ADULT XANTHOGRANULOMA – OBSERVATIONS ON A CASE WITH RARE LOCALIZATION

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Summary

Xanthogranulomas belong to non-Langerhans cell histiocytoses. Juvenile xanthogranuloma occurs in children clinically presenting as a nodule or papule with a diameter of 0.5-2 cm, red-yellow, asymptomatic, especially located on the head and neck. Histology highlights an infiltrate of foamy histiocytes and giant Touton cells in the dermis. Immunohistochemistry is positive for vimentin, CD68 and factor XIII.

A 60 years old patient presents two papules with a diameter of 4 mm and 6 mm respectively, red-yellow, asymptomatic, located on the penis. Hematological and biochemical analyzes were normal. Biopsy revealed an infiltrate of numerous foamy histiocytes and lymphocytes in the dermis. Immunohistochemistry was positive for vimentin, CD68 and factor XIII and negative for S100 and CD1a protein. Diagnosis of xanthogranuloma has been established. The lesions disappeared spontaneously after 2 months after the biopsy without relapse within the next 6 months.

Xanthogranuloma in adults is rarely encountered, the clinical, histological and immunohistochemical aspect being identical to juvenile xanthogranuloma. In contrast, adult xanthogranuloma does not show systemic damage and lesion regression is exceptional. The location of penile xanthogranuloma is very rare.

Xanthogranuloma should be considered when it comes to papulo-nodular lesions located on the penis.

***Key-words**: xanthogranuloma, unusual location, non-Langerhans-cell histiocytoses.*

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Introduction

The adult xanthogranuloma is more rarely seen than juvenile xanthogranuloma (JXG). It appears after the age of 15 years old, the clinical, histological and immunohistochemical aspect being identical to that of JXG. Unlike JXG, it is not associated with systemic damage and spontaneous regression is very rare.

Clinical case

A 60-year-old patient is consulted for the presence of two lesions on the penis that occurred two months ago. The dermatological examination reveals on the glans penis two papules slightly raised by the surface of the skin, with a smooth, well-defined surface having diameters of 4 mm and 6 mm respectively, asymptomatic, of red-yellow coloration (Fig. 1, Fig. 2). Hematological and biochemical laboratory tests including triglycerides, cholesterol, liver and kidney tests were within normal range, and syphilis tests were negative. Biopsy revealed in the dermis an infiltrate of numerous clustered foamy histiocytes and lymphocytes, as well as a small fragment of the erectile body (Fig. 3, Fig. 4). The epidermis is absent. Immunohistochemistry

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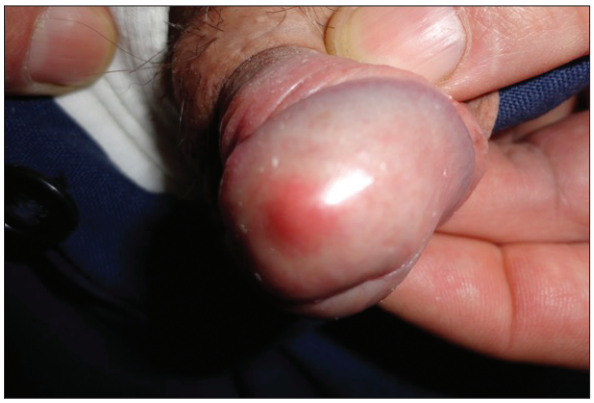

Fig. 1. Clinical appearance. Smooth-tipped papula with a red-yellowish 6 mm diameter, asymptomatic

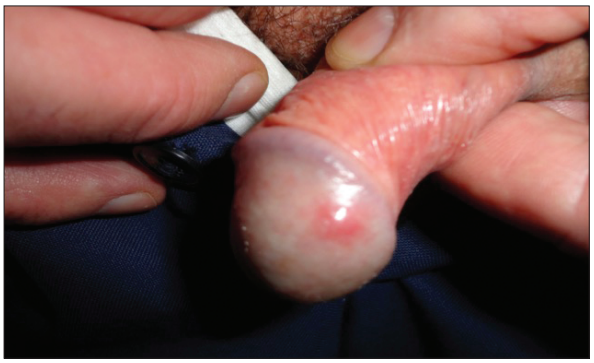

Fig. 2. Clinical appearance. Smooth-tipped papula with a red-yellowish 4 mm diameter, asymptomatic

was positive for vimentin, CD68 and factor XIII and negative for PS100 and CD1a. Clinical appearance, histopathology and immunohistochemistry have established the diagnosis of xanthogranuloma. The ophthalmologic examination was normal. The lesions disappeared spontaneously two months after the biopsy without relapses within the next 6 months.

Discussions

Xanthogranulomas belong to non-Langerhans-cell histiocytoses comprising a series of histologically entities characterized by the presence of histiocytes with different aspect (vacuolized, foamy, spindle, oncocytic or laced) associated with giant and also various aspect (Touton, en verre dépoli, Langhans type)[1].

