Saudi Journal of Ophthalmology (2016) 30, 49–52

Case Report

Corneal xanthogranuloma in association with multiple endocrine neoplasia 1: A clinicopathologic case report and review of the literature

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Abstract

Juvenile xanthogranuloma (JXG) is a benign inflammatory condition of uncertain pathogenesis. It is characterized by skin and ocular involvement – typically in the iris – in children. It has been reported in older age groups and has been also observed to involve other ocular structures such as the cornea and conjunctiva.
In this case report, we are presenting an extensive right eye corneal lesion in a 43-year old male which showed the typical histopathological feature of JXG and in association with multiple endocrine neoplasia (type 1). Similar cases in the English-language literature have been also reviewed.

Keywords: Cornea, Xanthogranuloma, Multiple endocrine neoplasia 1

http://dx.doi.org/10.1016/j.sjopt.2015.11.006

Introduction

Juvenile xanthogranuloma (JXG) is a benign inflammatory condition of uncertain pathogenesis characterized by skin and ocular involvement – typically in the iris – in children.[1] However, it can involve the cornea and the conjunctiva presenting as epibulbar lesions and has been also reported in older age groups.[1,2] We are reporting this lesion in the cornea of an adult patient with typical histopathological appearance.

Case report

A 43-year old male presented with a right painless corneal lesion, brownish in color, and slowly enlarging over a period of 7 years. He described an almost identical similar corneal lesion in his left eye for which penetrating keratoplasty (PKP) was performed in Germany 8 years prior to his current presentation. Systematically he was known to have multiple endocrine neoplasia type 1 with pan hypopituitarism and hypoadrenalism, on replacement therapy. He was a known diabetic on oral hypoglycemic and was recently diagnosed with ischemic heart disease.

The patient's vision in his right eye was light perception owing to his corneal lesion. The corneal mass was yellowish-pink in color, showed feeding blood vessels and obscured any further view of the anterior chamber and the posterior pole in his right eye (Fig. 1a). His left eye showed a centrally clear corneal graft with peripheral corneal haze and vascularization involving the edge of the graft (Fig. 1b).

Received 29 June 2015; received in revised form 8 November 2015; accepted 10 November 2015; available online 25 November 2015.

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Peer review under responsibility of Saudi Ophthalmological Society, King Saud University

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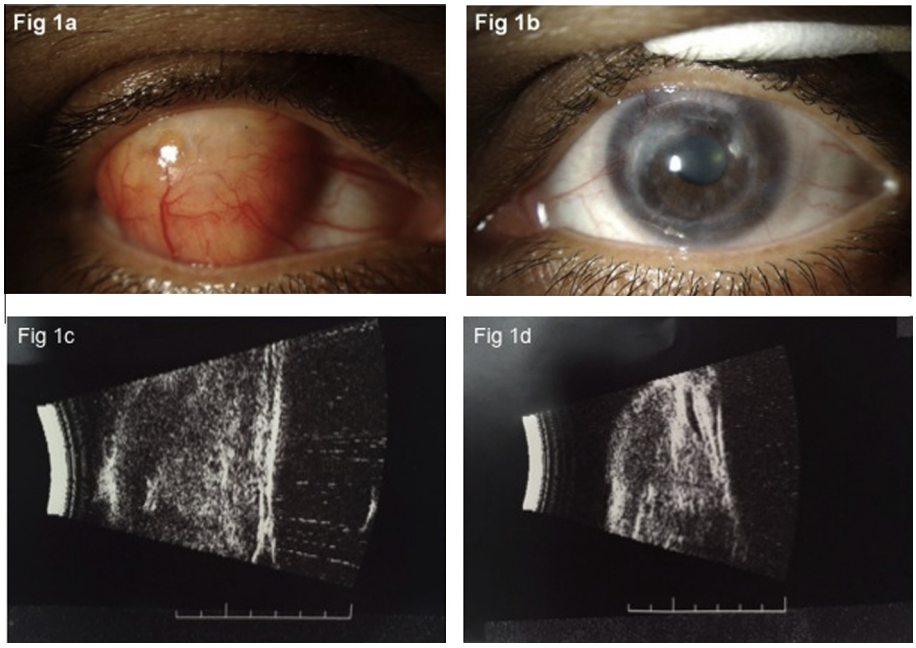

