Juvenile Xanthogranuloma Typical and Atypical forms in the same patient. A case report and review of the literature

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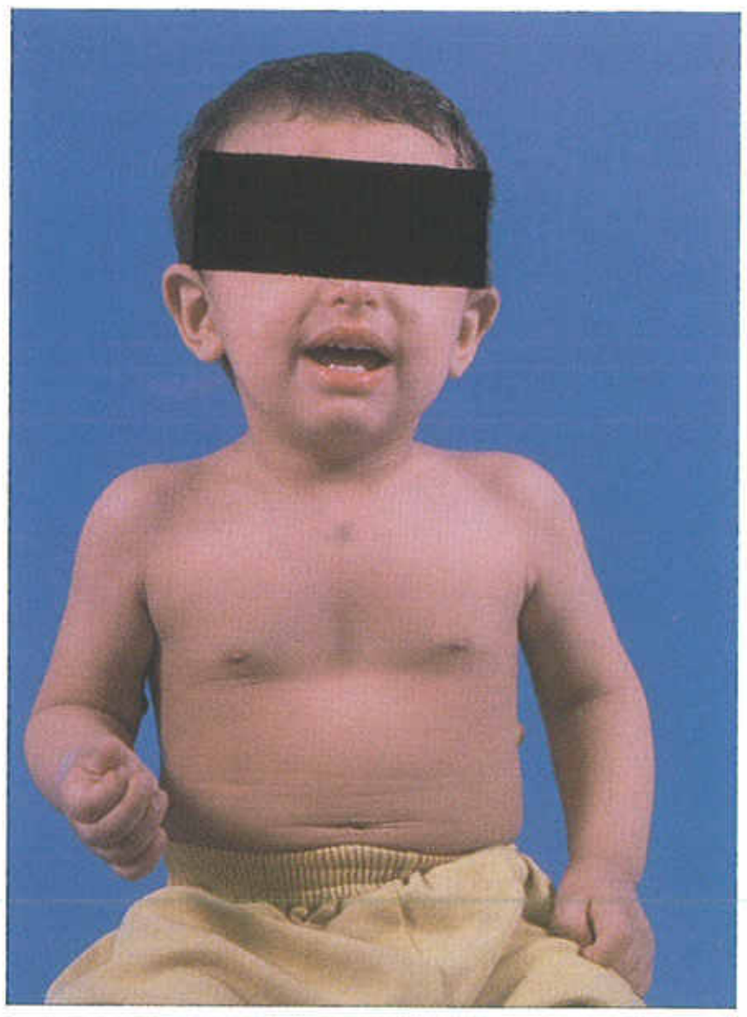
Introduction.

Juvenile Xanthogranuloma (JXG) is a benign self-healing disorder characterized by solitary or multiple yellowish-red nodules on the skin and in other organs. It is predominantly a disease of infancy and early childhood. The patient's general health is not impaired, there is normal lipid metabolism and the prognosis is excellent.

Histologically, JXG represents an accumulation of histiocytes. Diagnosis is readily made in typical cases but may be more difficult in atypical variants.[1]

We report a new case of Juvenile Xanthogranuloma with a combination of typical and atypical clinical variants.

