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Juvenile Xanthogranuloma - An Image to Remember

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Description

A 2-month-old male infant, presented with a papulonodular skin lesion on the left malar region since birth. It was tan-orange but blanchable, well-defined and about 10 mm long (Figures 1-3). He had appropriate general appearance, anterior fontanelle open and flat; chest with symmetric aeration, normal S1 and S2 and no murmur in heart auscultation; soft abdomen, with no pain at palpation, no abdominal masses; no other skin lesions were observed.

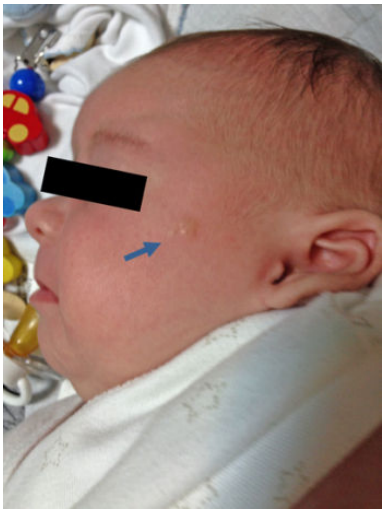

Figure 1: Orange papular lesion in the left malar region.

Medical history and clinical findings were compatible with juvenile xanthogranuloma (JXG).

JXG belongs to the broad group of non-Langerhans cell histiocytoses and typically presents in the first two years of life. 5% to 17% of JXG occur soon after birth and up to 75% occur within 9 months of birth. This patient reflects a typical case: a single reddish or yellowish papule more commonly on the head, neck, and upper trunk, with a size 0.5 to 2 cm. The etiology is unknown.

Involvement of internal organs is rare, being more frequent in the presence of multiple skin lesions. Therefore, an early diagnosis is essential. The most frequent extracutaneous manifestation is a solitary nodule or mass in the subcutaneous or deep soft tissues. Other presentations may occur, as the ocular (especially in the iris), bone (Erdheim-Chester disease), pulmonary and hepatic involvement. There seems to be an association between JXG, neurofibromatosis type 1 (in 5-10% of these patients) and juvenile myelomonocytic leukemia.

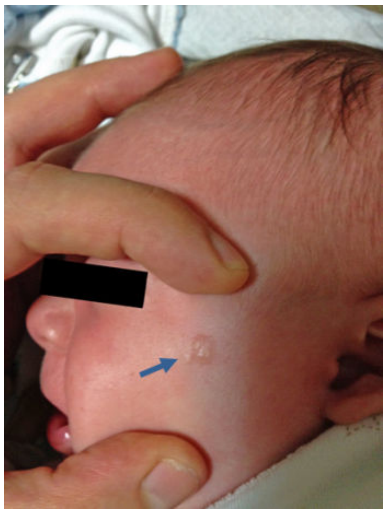

Figure 2: Papule almost disappears on finger pressure.

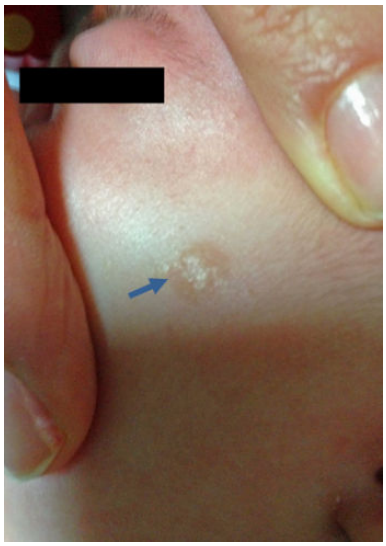

Figure 3: Orange papular lesion in the left malar region is blanchable.

The differential diagnosis includes xanthoma, Spitz nevus, contagious molluscum, hemangioma and neurofibroma. The diagnosis

is clinical in most cases, but skin biopsy is considered when there is diagnosis suspicion. Up to 10% cases occur in adulthood known as adult xanthogranuloma. Histologically, JXG is characterized by the presence of histiocytes, foam cells and giant cells of Touton.

JXG is usually a self-limited process with spontaneous regression in up to 5 years. It may leave residual hyperpigmentation, minimal atrophy or loss of elastic fibers in the dermis following regression. No treatment is required; however, lesions excision may be performed for diagnostic purposes.

Conflict of Interest

None declared.

The article has been peer-reviewed.

The authors have obtained patient consent.