Figure 1. (a) The clinical appearance of the right eye corneal vascularized mass. (b) The clinical appearance of the centrally clear corneal graft in the patient's left eye. (c) Ultrasound bio microscopy showing the large right corneal lesion approaching the iris centrally. (d) Ultrasound bio microscopy showing the same right eye lesion with shallow anterior chamber near the angle.

The left eye uncorrected visual acuity was 20/40 (improving with pinhole to 20/30). The rest of the left eye examination was unremarkable. Ultrasound Bio microscopy (UBM) study of the right eye showed a large corneal lesion approaching the iris centrally with extremely shallow space between the lesion and the iris near the anterior chamber angle for 360 degrees (Fig. 1c and d).

The patient underwent PKP procedure with a 10 mm size graft for his right eye because of the extensive corneal involvement reaching the limbus. The corneal tissue was sent for histopathological examination. The corneal tissue showed acanthotic epithelium and absent Bowman's layer. Sub epithelial calcification and hemosiderin-laden macrophages were noted. The corneal stroma was replaced by a nodular-like lesion consisting of mixed epithelioid cells and lymphocytes. Foamy histiocytes and multinucleated Touton giant cells were also observed. Descemet's membrane showed a focal area of delamination and few endothelial cells with flattened nuclei. A focal area of adherent iris tissue and pigment deposition was also observed. The histiocytes were all positively stained with CD68 while showing negative staining with S-100 and CD 1a (Fig. 2a–d). The diagnosis of corneal xanthogranuloma was made.

Unfortunately our patient developed cataract and secondary glaucoma 1 year after his PKP with evidence of peripheral anterior synechiae and iris–corneal touch (Fig. 3). His corneal graft eventually failed 3 years following his PKP and then he was lost to follow-up.

Discussion

Limbal xanthogranuloma was first reported in 1958 by Cogan who described this lesion as the so-called: ''Nevoxan thoendothelioma''. His case was a 5-year old girl who presented with multiple limbal lesions consisting of lymphocytes and foamy histiocytes with no Touton giant cells.[3]

On the other hand, Collum was the first to describe limbal xanthogranuloma in an adult in 1984.[4] Several case reports have been published describing similar lesions involving the cornea, episclera and conjunctiva in different age groups.[5–9] Mocan and his coauthors did an extensive review and summary of all reported limbal cases in 2008.[1] They concluded the rare occurrence of juvenile xanthogranuloma as an isolated lesion at the corneoscleral limbus in general. The clinical presentation in this location varied from a yellowish to red raised lesion or fleshy mass often observed with associated vascularization such as in our case. The lesions shared the typical histopathological finding of foamy histiocytes and Touton giant cells as described in our lesion. Their review however included all age groups. Zelger and his coauthors studied the histological and IHC staining features of JXG and the adult-type xanthogranuloma confirming similar results.[10]

Lee and his coauthors[11] described a case with an identical age to our patient: a 43-year old man developed 2 painless conjunctival masses over 2 months described as round yellowish masses approaching the limbus, however these did not involve the cornea. The lesions were excised and were found to be infiltrative and adherent to the underlying sclera. Their immunohistochemical (IHC) staining revealed diffuse CD3 positive reactive T lymphocytes and CD68 positive histiocytes, which were negative with S-100 stain indicating that these were non-Langerhans histiocytes. Our IHC staining showed similar results.

Taking into consideration that our patient is a 43-year old, we have tried to focus on the occurrence of such a lesion in adults by reviewing the available English-language literature since the first reported case of adult limbal xanthogranuloma by Collum in 1984.[4] More cases of limbal and conjunctival lesions have been identified in older age group other than infants and young children with an estimated prevalence of 13–18% of JXG cases being encountered in the second decade of life.[12,13]