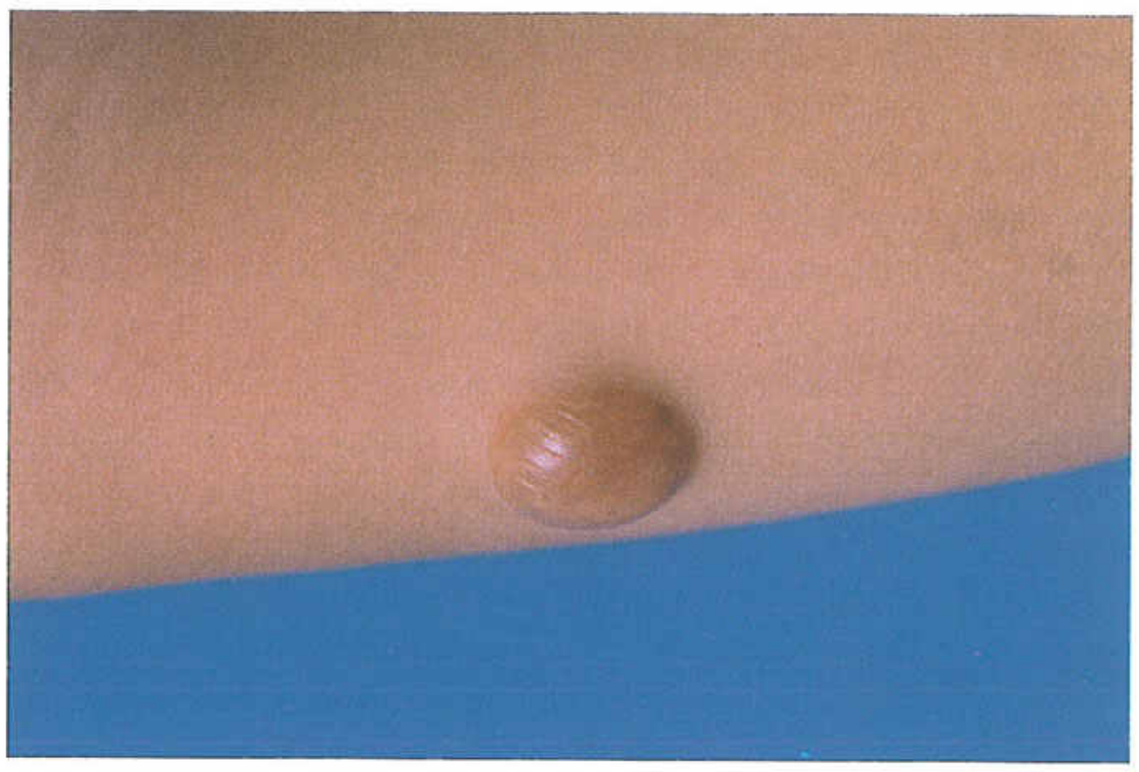


[Fig. 1]. Nodules of JXG on the arm and trunk.

