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Case report

Juvenile xanthogranuloma: an unusual and rare presentation as an eyelid nodule

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ABSTRACT

Juvenile xanthogranuloma (JXG) is an uncommon disorder. It belongs to the broad group of non-Langerhans cell histiocytosis. It is characterized by one or more cutaneous nodules and less often by additional lesions in the deeper soft tissue or organs themselves. Skin lesions are well-demarcated, rubbery, tan-orange papulonodules ranging from 1 to 20 mm in size. They may be single or multiple and usually occur on the head and neck. Extracutaneous locations of JXG are the lung, heart, gastrointestinal system, central nervous system, adrenal gland, pituitary gland, bones, bone marrow, and kidney. It also has a predilection for the ocular structures, especially the uveal tract. Although JXG of the eyelid is uncommon and its occurrence anywhere in the eye is rare, it should be included in the differential diagnosis of eyelid mass lesions. We report a case of JXG in a 4-year-old child with an unusual presentation over the eyelid.

1. Introduction

Juvenile xanthogranuloma (JXG) belongs to the group of non-Langerhans cell histiocytosis.[1] It is characterized by one or more cutaneous nodules and less often by additional lesions in the deeper soft tissue or organs themselves.[2] Skin lesions are well-demarcated, rubbery, tan-orange papulonodules ranging from 1 to 20 mm in size. They may be single or multiple and usually occur on the head and neck. They may appear at any site including the groin, scrotum, toe nail, palm, sole, and lip.[2,3] It also has a predilection for the ocular structures, especially the uveal tract.[4,5] Although JXG of the eyelid is uncommon and its occurrence anywhere in the eye is rare, it should be included in the differential diagnosis of eyelid mass lesions.[6] We report a case of JXG in a 4-year-old child with an unusual presentation over the eyelid.

2. Case report

A 4-year-old girl presented to the ophthalmology department at our hospital with a 1.5-month history of slowly growing nodular growth over the right eyelid near the lateral canthus. She was otherwise healthy, and ocular examination was normal. There were no other skin lesions. Family history was not significant. Excision was performed, and the specimen was sent to the histopathology department with a clinical diagnosis of granuloma pyogenicum. Grossly, it was a skin-covered nodular piece of tissue measuring $0.5 \times 0.4 \times 0.3$ cm. Cut surface was grayish white. On microscopy, the section was lined by skin with epidermis showing focal attenuation and flattening of rete ridges. The dermis showed a poorly circumscribed lesion composed of histiocytic and fibrohistiocytic cells, focally showing storiform pattern. Cells had an indistinct cell borders with amphophilic to eosinophilic cytoplasm. Few cells showed vacuolated cytoplasm. Touton giant cells were also identified. In addition, inflammatory infiltrate, including lymphocytes, neutrophils, and eosinophils, was seen (Fig. 1a,b). On immunohistochemistry, histiocytes were positive for CD68 and negative for CD1a and S-100 (Fig. 2a,b,c).

On the basis of microscopic and immunohistochemical findings, the diagnosis of JXG was made.

3. Discussion

JXG is a benign cutaneous fibrohistiocytic lesion and a type of granulomatous process, at times accompanied by lipid deposits. It most often affects young children but occasionally arises as a solitary tumor in adolescents and adults.[7]

The main clinical feature is single or multiple papulonodular lesions, tan orange in color, and several millimeters in diameter.

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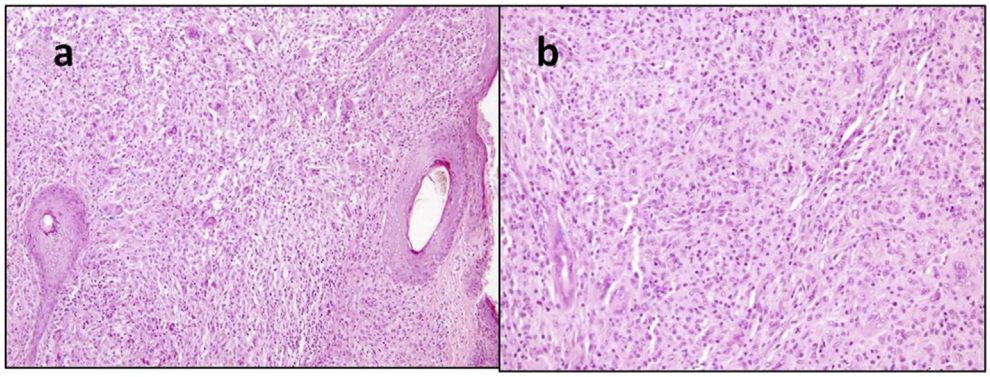

Fig. 1. a, b - H&E, 10x, 20x: Numerous touton giant cells with mononuclear histiocytes and scattered lymphocytes.

