

## Case Reports

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Received on: 21/01/2020

Approved on: 18/02/2021

Study conducted at the Universidade Federal da Fronteira Sul, Passo Fundo (RS), Brazil.

Financial support: None.

Conflict of interest: None.



# Multiple adult xanthogranuloma: case report of successful treatment with CO<sub>2</sub> Laser

*Xantogranuloma múltiplo do adulto: relato de caso de tratamento exitoso com laser CO<sub>2</sub>*

DOI: <https://www.dx.doi.org/10.5935/scd1984-8773.20201242515>

### ABSTRACT

Multiple adult xanthogranuloma is a rare and late variant of Juvenile xanthogranuloma, a non-Langerhans cell histiocytosis. It usually corresponds to a single lesion in adults, and the manifestation of multiples lesions is uncommon. We report a case of multiple adult xanthogranuloma, with Isotretinoin therapy failure and optimal response to CO<sub>2</sub> Laser treatment in the surgical mode.

Keywords: Histiocytosis non-Langerhans-cell; Lasers gas; Xanthogranuloma juvenile

### RESUMO

O xantogranuloma múltiplo do adulto é uma apresentação mais rara e tardia do xantogranuloma juvenil, uma histiocitose de células não Langerhans. No adulto, normalmente, é uma lesão única, sendo a manifestação por múltiplas lesões infrequente e pouco descrita na literatura. Relatamos um caso de xantogranuloma múltiplo do adulto, com falha terapêutica à isotretinoína e ótima resposta ao tratamento com laser CO<sub>2</sub> no modo cirúrgico.

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

### INTRODUCTION

Xanthogranuloma is a benign and self-limited dermatosis characterized by non-neoplastic proliferation of histiocytes. It has phenotypic features different from Langerhans cells, with CD1a and S100 negative in immunohistochemistry and absence of Birbeck granules (cytoplasmic organelles identical to those of the Langerhans cell) in electron microscopy.<sup>1</sup> Juvenile xanthogranuloma, the most common form of non-Langerhans cell histiocytosis, is prevalent in children, presenting as multiple yellowish-brown papules or nodules distributed preferentially in the head and neck region. The appearance in adults is rare, with few cases published since its first description in 1963 by Gartmann and Titsch. It usually manifests as a single lesion and, exceptionally, as multiple lesions, this form being called multiple adult xanthogranuloma.<sup>2</sup> Cutaneous involvement is the rule, but extracutaneous manifestations have also been reported.

Possible spontaneous involution may occur. However, treatment with local cryotherapy, lesion excision, carbon dioxide laser (CO<sub>2</sub> laser), and oral retinoids<sup>5</sup> are potential therapies. We present a case of multiple adult xanthogranuloma without extra-cutaneous manifestation.

### CASE REPORT

A 37-year-old man, previously healthy, living in an urban area, presented asymptomatic symptoms but progressive papular lesions on the face for 15 days, without any other complaints. He denied external factors related to the onset of the condition.

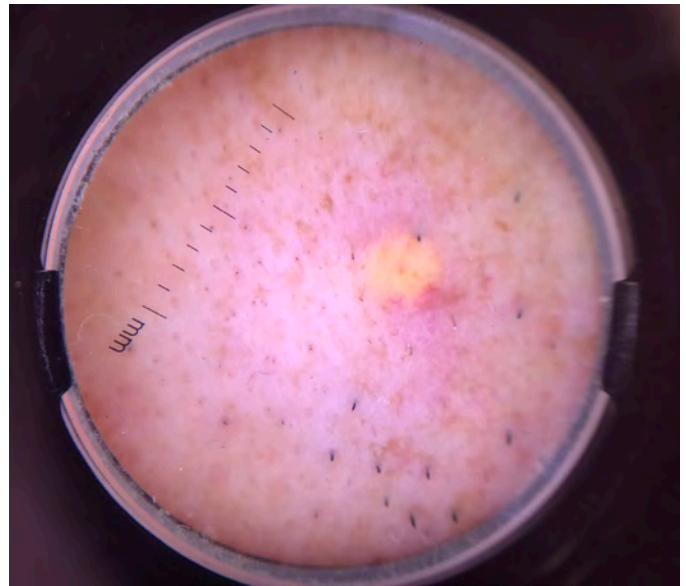
The dermatological examination showed several papules, normochromic to slightly yellow, firm, shiny, and well defined. The papules were diffuse on the face and some on the anterior chest, without central umbilication (Figures 1 and 2). The patient underwent a complete physical examination without adenomegaly or visceromegaly.

