

## Juvenile Xanthogranuloma of the Eyelid in Two Children

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### Abstract

Juvenile Xanthogranuloma is a benign dermatosis in children and teenagers, characterized by one or several small yellow or brown nodules which appear in the first years of life, having spontaneous involution during a year. We report clinical histories, ophthalmic examination and histopathology results of 2 cases with solitary Juvenile Xanthogranuloma on upper right eyelid. A 4 year-old boy was evaluated for a lesion on the right upper eyelid. The patient gave history of this swelling for last 8 months, for which he was being treated by a local ophthalmologist thinking it to be chalazion, for which incision and curettage has been done in the last month. The lesion was surgically removed in its entirety. Biopsy of his eyelid mass confirmed the diagnosis of juvenile xanthogranuloma. An 8 year-old girl presented with upper right eyelid margin nodule. Excisional biopsy of the nodule performed and confirmed it was a juvenile xanthogranuloma. All two cases yielded a good prognosis and at the 5-years follow-up, here was no recurrence of the lesion. Complete resection of the mass is essential to confirming the diagnosis and avoid recurrence.

**Key words:** Eyelid juvenile xanthogranuloma; upper eyelid; eye. Biopsy; chalazion.

### INTRODUCTION

Juvenile xanthogranuloma (JXG) is an uncommon, benign dermatologic condition, which occasionally involves the eye, orbit and ocular adnexa affecting primarily infants and young children [1-6]; more than 80% of the cases occurring during the first years of life [4]. Approximately 10% of these patient's exhibit ocular manifestations, whose presentations vary [3]. The ocular lesions are typically unilateral, although in few cases bilateral lesions have been described with variable presentation [4]. We describe two cases of eyelid juvenile xanthogranuloma. One of the cases was a recurrent tumor. In this case biopsy and complete resection of the mass is essential to confirming the diagnosis and avoid recurrence.

### CASE REPORT

#### Case 1

A 4-year-old infant boy was evaluated for an upper eyelid lesion of 6 month duration. For long, he was being treated by a local ophthalmologist thinking it

to be chalazion, for which incision and curettage has been done in the last month. After the surgery, the lesion enlarged and the patient was presented to us. His visual acuity was 20/20 in both eyes. Ophthalmic examination revealed a 0.5 cm diameter elevated, nontender, subcutaneous yellowish nodule of right upper eyelid (Figure 1).

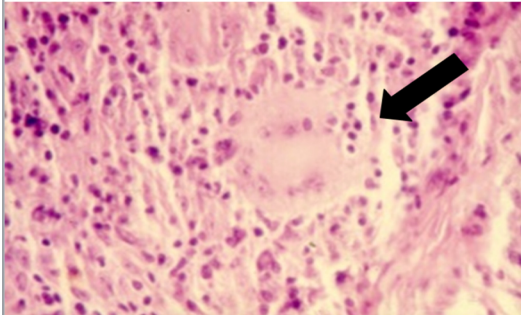


**Figure 1.** The yellowish nodule after partial resection

The remainder of the ocular and skin examination was normal. There were no similar cutaneous lesions elsewhere. The lesion was surgically removed. Histopathological examination of the lesion revealed

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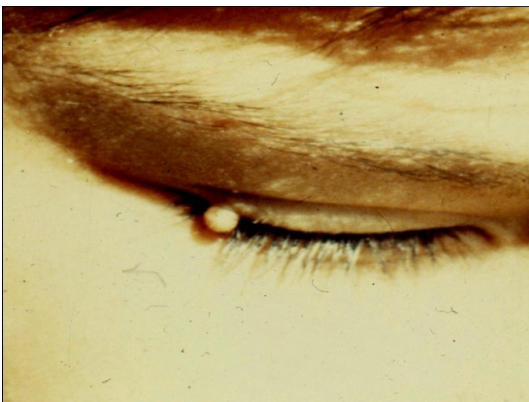
that the epidermis did not present alterations. In the dermis was observed an infiltrate of lymphocytes, plasma cells, numerous histiocytes and multinucleated Touton giant cells suggestive of JXG (Figure 2).