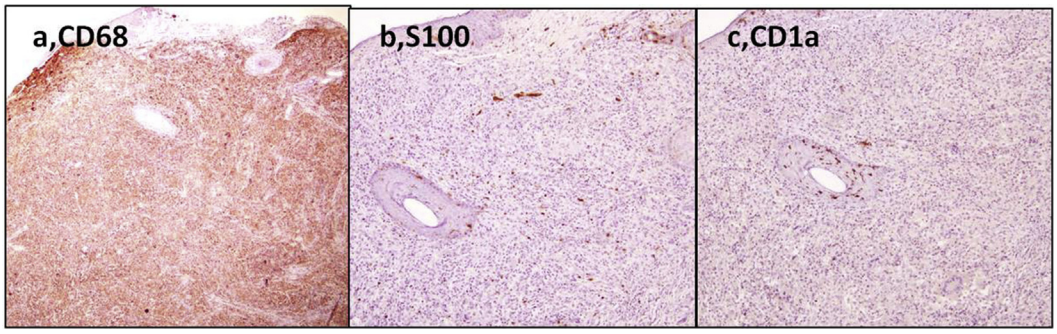

Fig. 2. a,b,c – CD68, S100, CD1a, 4x: Tumor cells show diffuse positivity with CD68 and negative staining with S100 and CD1a.

The predilection sites of occurrence are the skin of the head and neck, but these lesions may occur on the trunk and extremities also.[4,5]

The eye is the most frequent extracutaneous location of JXG, involving the uveal tract mostly; however, eyelids are a rather rare site.[4,5] Ocular involvement may occur without concomitant skin involvement as in our case. Of patients with cutaneous JXG, ocular involvement is reported in 0.5% of them,[8,9] although a higher incidence is reported in some studies.[9,10] Children with multiple skin lesions and those who are younger than 2 years are at a greater risk for ocular involvement.[2,10]

The male-to-female ratio of cutaneous JXG is about 1.4:1 in children, whereas in adults, no sex predilection exists.[11]

Helwig and Hackney[12] showed the fibrohistiocytic origin of tumors, and since then, the term “juvenile xanthogranuloma” has been commonly used.

The diagnosis is mainly based on characteristic clinical features. The clinical differential diagnosis includes Spitz nevi, mastocytomas, and dermatofibromas.[1] The confirmation of clinical diagnosis can be made by skin biopsy.

Microscopically, the typical JXG contains macrophages with a variety of cellular features. On a low-power microscope, a well-circumscribed nodule, often exophytic and with an epidermal collarette, is seen. Typically, many morphologic variants of macrophages can be seen.[13,14]

The lesions show characteristic progression as they mature. Early lesions may show large accumulations of vacuolated cells without significant lipid infiltration intermingled with only a few lymphocytes and eosinophils. When no foamy cells or giant cells are seen, the possibility of JXG is often overlooked. Usually some degree of lipidization is present, even in very early lesions, manifested by pale cells. In mature lesions, a granulomatous infiltrate is usually present containing foamy cells, foreign-body giant cells, and Touton giant cells, as well as macrophages, lymphocytes, and eosinophils. Occasionally, Touton giant cells are absent even in mature lesions. Older, regressing lesions show proliferation of fibroblasts and fibrosis replacing part of the infiltrate. Two types of spindle cells may be identified; the spindle-cell or fusiform macrophages are dispersed throughout the lesion. Langerhans cells may be found as bystanders, generally at the periphery of the lesion.[13]

JXG lesions usually label strongly with CD68, whereas S-100 and CD1a protein immunoreactivity is typically absent.[11] In most cases with JXG, S-100 protein is nonreactive, but scattered cells may show weak cytoplasmic reactivity, unlike the more diffuse and intense reaction of Langerhans cells.[14]