We investigated one of the lesions through excisional biopsy. The anatomopathological examination showed dermal cell proliferation with numerous foamy histiocytes and giant cells (Touton cells). These findings are compatible with xanthogranuloma (Figure 3). Immunohistochemical analysis revealed expression of CD68 (clone PGM1) and absence of expression of Langerhans cell markers S100 (polyclonal) and CD1a (clone 010) (Figure 4).

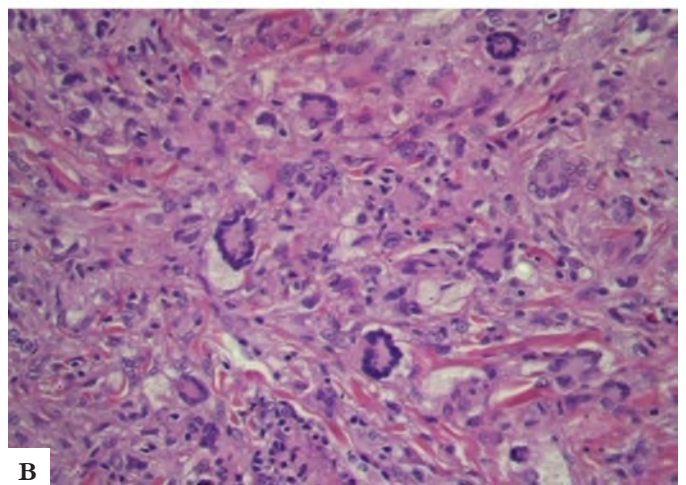
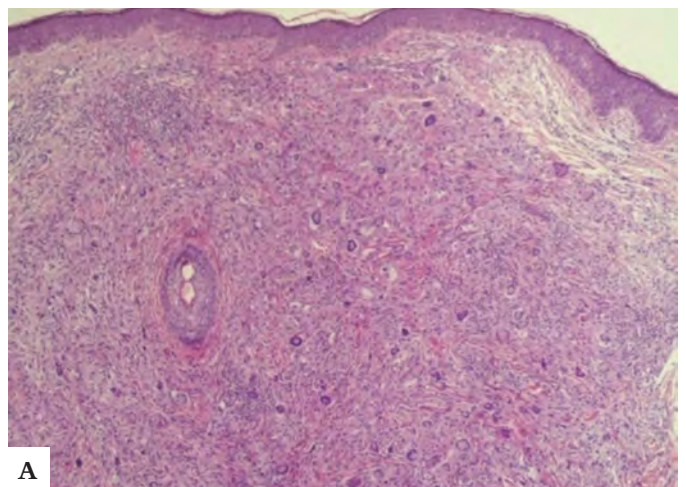
After assessing the clinical aspect of the patient and the anatomopathological and immunohistochemical findings, we concluded the diagnosis of multiple adult xanthogranuloma. Visceral involvement was assessed, with laboratory tests, chest radiography, ultrasound of the total abdomen, and ophthalmological evaluation, with no evidence of abnormalities.



**FIGURE 1:** Observation of several normochromic papules, slightly yellow, firm, and shiny

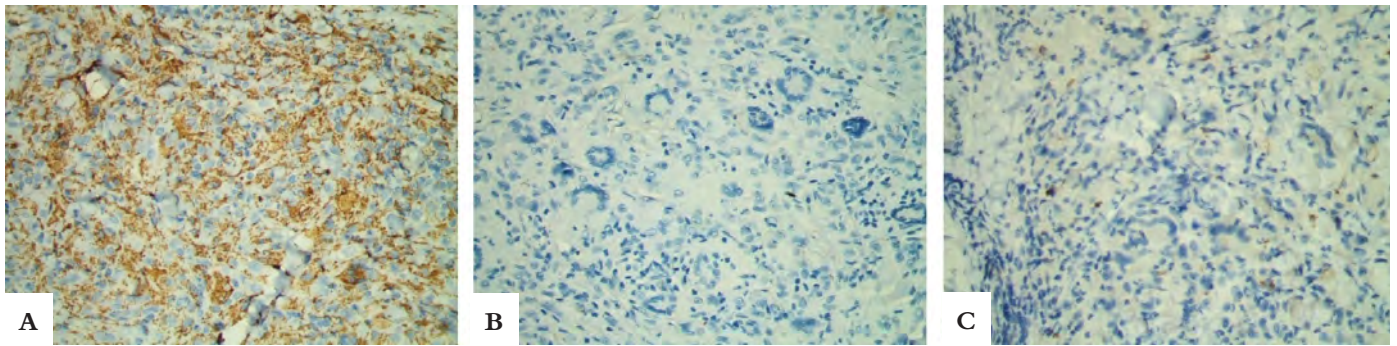


**FIGURE 2:** Dermoscopy showed yellowish papule with slightly erythematous halo and vessels on the periphery of the lesion



**FIGURE 3:** Skin showing dermal cell proliferation with the presence of numerous foamy histiocytes and Touton giant cells, compatible with xanthogranuloma. Magnifications 10x (image A) and 40x (image B)





**Figure 4:** Immunohistochemical examination with CD68 expression observed in image A, absence of expression of Langerhans cell markers S100 and CD1a observed, respectively, in images B and C

Due to the extent of cutaneous involvement and the patient's discomfort, we decided to start isotretinoin 20 mg/day. After 30 days, there was an increase in the number of lesions. Thus, the medication was suspended, and CO<sub>2</sub> laser sessions were performed in the surgical mode with excellent aesthetic results (Figure 5).

