A Rare Spontaneous Corneal Keloid Presentation: clinicopathological features, genetics and surgical outcome

Petya Popova, Matteo Posarelli, Matteo Airaldi, Alfredo Borgia , Yamini Krishna, Nardine Menassa

INTRODUCTION

Corneal keloids are rare, benign, grey-white elevated lesions characterized by epithelial hyperplasia and the accumulation of disorganized collagen fibrils. They may develop following ocular trauma, surgery, or inflammation, but can also occur idiopathically or in association with congenital conditions such as Lowe syndrome or Rubinstein-Taybi syndrome. Surgical intervention is typically reserved for visually significant cases; however, treatment approaches and outcomes vary widely due to the rarity of the condition.

CASE PRESENTATION

A 41-year-old Caucasian male presented with gradual vision deterioration in his left eye. His past medical and ocular history included sleep apnoea, obesity and floppy eyelid syndrome. Initial visual acuity was 6/24 in the right eye and 6/60 in the left. Bilateral corneal stromal oedema, endothelial vesicular changes and guttata suggested corneal endothelial dystrophy. Left DSAEK was planned, but surgery was delayed due to the COVID-19 pandemic. Two years later, a new white corneal lesion with neovascularization was noted . Excision and biopsy revealed a left corneal keloid.



INVESTIGATIONS & TREATMENT

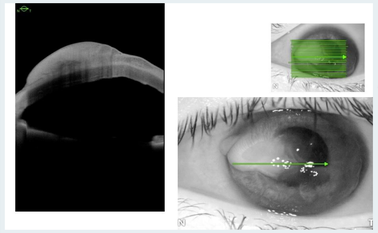


Figure 1: *Anterior segment SPECTRALIS Heidelberg OCT at the first presentation of the left corneal lesion . Visual acuity is hand movements.*

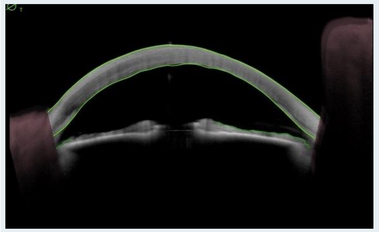


Figure 2: *Anterior segment SPECTRALIS Heidelberg OCT of the left cornea six weeks post excision and biopsy of the corneal lesion. Visual acuity is hand movements.*

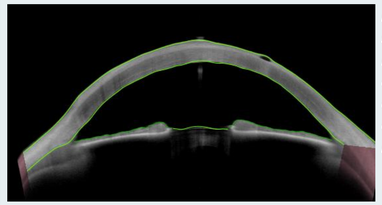


Figure 3: *Anterior segment SPECTRALIS Heidelberg OCT of the left cornea ten weeks post excision and biopsy of the corneal lesion with early recurrence.*

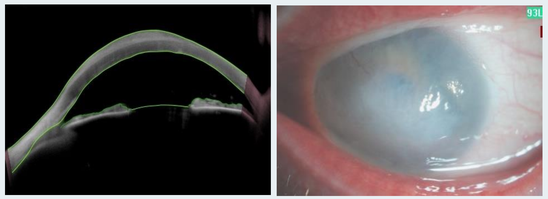


Figure 4: *Anterior segment SPECTRALIS Heidelberg OCT and colour photograph showing recurrence of the left corneal keloid extending diffusely across most of the cornea and its original borders ten months post the excision and biopsy. Visual Acuity is hand movements.*

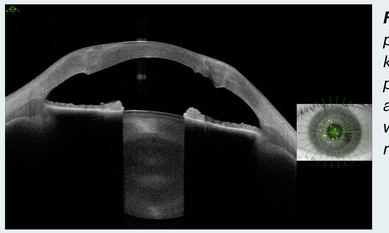


Figure 5 : *Seven months post left superficial keratectomy, penetrating keratoplasty and amniotic membrane with no signs of recurrence. VA 0.48*

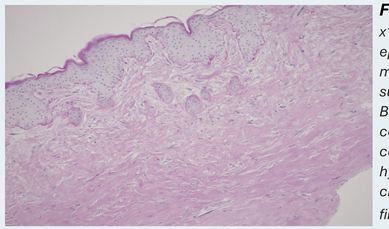
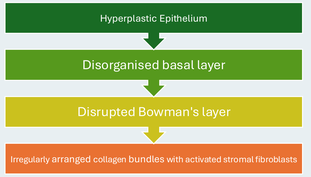


Figure 6 : *Periodic-Acid Schiff (PAS) x100 showing irregular hyperplastic epithelium, thickened basement membrane, swollen basal cells and subepithelial oedema and scarring. Bowman's layer has been completely lost. The stroma contains disorganised, thickened, hyalinised collagen with myxoid-type change and is cellular with fibroblasts.*

CONCLUSION

- Genetic testing showed an autosomal dominant ZEB1-related disease.
- ZEB1 is a key transcription factor involved in epithelial-to-mesenchymal transition (EMT) and cell differentiation, associated with several corneal dystrophies, including keratoconus, Fuchs endothelial dystrophy, and posterior polymorphous corneal dystrophy [1].
- Corneal keloids are reported more frequently in males [2].
- Superficial keratectomy or excision/biopsy often results in rapid recurrence and lesion enlargement.
- Amniotic membrane may improve surgical outcomes due to its anti-inflammatory and anti-fibrotic properties [3].



CONTACT INFORMATION
Petya Popova
Petya.Popova@rlbuht.nhs.uk
Liverpool University Hospital NHS Foundation Trust

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