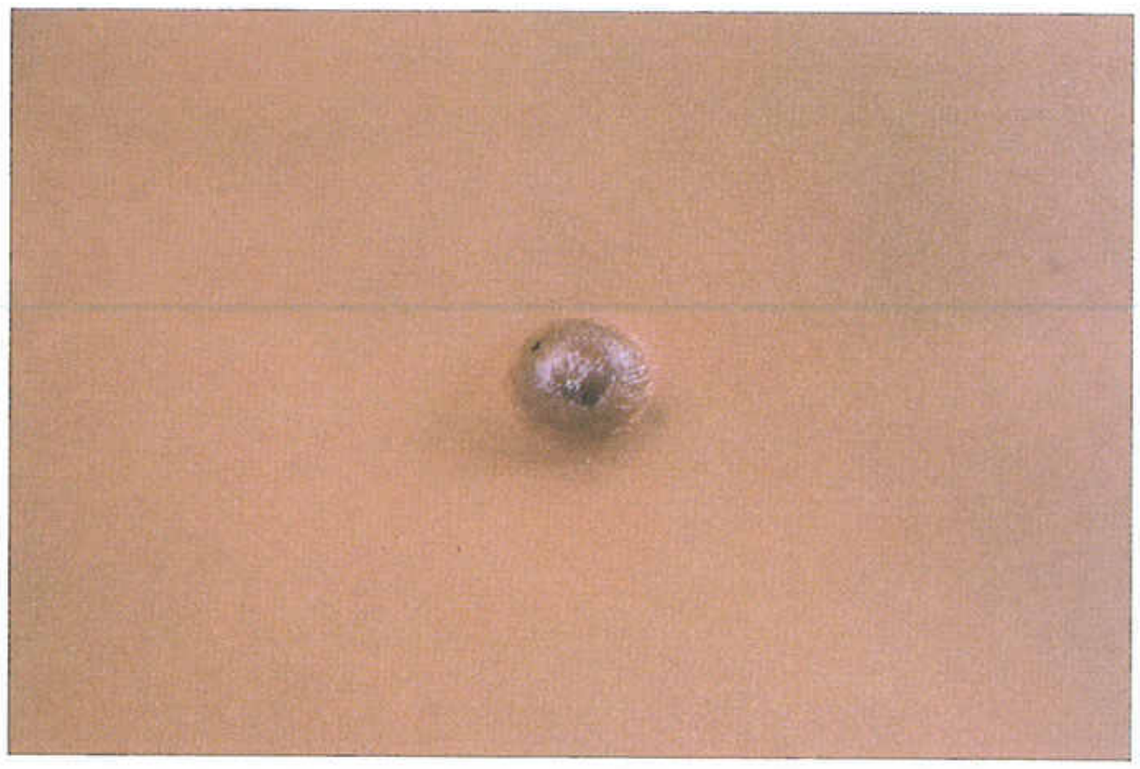
Case report

A 16 month-old boy presented with multiple nodular lesions on the arm and trunk. [Fig.1] The patient was in a good general health with no family history of similar lesions.

Physical examination revealed two different skin lesions, the first a shiny yellowish dome-shaped nodule on the inner aspect of the right arm [Fig.2]; the second a reddish-brown nodule mimicking a keratoacanthoma on the left side of the trunk. [Fig.3] No lesions were found elsewhere on the skin and there was no evidence of extra-cutaneous involvement. Laboratory investigations, which included a



[Fig.2]. Typical dome-shaped nodule of JXG on the arm.



[Fig.3]. Atypical JXG on the trunk mimicking a Keratoacanthoma.

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complete blood count, blood chemistry and lipid profile, were all normal as were also X-rays of the chest and skull and neurologic and ophthalmologic examinations.

Biopsy specimens of both lesions revealed similar histopathologic findings of Juvenile Xanthogranuloma; the entire dermis replaced by a fairly dense infiltrate of histiocytes [Fig.4] mixed with Touton-type giant cells, a few lymphocytes and eosinophils. [Fig.5]

As JXG is a self-healing disease no treatment has been given and the parents have been told about its expected natural course.

Discussion

Juvenile Xanthograuloma (JXG) is a benign, self-limiting disease of infants and children, characterized by the occurrence of discrete histiocytic tumors.(2) The majority of cases occur in infancy but older people can be affected. Infants are most likely to have multiple lesions while those older than one year tend to have solitary lesions.(3)

The etiology is unknown but the primary event seems to be a reactive hyperplasia of histiocyte elements.(4) Usually one or several reddish-yellow or brown papules or nodules erupt suddenly during the first few months of life commonly affecting the scalp, neck, trunk and extremities.(5) Atypical clinical forms of JXG, usually diagnosed after histopathologic analysis, have been reported.(6)

Extra-cutaneous involvement may occur in as many as ten per cent of cases, the eye being most commonly affected with hemorrhage into the anterior chamber. Secondary glaucoma are known to occur. Other reported extra-cutaneous sites include the lung, pericardium, liver, spleen, gastro-intestinal tract and testes.(7) Association of JXG with neurofibromatosis and chronic granulocytic