## DISCUSSION

Multiple adult xanthogranuloma is a non-Langerhans cell histiocytosis. It is considered a rare and later manifestation of juvenile xanthogranuloma, occurring most frequently between the third and fourth decades of life,<sup>3</sup> and the mean age at diagnosis is 47 years. The disease is slightly more common in men, at a ratio of 1.6/1.<sup>4</sup> Clinically, it presents with five or more lesions, characterized by erythematous-yellow, firm, well-defined, asymmetric, and asymptomatic papules affecting predominantly the trunk, followed by the face and, less commonly, in the limbs.<sup>4</sup> With the evolution of the condition, the lesions become brownish-yellow and present telangiectasias, which may result in small atrophic scars or hyperpigmentation after resolution.<sup>3,5</sup> Differing from the juvenile form, which tends to involve in three to six years, spontaneous involution of the lesion in adults is not common.<sup>6</sup>

Despite preferentially affecting the skin, the disease can have extracutaneous involvement in 4% of children and 5-10% of adults, and the eyeball is the most frequently affected. Extracutaneous manifestations, which include liver, spleen, lungs, central nervous system, and hematopoietic tissue,<sup>3,5</sup> can cause, for example, thrombocytosis and monoclonal gammopathy. As a dermatological manifestation, there are reports of underlying hematological neoplasia, which may appear before, concomitantly, or after the development of the disease, being considered a cutaneous marker of hematological disease.<sup>8</sup> There is a hypothesis that excess gamma globulins in leukemic states could stimulate histiocytic production, resulting in the development of lesions.<sup>4</sup> Serum lipid profiles are normal in patients with adult and juvenile xanthogranuloma.<sup>7</sup> The etiology of xanthogranuloma is unknown; associations with trauma, infections, and neoplasms have been suggested.<sup>8</sup>



**FIGURE 5:**  
Post CO<sub>2</sub> result

The tumor presents an accumulation of differentiated histiocytes that express the phenotype of dermal dendrocytes, although studies suggest that the cells of its origin could be plasmacytoid monocytes.<sup>7</sup>

From the histological point of view, the lesions present a dermal cell infiltrate composed of histiocytes, lymphocytes, eosinophils, and, occasionally, neutrophils. They exhibit a pleomorphic histiocytic infiltrate with a predominance of empty cells without lipids at the beginning. Later foam cells prevail, corresponding to lipid-filled histiocytes,<sup>2</sup> together with the appearance of multinucleated giant cells, the Touton cells, in 95% of cases (multinucleated xanthomatous giant cells with nuclei arranged in a wreath-like pattern).<sup>3</sup> The appearance of histiocytes laden

with lipids and giant cells occurs late and is probably a secondary event in response to cytokine production by the lesion histiocyte.<sup>7</sup> In the immunohistochemical study, histiocytes differ from Langerhans cells by positive CD68 and HAM56 macrophage cell markers as well as dermal dendrocyte markers (FXIIIa), presenting negative Langerhans cell markers S100 and CD1a. There is also a lack of Birbeck granules inside cells in the ultrastructural study.<sup>2</sup> In doubtful cases, the expression of factor XIIIa offers more evidence for the diagnosis of xanthogranuloma.<sup>6</sup>

Juvenile xanthogranuloma was noted in association with different diseases, such as neurofibromatosis, Niemann–Pick disease, urticaria pigmentosa, and juvenile chronic myelomonocytic leukemia.<sup>7</sup> The differential diagnosis is made between diseases classified as Langerhans cell histiocytosis, in addition to molluscum contagiosum, cryptococcosis, lepromatous leprosy, and neurofibromatosis.<sup>8</sup>

Therapeutic management is conservative, with spontaneous involution being possible but unlikely. Local cryotherapy, surgical excision, carbon dioxide laser (CO<sub>2</sub> laser), and oral retinoids are reported, with an emphasis on isotretinoin.<sup>5,7,8</sup>


Extensive skin involvement, with several lesions, of the adult form of the condition, can cause unsightly discomfort and require treatment to accelerate the improvement of the disease. ●

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