Juvenile xanthogranuloma (JXG) is the most commonly encountered and known condition of xanthogranulomas. Described initially in 1905 as congenital multiplex xanthoma, it is renamed juvenile xanthogranuloma by Helvig and Hackney in 1954 based on histological appearance (infiltrate of xanthomized histiocytes). It occurs early, being present at birth in 5-17% of cases, and in the first years of life in 40-70% of cases[2], having an equal gender distribution[3,4]. Clinically, the JXG presents as a well-defined, round or oval, papule or well defined node with a diameter of 0.5 to 2 cm, initially red-yellowish in color, which subsequently becomes orange-

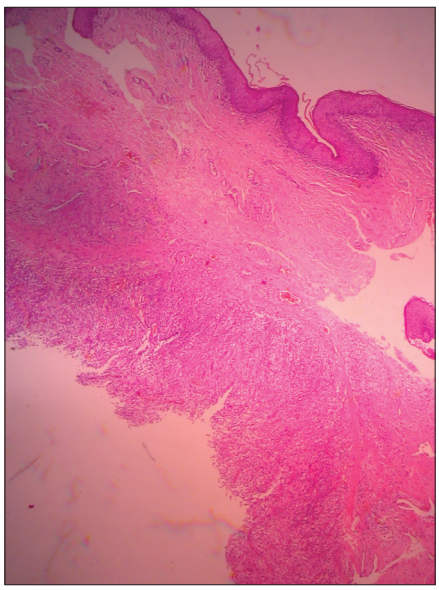

Fig. 3. Histopathological aspect. Dermic infiltrated of many histiocytes and lymphocytes

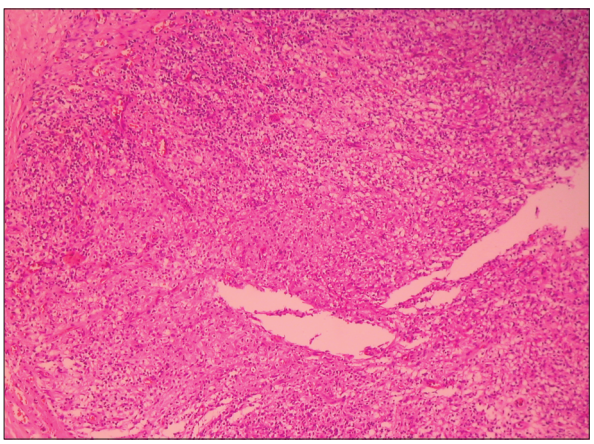

Fig. 4. Histopathological aspect. Infiltrated into the deep dermis. Epiderm absent

yellow or brown, asymptomatic. In over 40-60% of patients, the JXG presents as a single lesion, but multiple or disseminated forms also exists. It is often located on the head, neck, trunk and limbs, but can appear on any skin area like palms, plants, toes, external genitalia, eyelids, lips and mucous membranes[5,6,7]. JXG is regresing spontaneously with complete extinction after 3-6 months, residual hyperpigmentation, atrophy or anetodermia [8] may persist in place of the lesion. The excision of the lesion can be followed by relapses.

Histology of JXG consists of a histiocytic infiltrate located in the papillary dermis and sometimes in the reticular dermis with the possibility of extension in the subcutaneous tissue, fascia and muscles. The infiltrate is predominantly composed of foamy histiocytes, but also of giant Touton cells (xantomized multinucleated cell), lymphocytes, neutrophils and sometimes mast cells. Nuclear atypia and mitosis may be present. Infiltrate can lead to thinning of the epidermis or ulceration. Regression of the lesion is accompanied by progressive appearance of fibroblasts and fibrosis. Immunohistochemistry is positive for vimentin, CD68, factor XIII and negative for PS100, CD34 and CD1a.

Eye involvement is the most common extracutaneous affection of JXG followed by pulmonary and hepatic involvement. The coexistence of JXG with type 1 neurofibromatosis represents an increased risk of monocytic leukemia.

Adult xanthogranuloma is rarely encountered after 15 years of age, the peak of incidence being between 20 and 30 years of age but lesions can also appear at the age of 80[9]. The adult xanthogranuloma has a male predominance and is not associated with systemic impairment. The clinical, histological and immunohistochemical aspect is similar to JXG. Spontaneous involution is rarely encountered[10]. The presented case is of an adult showing two papullous XG lesions located on the penis, with only four cases cited in the literature with this localisation[11, 12, 13]. The two lesions involuated spontaneously two months after the biopsy without relapses within the next 6 months.

In conclusion, xanthogranuloma diagnosis should be considered in the differential diagnosis of penile papulo-nodular lesions.

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Conflict of interest
NONE DECLARED

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