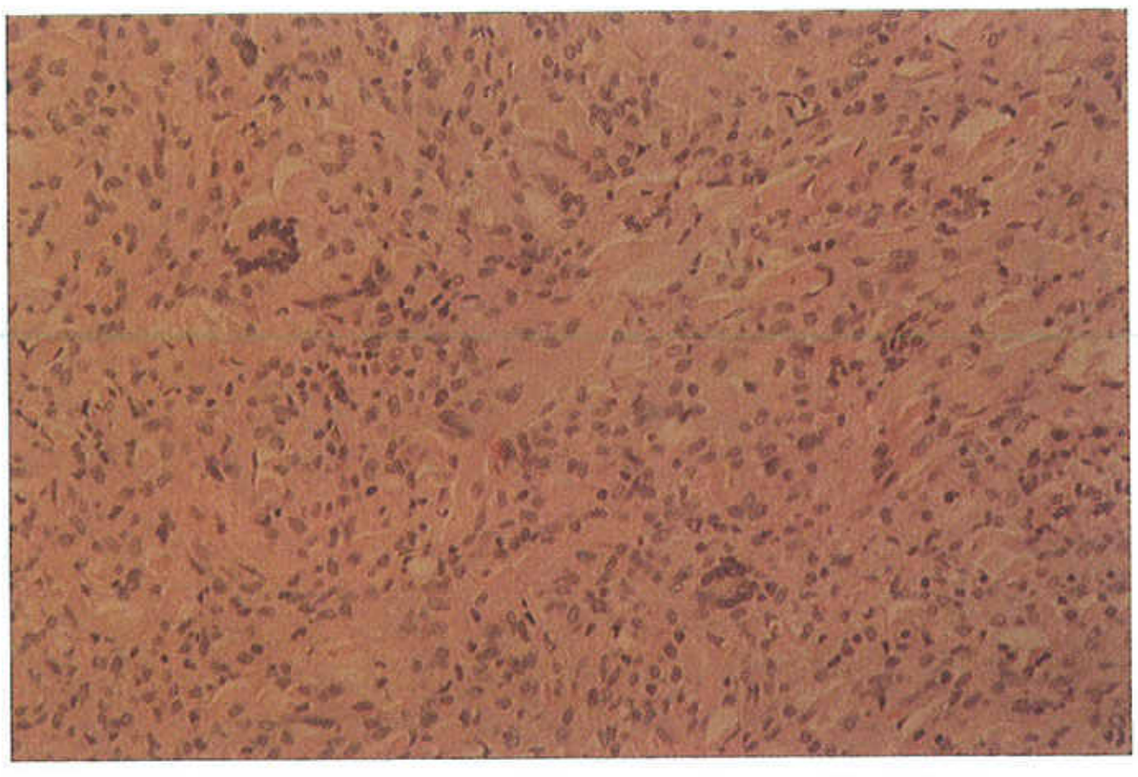
leukemia has been reported recently.(8)

Histopathologically JXG is characterized by a dense cellular infiltrate in the dermis of histiocytes, giant cells, Touton cells, lymphocytes, eosinophils and neutrophils.(9) Ultrastructurally histiocytes of JXG show no Birbeck granules (non-Langerhans histiocytic cells).(10) Immunohistochemical tests are necessary to distinguish JXG from other forms of Langerhans and non-Langerhans cell histiocytosis. The proliferating histiocytes of JXG are negative for S-100 protein and Mac-387 and positive for HAM 56 and Vimentin.(11)

Skin lesions of JXG alone do not usually require therapy because spontaneous resolution in 1-5 years is the natural course.(12) Surgery is the only other effective measure and recurrences are rare. However, the majority of ocular cases of JXG do not resolve spontaneously and treatment with topical and intra-lesional corticosteroids, radiation or surgery is indicated to prevent significant complications such as spontaneous hemorrhage or secondary glaucoma(13).

The first case of Juvenile Xanthogranuloma was reported by Adamson in 1905(1)and it has been considered the most common non-Langerhans cell histiocytosis of infants and childhood.(14) The dome-shaped nodular, yellow-brown lesions affecting the head, neck, trunk, and extremities are the usual clini-

[Fig.4]. Histiocytic cellular infiltrate occupying entire thickness of the dermis.



[Fig.5]. High-power view of Touton giant cells, histiocytes and lymphocytes cells.

cal presentation although JXG can appear at any site of the body surface including

under a toenail, on the penis, clitoris, eye lid, scrotum, lips, palms, soles and on the nose (mimicking the hemangiomatous Cyrano nose).[15]

Atypical clinical forms of JXG may mimic Dermatofibroma, Dermal nevus, Calcinosis cutis, Keloid, Pyogenic granuloma, Xanthoma, Cysts and Keratoacanthoma.[6] Giant variants of JXG have been reported to affect the scapula and the scalp.[16] JXG has also been reported as a generalized lichenoid eruption[17] and as subcutaneous lesions on the neck and scalp.[18] Extracutaneous involvement has included neurofibromatosis, mastocytosis and leukemia. [7,8]

Clinically our case had two different lesions, one typical dome-shaped nodule on the right arm and a second non-typical nodular lesion mimicking keratoacanthoma on the trunk. There was no extracutaneous involvement. Both lesions showed histopathological findings characteristic of JXG .

We felt that the presence of two clinical forms of JXG in the same patient should be recorded.

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