Short communication

Juvenile xanthogranuloma of the corneoscleral limbus in adults

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ABSTRACT

Case report: A 25-year-old woman noticed a painless yellow-orange mass on her right eye. 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. No other ocular abnormalities were observed.

Discussion: Surgical excision was carried out and the lesion was sent for histological examination. This showed a granulomatous lesion, rich in Touton-type giant cells, features that are strongly suggestive of juvenile xanthogranuloma (JXG). Ocular involvement occurs in 10% of cases of JXG.

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Xantogranuloma juvenil en limbo corneoescleral en el adulto

RESUMEN

Caso clínico: Mujer de 25 años de edad que refería una tumoralización no dolorosa amarillenta en el ojo derecho. Su agudeza visual era de la unidad. En la exploración en lámpara de hendidura se observa una masa amarilla-anaranjada localizada en el limbo superior del ojo derecho. No se observaron otras anormalidades.

Discusión: Se realizó una escisión quirúrgica de la lesión y se remitió a anatomía patológica para su examen. La lesión granulomatosa mostraba células de Touton. Este hecho sugiere de forma importante el diagnóstico de xantogranuloma juvenil (JXG). La afectación ocular ocurre en el 10% de los casos de JXG.

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Introduction

Juvenile xanthogranuloma (JXG) is an infrequent, benign and non-hereditary histiocytosis of non-Langerhans cells. In most cases the lesions appear in the first year of life and even at birth, and in smaller percentages in adolescents or adults.\(^1\)

Skin lesions are unique in 60–80% of cases and are located in head and neck, although they can also appear in the top part of the chest and the proximal area of limbs. Expression is in the form of papules or well-defined and firm tuberous lesions, initially red-orange and subsequently brownish. Extracutaneous involvement is rare and predominantly in the eyes in which the most frequent location is the iris, with possible invasion of the angle and causing hemorrhage (hyphema) and glaucoma. It can also involve the retina, orbit, choroids and optic nerve. Sclerocorneal limbus involvement is rare.\(^2\)

Ocular lesion occurs in 10%\(^3\) of JXG cases and normally expresses in infants under one year of age and is generally unilateral. Internal organ involvement is even less frequent (liver, lungs, SNC). Association with neurofibromatosis and chronic juvenile myeloid leukemia has been described.

Case report

Female, 25, who consulted due to orange raised lesion with several months evolution located at 11 o’clock in the limbus and invading 1.5 mm of the cornea. Patient exhibited best corrected visual acuity (BCVA) of one in both eyes. Ocular fundus did not exhibit relevant findings. In the absence of diagnostic certainty, surgical excision was indicated (Fig. 1). The intervention began in the conjunctiva and upon reaching the limbus it separated easily from the cornea without requiring lamellar keratotomy. Histopathological study revealed that the main composition of the lesion was a proliferation of histiocytes and numerous multinucleated Touton-type giant cells (Fig. 2) with microvacuolated cytoplasm expressing monocytic/macrophagic markers (CD68 positive) and negative for Langerhans cell markers (protein S100 and CD1a).

The presence of mature lymphocytic infiltrate and very few eosinophiles was observed.

Discussion

The incidence of JXG ocular alterations is approximately 10%\(^2\) and, although it has been described in adults, it expresses with greater frequency in children with multiple skin lesions. The most frequent ocular involvement is at the anterior uvea and very rarely it can present as a mass in the sclerocorneal limbus, with very few cases being described in the literature.\(^2,4–6\) Approximately less than 10 documented cases.\(^5\)

Differential diagnostic must be considered for any tumor lesion of the sclerocorneal limbus such as atypical skin lipoma, lipoma, pyogenic granuloma, neurofibroma, leiomyoma, neurilemmoma, amelanocytic melanoma.\(^5,7\)

Several therapeutical options have been described: simple excision with or without lamellar keratectomy, excision combined with radiotherapy, intralesional administration of corticoids, excision with intra- and post-surgery corticoids or only corticoids treatment.\(^2,3,9\)

In the case reported herein, a simple excision of the lesion was performed without requiring lamellar keratectomy even though in slit lamp examination it appeared firmly adhered to the cornea, upon reaching the limbus it detached with ease. For the majority of authors, surgical intervention is necessary\(^10\) although there are some discrepancies about the healing effect of the excision\(^1\) as some cases have exhibited relapses,\(^8\) in which case supplementary treatment with corticoids would be necessary. In this case no corticoid treatment was prescribed and in successive examinations in the course of 5 years no relapse or other kinds of lesions have been observed (Fig. 3).

JXG diagnostic is established after histopathological study, characterized by histiocyte infiltrates with varied morphology, microvacuolated or xanthomatous cytoplasm, spindle cells with storiform pattern, etc. The most characteristic cells are

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**Fig. 1** - Raised vascularized yellowish-orange in sclerocorneal limbus.

**Fig. 2** - JXG microphotography with presence of numerous Touton-type histiocytic multinucleated cells (asterisk) (hematoxylin-eosine, 20×). Bar: 100 μ.
Fig. 3 – Appearance 2 months after surgery.

Touton-type multinucleated giant cells with centralized nuclei and vacuolated cytoplasm although these are not constant in extracutaneous lesions. After establishing the diagnostic, the patient was referred to Dermatology for screening other possible lesions, verifying the absence of JXG cutaneous lesions.

The case reported herein differs from other groups of patients in what concerns age because this disease, even considering the low frequency of sclerocorneal limbus presentation, is even less frequent in adulthood. An additional difference is the evolution as simple excision was sufficient, without requiring adjuvant therapy.

Conflicts of interest

The authors declare no conflicts of interest.

REFERENCES