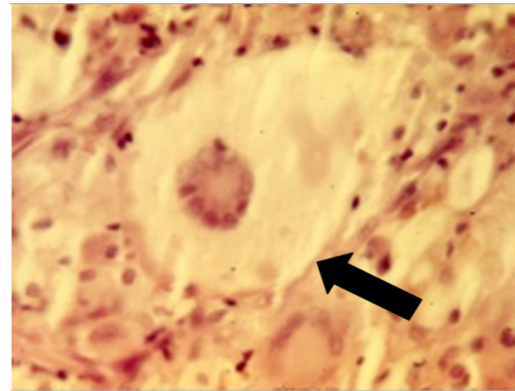
**Figure 2.** Histopathological examination showed an infiltrate of lymphocytes, plasma cells, numerous histiocytes and multinucleated Touton giant cells.. (Haematoxylin and eosin stain at x 400 magnification)

### Case 2

An 8-year-old girl presented to us with a 4-month history of a slowly enlarging, spherical right upper eyelid painless tumour that was 4 mm in diameter without umbilication (Figure 3). On presentation, the patient's eye looked relatively normal, and she had uncorrected visual acuity of 20/20 in both eyes. Ophthalmic examination revealed a 4 mm diameter pedunculated spherical right upper eyelid margin, painless yellowish nodule. The remainder of the ocular examination was normal. There were no skin lesions and the systemic pediatric workup noncontributory. An excisional biopsy was then performed. Histopathological examination showed the existence of an infiltrate of lymphocytes plasma cells, eosinophils, with numerous histiocytes and Touton giant cells (Figure 4)



**Figure 3.** The solitary nodule in the right eyelid margin



**Figure 4.** Histopathology showing the existence of an infiltrate of lymphocytes, plasma cells, eosinophils, with numerous histiocytes and Touton giant cells.. (Haematoxylin and eosin stain at x 400 magnification)

### DISCUSSION

JXG is a rare, benign cutaneous disorder of unknown etiology [6, 7] that occurs in infants and young children [1-6]. Presentation usually occurs in childhood with spontaneous regression by adolescence [1, 3, 8, 9]. JXG most commonly affects children less than one year of age and nearly 64% are younger than 7 months [1] as solitary or multiple red, yellowish or brown cutaneous nodule [6, 8] showing a predilection for the head and neck region [5, 8, 10, 11] but these papulonodular lesions may occur on the trunk and extremities also [10]. The appearance of multiple lesions is more frequent in children younger than 6 months, which are not our cases. Visceral involvement by JXG is rare, with a predilection for lung, spleen, testis, pericardium, gastrointestinal tract [6] brain liver [9] kidney, central nervous system, adrenal gland, pituitary gland, bones, bone marrow and heart. [10]. The most common ocular finding is diffuse or discrete iris nodules, which could be quite vascular and may bleed spontaneously, resulting in hyphema [3,4] and secondary glaucoma [6]. This condition can affect occasionally, the orbits [3, 4, 6] eyelids [4-6, 9, 11] conjunctiva, optic nerve, choroid [9] cornea [4, 8] retina [8] and episclera [3]. Eyelid involvement represented 25% of cases in a single series of ocular JXG [6]. Our first patient was previously mistakenly treated as chalazion by incision and curettage instead of excision biopsy, omitting the diagnosis. In this case due to incomplete resection, recurrence and rapid growth of the tumor, arousing concern for malignancy, surgical resection in toto had been used for both correct diagnostic and therapeutic reasons.

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The clinical differential diagnosis includes spitz nevi, mastocytomas and dermatofibromas [10] lymphangioma, dermoid cyst, pilomatricoma [5]. The confirmation of clinical diagnosis can be made by skin biopsy. Characteristic histologic findings in JXG are: dense dermal histiocytic infiltrate and Touton Giant cells which are multinucleated, with homogeneous eosinophilic cytoplasmic center and xanthomatization in the periphery [10], In conclusion childhood tumors tend to benign and self-limited like ours cases. Eyelids JXG can be diagnosed by a skin biopsy if skin lesions are present. JXG should be considered in the differential diagnosis of a solitary eyelid yellowish nodule in the pediatric population. Surgical treatment is required on those with eyelids involvement for fear of malignancy, diagnostic and therapeutic reasons. In our first case the nodule enlarged after the partial resection. For these reasons the nodule must be completely resected otherwise they recur. All two cases yielded a good prognosis and at the 5-years follow-up, here was no recurrence of the lesions.

### CONSENT

Written informed consent was obtained from the patient's parents for publication of this Case Report and any accompanying images.

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