When the tumor gradually became larger, we excised it for a precise diagnosis, which showed that it was JXG. The present case is rare because (1) only six cases of JXG involving the corneoscleral limbus have been previously reported as occurring in patients at less than 5 years of age; and (2) the extensive time course of the lesion was photographed.

Of the 18 reported patients with JXG involving the corneoscleral limbus, three patients underwent partial excision. In all three cases, the tumor recurred. In these recurring cases, all tumors were located on the lower side of the limbus. Recurrence may be related to the choice of surgery and the location of the tumor. It has been the main choice of therapy to totally excise the tumor, since complete excision decreases the chance of limbal JXG recurrence. In our case, observation 12 months after the surgery has not shown any evidence of tumor recurrence, but we will have to continue to follow this patient.

Solitary JXG affecting the corneoscleral limbus is a rare form of limbal lesion. If such a lesion becomes enlarged and extends to the stromal cornea rapidly, total excision is required, and occasionally, lamellar keratoplasty. JXG should be considered in the differential diagnosis of any lesions occurring in the limbus.

**Key Words:** juvenile xanthogranuloma, limbus

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Received: October 26, 2006 / Accepted: March 19, 2007
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DOI 10.1007/s10384-007-0450-5

**References**


**A Case of Adult Limbal Xanthogranuloma**

Juvenile xanthogranuloma (JXG) is an uncommon benign skin disorder usually seen in infants and children, occurring...
at less than 1 year of age in 70% of the cases. Only 10% of affected patients develop ocular involvement. Although any part of the eye may be affected, the iris is the most frequently affected region. Solitary JXG occurring at the corneoscleral limbus is an uncommon type. We studied a case of JXG of the limbus in a 48-year-old woman.

Case Report

A 48-year-old woman noticed a painless yellow-orange mass on her right eye of 2 weeks’ duration. Her visual acuity was 20/20 in both eyes, and a slit-lamp examination showed a yellow-orange mass located at the superior limbus of the right eye (Fig. 1). The mass measured approximately 4 × 5 × 2 mm. No other ocular abnormalities were observed. The patient’s medical history and past ocular history were non-contributory. Results of all laboratory tests were within normal limits, and no skin lesions were noted.

Local steroid therapy and betamethasone eye drops, for 1 month, were ineffective; therefore, the mass was excised, with the patient under local anesthesia. The surgery included lamellar dissection of the mass, which had penetrated the surface of the corneal stroma. Microscopically, the mass was covered with stratified squamous epithelium, and inflammatory cells had permeated under the cuticle (Fig. 2A). Fibroblasts, lymphocytes, and histiocytes were detected. Numerous Touton giant cells were also noted (Fig. 2B). Touton giant cells showed lipid deposition in their outer circumference, between the outer cell membrane and the center ring of cellular nuclei. There were no cellular atypia or foreign body. Postoperative visual acuity was 20/20 in the patient’s right eye. There was no evidence of recurrence during a follow-up period of 15 months.

Comments

Juvenile xanthogranuloma is a rare benign skin disease primarily affecting infants and young children. An ocular manifestation, mainly in the iris, has been found in approximately 10% of affected patients. It is the most common cause of spontaneous hyphema in children. Secondary glaucoma is also a potential complication. It has a tendency to affect the iris, but other ocular involvement in the eyelid, conjunctiva, sclera, corneoscleral limbus, ciliary body, orbit, or optic nerve has also been reported. The etiology of JXG is poorly understood; however, several indications point to the presence of inflammation. It has been considered a form of local irritation, which is the underlying stimulus for the accumulation of the activated macrophages that characterize the lesion. However, we could not find any sign of irritation in our patient’s past history. Fungi or bacteria,