Differential diagnosis of JXG includes several entities. Histiocytosis X may be differentiated on the basis of demonstrating the proliferation of Langerhans histiocytes with oblong and notched nuclear morphology, epidermal and adnexal invasion, positive S-100 and CD1a immunostain within the tumor cells, and ultrastructural evidence of Birbeck granules.[7]

Xanthoma associated with hyperlipidemia is distinguished by the presence of wide and evenly distributed foamy histiocytes. Reticulohistiocytomas usually show random arrangement of multinucleated histiocytes with eosinophilic glassy cytoplasm and a reticulin-rich matrix. These distinctive cells are few or absent in xanthogranuloma. Dermatofibromas usually have a dense collagenous stroma with a uniform growth pattern and a hyperplastic epithelium, whereas in JXG, a storiform pattern is indistinct and the covering epidermis is thin.[11]

JXG is a self-limited disorder, and skin lesions usually resolve spontaneously. Treatment is required on those with extracutaneous involvement, who may have increased morbidity. Sometimes the

excision is performed for diagnosis and esthetic reasons as in our case.[11]

JXG of the eyelid is uncommon, and its occurrence anywhere in the eye is rare. It should be included in the differential diagnosis of eyelid mass lesions. We recommend that such lesions should be biopsied, including total excision when no functional compromise from surgery is anticipated.

Source(s) of support

None.

Conflict of interest

None.

References

1. Dehner LP. Juvenile xanthogranuloma in the first decades of life. A clinicopathologic study of 174 cases with cutaneous an extracutaneous manifestations. *Am J Surg Pathol*. 2003;5:579–593.
2. Cypel TKS, Zuker RM. Juvenile xanthogranuloma: case report and review of the literature. *Can J Plast Surg*. 2008;16:175–177.
3. Chu MW, Werner A, Moody-Antonio SA. Juvenile xanthogranuloma of the tympanic membrane: a case report. *Ear Nose Throat J*. 2012;91:364–368.
4. Shields CL, Shields JA, Buchanon HW. Solitary orbital involvement with juvenile xanthogranuloma. *Arch Ophthalmol*. 1990;108:1587–1589.
5. Kaur H, Cameron JD, Mohney BG. Severe astigmatic amblyopia secondary to subcutaneous juvenile xanthogranuloma of the eyelid. *JAAPOS*. 2006;10:277–278.
6. Hayashi N, Komatsu T, Komatsu T, Hiroi M, Ueno H. Juvenile xanthogranuloma presenting with unilateral prominent nodule of the eyelid: report of a case and clinicopathological findings. *Jpn J Ophthalmol*. 2004;48:435–439.
7. Daniel J, Mark A. Non melanocytic cutaneous tumors. In: Mills SE, Carter D, Reuter VE, Greenson JK, Stoler MH, Oberman HA, eds. *Sternberg's Diagnostic Surgical Pathology*. Philadelphia: Lippincott Williams & Wilkins; 2015:67–68.
8. Liang S, Liu YH, Fang K. Juvenile xanthogranuloma with ocular involvement. *Pediatr Dermatol*. 2009;26:232–234.
9. Karcioglu ZA, Mullaney PB. Diagnosis and management of iris juvenile xanthogranuloma. *J Pediatr Ophthalmol Strabismus*. 1997;34:44–51.
10. Pajaziti L, Hapçiu SR, Pajaziti A. Juvenile xanthogranuloma: a case report and review of the literature. *BMC Res Notes*. 2014;7:174.
11. Sonoda T, Hashimoto H, Enjoji M. Juvenile xanthogranuloma-Clinicopathologic analysis and immunohistochemical study of 57 patients. *Cancer*. 1985;56:2280–2286.
12. Helwig EB, Hackney VC. Juvenile xanthogranuloma (nevoxanthoendothelioma). *Am J Pathol*. 1954;30:625–626.
13. Kubota Y, Kiryu H, Nakayama J, Koga T. Histopathologic maturation of juvenile xanthogranuloma in a short period. *Pediatr Dermatol*. 2001;18:127–130.
14. Tomaszewski MM, Lipton GP. Unusual expression of S-100 protein in histiocytic neoplasms. *J Cutan Pathol*. 1998;25:129–135.