Case Report

Crumbs, Criss Cross across Cornea-Avellino Dystrophy (A Rare Mixed Stromal Entity)

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Received: 08-06-2021 / Revised: 19-08-2021 / Accepted: 30-09-2021

Abstract

Male patient aged 50 years presented with redness, pain and watering in right eye since 8 days. Examination revealed that right eye corneal transparency was lost in central and temporal half of cornea due to multiple small and white dot without any clear space in between and with presence of network of lines called lattice in stroma. Transparency was present in peripheral cornea. Surface was irregular with presence of foreign particle (hair). Foreign body removal was done. On follow-up, due to poor compliance, we observed fungal ulcer for which therapeutic keratoplasty was done which lead to graft rejection after few months.

Keywords: Crumbs, Criss Cross, Cornea-Avellino Dystrophy.

Introduction

The corneal dystrophies are a group of non-inflammatory, inherited, bilateral disorders of the cornea characterized by pathognomonic patterns of corneal deposition and morphological changes[1]. Classically, the stromal corneal dystrophies have been classified based on their phenotypic appearance at the slit lamp and divided into the three main types of granular, lattice, and macular.Avellino Dystrophy also known as Granular-Lattice Dystrophy, since it displays findings of both diseases and has autosomal dominant inheritance[2]. It is characterized by tiny whitish dots in the anterior stromal layer in childhood which progress into larger stellate, ring, or snowflake-like opacities over time[2].

Case Report

A 50-year-old male presented with redness, pain and watering in right eye since 8 days. On examination, right eye corneal transparency was lost in central and temporal half of cornea due to multiple small, white dot without any clear space in between and with presence of network of lines called lattice in stroma, not involving endothelium was observed s/o Avellino dystrophy. Transparency was present in peripheral cornea. Surface was irregular with presence of foreign particle (hair) in inferotemporal part of cornea along with infiltration in subepithelial and stromal area which was suggestive of superficial keratitis (Fig 1A & B).

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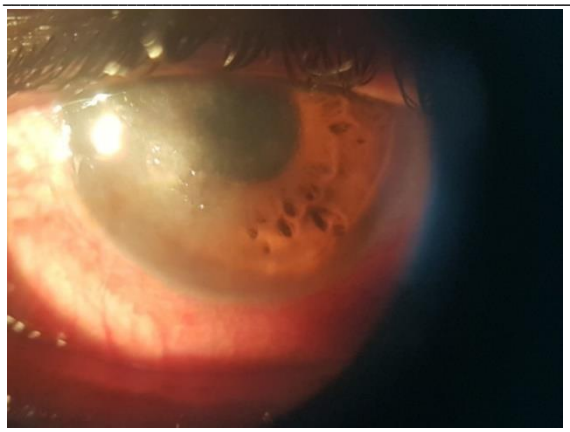
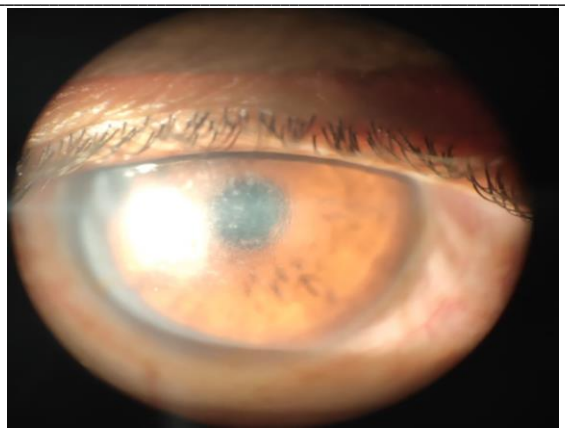

Fig. 1a and Fig. 1b: Superficial keratitis with foreign particle in inferotemporal part of cornea

Rest of the anterior segment was within normal limits. Left eye examination revealed status post keratoplasty for similar complain 2 years back with no sign of recurrence till date (Fig 1C). History of similar complaints was observed in sister and father. Foreign body removal was done and topical antibiotics, cycloplegic and lubricating eye drop was advised. But on subsequent follow-up due to poor compliance, the patient developed fungal ulcer for which therapeutic keratoplasty was done, which eventually lead to graft rejection few months later (Fig. 1D). Patient further planned for keratoplasty.

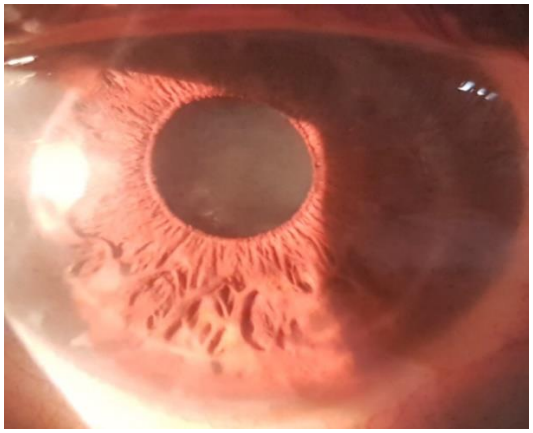
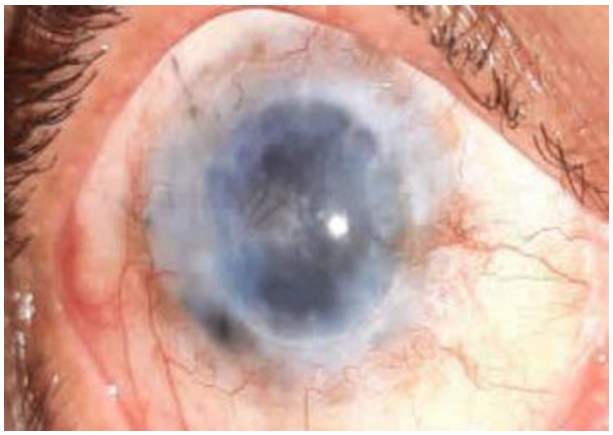

Fig. 1c: left eye post keratoplasty

Fig.1d:Right eye graft rejection following therapeutic keratoplasty

Conclusion

Vision loss may occur earlier than in Type 1 Granular, with many patients noting vision loss in adolescence. In avellino vision loss occur later than type 1 for which Keratoplasty was done. Recurrent erosions and photophobia are also more common in Type 2 Granular than Type 1. Due to higher incidence of recurrence post keratoplasty and progressive vision diminution, patients of avellino dystrophy should be kept in regular follow up.

References

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Conflict of Interest: Nil
Source of support:Nil