Adult limbal xanthogranuloma

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SUMMARY An 18-year-old white man presented with a non-painful yellow raised swelling on the inferior limbus of his right eye. Systemic and ocular examination revealed no other abnormalities. The lesion was dealt with by simple excision, but when it recurred fairly soon it was removed in toto and replaced by a lamellar graft, without recurrence. Histological examination revealed a typical xanthogranuloma. The question is, why should a healthy male with no other manifestations develop a lesion like this on his limbus?

Juvenile xanthogranuloma is said to be typically a benign skin disorder found in babies and young children.¹ The skin nodules often disappear spontaneously and are rarely associated with visceral manifestations. Studies of this condition have given rise to the conclusion that juvenile xanthogranuloma is not a member of the histiocytosis X group.² A search of the literature would suggest that xanthogranuloma at the limbus has been infrequently studied histologically.

This case is reported in view of the rarity of previous occurrences at this site in juveniles and because this is the first xanthogranuloma at the limbus to be described in an adult.

Case report

A 17-year-old male was first seen in April 1979 having noticed a painless swelling on the limbus of his right eye at 6 o'clock for about 3 months. This had gradually increased in size, and when he was examined by us he was found to have a solid pink tumour there measuring 5×3×6 mm (Fig. 1). It extended for approximately 3 mm on to the cornea and a similar distance on to the sclera with deep extension into both tissues. Apart from this both eyes were normal, with full visual acuity. There were no other systemic abnormalities on clinical examination, and all clinical biochemistry tests were negative.

It was decided that it might be possible to remove the tumour by simple excision, and this was done in April 1979. The postoperative result was not entirely

satisfactory in that a white localised area remained. This was observed, but over the next couple of months a recurrence appeared to be beginning at the site of the original lesion with slight elevation in the area of the excision.

Six months later in December 1979, it was felt that the lesion was progressing to the extent that it would have to be re-excised. On this occasion a superficial keratectomy was performed to get at the base of the tumour, and a lamellar graft was inserted. There was no postoperative complications, and in the intervening 4 years the cornea has remained quite clear with no evidence at all of any recurrence of the disease (Fig. 2). Clinical examination of the eye at this time again shows no abnormality, and the vision remains normal in both eyes.

Fig. 1 Initial appearance of limbal mass.

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Fig. 2 Postoperative appearance of limbus.

HISTOPATHOLOGY

A small piece of tissue was received which showed microscopically modified squamous epithelium with intra- and subepithelial mixed inflammatory cells. There were numerous histiocytes and Touton giant cells (Fig. 3) as well as fibroblasts, sometimes in whorled pattern. A homogeneous structure under the epithelium indicated the presence of Bowman's membrane at some levels (Fig. 4). The histology was typical of that of xanthogranuloma. The pathological areas extended to the edges of the sections.

Specimen 2. The recurrence consisted of small fragments of interstitial tissue covered by stratified squamous epithelium containing mixed inflammatory cells, including histiocytes, and an occasional Touton giant cell similar to the previous histology.

Fig. 3 Touton giant cell lying beneath corneal epithelium. (Haematoxylin-eosin, $\times 79$).

Discussion

Five histologically confirmed and reported cases²⁻⁴ classified as limbal xanthogranuloma, that is, stated to contain giant cells, are available to us to date (Table 1). This is in addition to other yellow lesions at the limbus with lipid-bearing histiocytes without giant

Fig. 4 First excision. Bowman's membrane fragmented, histiocytic infiltration, and scattered giant cells. (Haematoxylin-eosin, $\times 22$).

Table 1 *Reported cases of limbal xanthogranuloma*

Author	Patient	Ocular findings	Skin lesions	Treatment	Follow up
Cogan <i>et al.</i> ³	5-year-old female	Bilateral limbal yellowish masses	Present	Excised partially	Recurrence right eye after 8 months
Zimmerman ²	Case 1. 10-month-old female	Left pink epibulbar mass, sclero-limbal	None	Excision at 1 year old	—
	Case 2. Newborn child	Bilateral yellowish lesions extending to limbus. Cornea free	Several skin lesions	Epibulbar lesions excised at 1 year old	—
	Case 3. 4-month-old male	Left temporal limbus Hyphaema. Cornea free	Not stated	Excision of limbal lesion. Radiation for intraocular lesion	—
Nordentoft, Andersen ⁴	18-month-old female	Right reddish yellow epibulbar and limbal mass. Cornea free	None	Excision and irradiation	6 months later no recurrence

cells⁵⁻⁹ (Table 2). The case now presented differs from others in the first group in that the patient was not a juvenile. The reported cases described have all been treated either by simple excision or excision combined with radiotherapy. Our case is the only one which appears to have had a keratectomy and lamellar graft. We would suggest that a graft is necessary to eradicate the mass completely and to give a good cosmetic result. The surgery is straightforward, but complete removal of the lesion does leave a defect which must be filled with a graft in order to avoid the

occurrence of pseudo-ptyerygium or an unsightly scar on the cornea.

It is not clear why such a lesion should appear in a completely healthy young man. It has been suggested⁷ that local factors such as lymphostasis could bring about the formation of xanthomas and nodular sub-epidermal fibrosis by irritation, but only one patient gave a history of redness and photophobia over a period of 3 months.⁴

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Table 2 *Cases reported as xanthoma/fibrous histiocytoma*

Author	Patient	Ocular findings	Other lesions	Treatment	Follow up	Diagnosis
Liebman <i>et al.</i> ⁸	Case 1. 3-year-old male	Yellow/orange episcleral and corneal lesions bilateral. (Xanthoma syndrome)	Multiple dermal and visceral masses	Radiotherapy. Recurrence: lamellar keratoplasties	Right and left cornea opacifying at 12½ years of age	Xanthoma
	Case 2. 2½-year-old female	Yellow conjunctival and limbal nodules bilateral	Myelocytic leukaemia multiple dermal xanthoma	β-radiation Strontium 90	Eye lesions resolved. Patient died of leukaemia at 5 years of age	Xanthoma
Grayson, Pieroni ⁶	11-year-old male	Left yellow limbal lesion. Cornea involved	None	Excision and conjunctival flap	No recurrence 7 years after operation	Xanthoma
Jacobiec ⁷	3-year-old female	Right corneo scleral tan/pink limbal mass. Cornea involved.	None	Excision and lamellar keratectomy	No recurrence 2 years later	Fibrous histiocytoma
Faludi <i>et al.</i> ⁵	21-year-old female	Left limbal yellowish/tan lesion	None	Excisional biopsy. Lamellar keratectomy and sclerotomy with cryotherapy		Fibrous histiocytoma
Litricin ⁹	65-year-old female	Greyish/yellow corneal thickening	Rheumatoid arthritis	4 operations. 2 keratectomies on nodule, lamellar excision, keratoscleroplasty	Enucleation after 5 years due to perilimbal thickening	Fibrous histiocytoma

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