

## Case for diagnosis

### Caso para diagnóstico

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#### CASE REPORT

A 28 year-old male patient, military officer, presented with an asymptomatic solitary papule on his right thigh, which had appeared two months before his first doctor's appointment. He lacked any systemic signs or symptoms and had no relevant personal medical records. On physical examination, there was a firm, well-demarcated, smooth-surfaced papule, 0.5 cm in diameter, located in the medial aspect of his right thigh, without adenopathy (Figure 1). Histopathology showed thinned epidermis and dermis with diffuse inflammatory infiltrate of lymphocytes and histiocytes associated with giant multinucleated cells (Figure 2), containing a smooth eosinophilic central cytoplasm with a ring of nuclei and a foamy external cytoplasm characterizing Touton cells. Immunohistochemistry revealed CD68 positivity, CD4 and vimentin, and CD1a and S100 negativity (Figure 3). A complete blood count and a lipid profile were ordered, both with normal results. The patient was evaluated by an ophthalmologist, with no abnormal findings.



FIGURE 1: Yellowish, firm, well-demarcated, smooth-surfaced, erythematous papule, with normal adjacent skin

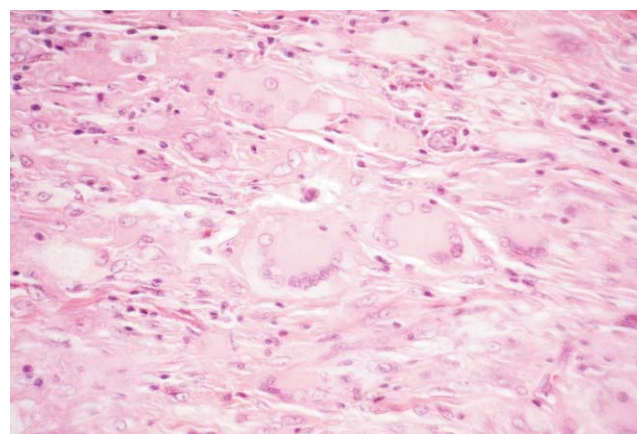


FIGURE 2: H&E 400X. Dermis with diffuse inflammatory infiltrate, composed mainly of lymphocytes and histiocytes, associated with Touton giant cells

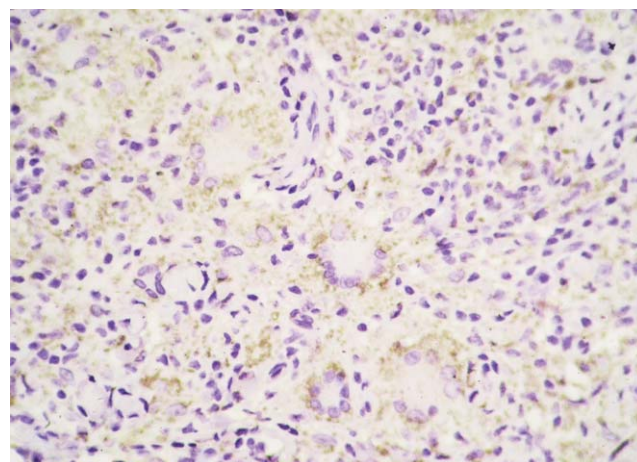


FIGURE 3: Immunohistochemistry - CD68 marker. Immunohistochemistry positive for CD68. It was positive for CD4 and vimentin, but negative for CD1a and S100

Received on 10.11.2011.

Approved by the Advisory Board and accepted for publication on 08.12.2011.

\* Work conducted at Hospital Central do Exército (HCE) – Benfica (RJ), Brazil.

Conflict of interest: None

Financial funding: None

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## DISCUSSION

Xanthogranuloma (XG) is a form of histiocytosis classified by the Histiocyte Society as class IIa, derived from dermal dendrocytes.<sup>1</sup> Its etiology is unknown and it has a benign and self-limited course. It most often occurs in childhood, especially early in life, but may be present at birth.<sup>2</sup> Men are more commonly affected than women by a ratio of 12:1.<sup>3</sup> It may occur in adulthood in 10-30% of the cases, usually in the second and third decades of life.<sup>4</sup>

Clinically, XG presents as one or multiple yellowish-erythematous papules and nodules, usually a few millimeters to centimeters in diameter, mainly on the face, neck and back. Mucosal involvement has been described. It can occur at any anatomical site, but the eyes are the most common extracutaneous site, followed by lung, kidney, central nervous system, colon, pericardium, ovary, liver and spleen.<sup>5</sup>

Histopathology varies according to the evolutive phase, but it classically presents dense inflammatory infiltrate, unencapsulated, composed mainly of lymphocytes and histiocytes, associated with Touton giant cells.<sup>6</sup> Immunohistochemistry is negative for CD1a and S-100 and positive for CD68, CD4, vimentin and factor XIIIa.

XG tends to spontaneous involution in 3-6 years, leaving a residual hyperpigmented and atrophic site. Surgical excision may be performed. In patients with multiple lesions or systemic involvement, the use of steroids, vincristine, vinblastine and 5-fluorouracil has been reported with conflicting results. Although current literature describes juvenile and adult-type XG as a single entity, some authors claim they are distinct diseases.<sup>7,8</sup> □

**Abstract:** Male patient, 28 years old, presented with an asymptomatic yellowish erythematous papule on his right thigh. Excisional biopsy was performed for histopathological examination of the lesion. Multinucleated cells (Touton giant cells) were observed. S100 immunohistochemistry was negative for CD1a and positive for CD4 and CD68. Based on clinical and histopathological findings associated with immunohistochemistry, we concluded that it was a case of adult-type xanthogranuloma. Because it was a solitary lesion without other clinical signs and symptoms, the medical conduct adopted was patient orientation.

**Keywords:** Histiocytosis; Histiocytosis, non-Langerhans-cell; Xanthogranuloma, juvenile

**Resumo:** Paciente do sexo masculino, 28 anos, com surgimento de pápula eritemato-amarelada na coxa direita, assintomática. Foi realizado biópsia excisional da lesão para exame histopatológico, tendo sido observado células gigantes multinucleares do tipo Touton e imunohistoquímica com CD1a e S100 negativos e CD4 e CD68 positivos. Pelos achados clínico-histopatológicos associados a imunohistoquímica concluímos ser um caso de xantogranuloma do tipo adulto. Por ter sido uma lesão solitária sem outros sinais e sintomas clínicos, a conduta adotada foi orientação quanto a doença.

**Palavras-chave:** Histiocitose; Histiocitose de células não Langerhans; Xantogranuloma juvenil

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How to cite this article: Sampaio FMS, Lourenço FT, Obadia DL, Nascimento LV. Case for diagnosis. Solitary papule in the thigh: an uncommon diagnosis. *An Bras Dermatol.* 2012;87(5):789-90.