.(2020)[9]	*1, 10*	*-*	*corneal decompensation (8.2%); PED (6.56%)*
Puranick et al .(2013)[10]	*0.2, 2*	*22%*	*SPK (11%); PED (11%)*
Narang et al. (2016)[11]	*1.2, 12*	*-*	*PED (5%); FK (2.5%)*

Table 2: Overview of literature on visual outcomes and complications.VA: Visual acuity; FK: filamentary keratitis; PED: Persistent epithelial defects; SPK: superficial punctate keratitis.

Conclusion

Successful phacoemulsification outcomes in PCC requires careful perioperative management strategies. Good visual outcomes are reported although careful monitoring is required to manage post-surgical exacerbations of dry eye disease. Visual improvement may however be temporary due to the progressive nature of the underlying disease.

References

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