Granulomatosis disciformis in a non-diabetic patient*

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DOI: http://dx.doi.org/10.1590/abd1806-4841.20175761

Dear Editor,

Miescher's Granulomatosis (also known as *granulomatosis* disciformis chronica et progressiva) (GDML) is a chronic disease of unknown etiology, with clinical characteristics that are quite similar to lipoidic necrobiosis (LN) but that differ from this disease due to its anatomopathologic characteristics. In GDML, one can generally observe an exuberant granulomatous reaction, comprised of epithelioid cells, with gigantocytes and plasmocytes, and without necrosis, whereas in the LN, one can observe palisading granulomas with necrobiosis.[1,2]

Few studies have reported on this disease in the literature; however, it is believed that GDML is more commonly found in middle-aged women. Initially, it was proposed that the core difference between GDML and LN was the fact that the former was not associated with diabetes mellitus, with only LN associated to this condition. However, this theory was proven to be unfounded, as it was discovered that both diseases can be associated or not with diabetes mellitus.[1,2,3]

In the present case, a 46-year-old female patient reported the appearance of annular lesions on her forearms and legs over the past three years, with local yet discreet pruritis. The patient presented systemic arterial hypertension, which was treated with propranolol, and the patient denied any prior diagnosis of diabetes mellitus. Upon physical examination, it was possible to observe annular erythematous plaques, with infiltrated edges, together with an atrophic and telangiectatic center, with a yellowish color and an aspect of cigarette paper when wrinkled (Figures 1 and 2). The initial hypothetical diagnosis was of LN, and a biopsy of a lesion on the right forearm was, therefore, performed. The anatomopathologic exam showed a perivascular and interstitial inflammatory infiltrate, comprised of lymphocytes, plasmocytes, and epithelioid macrophages, with the presence of Langhans multinucleated giant cells and thick collagen fibers (Figure 3). The biopsy proved to be compatible with the diagnosis of GDML. Laboratory exams were also requested to investigate the lipid profile and the change in the glycemic curve, which presented no changes. The patient began her treatment with sulfone, administered orally, and topical clobetasol presenting, to date, partial regression of the lesions.

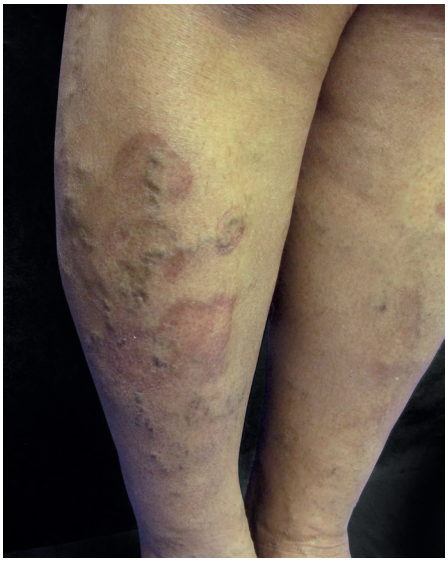

Figure 1: Annular erythematous plaques, with infiltrated edges and a yellowish, atrophic, and telangiectatic center on the legs

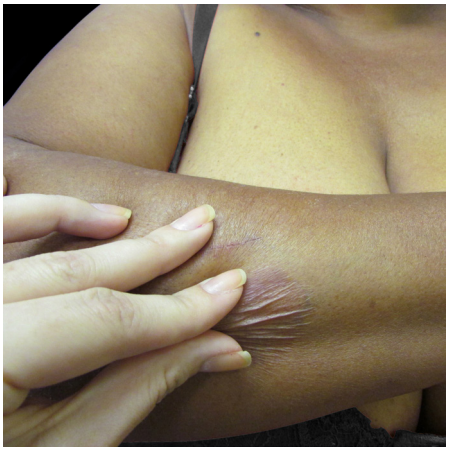

Figure 2: Lesion with an aspect of "cigarette paper" when wrinkled

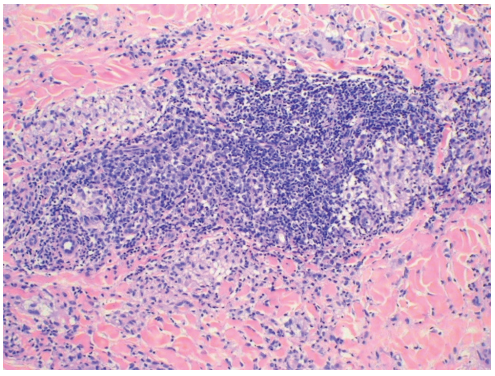

Figure 3: Sclerotic dermis, with tuberculoid granuloma and dense perivascular lymphoplasmacytic component (Hematoxylin & eosin x100)

Received on 04.03.2016
Approved by the Advisory Board and accepted for publication on 17.10.2016
* Work conducted at Santo Amaro University, São Paulo, SP, Brazil.
Financial support: None.
Conflict of interests: None.

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GDML was described for the first time by Miescher and Leder in 1948. The authors illustrated great clinical similarities of this entity with LN, a chronic disease of unknown origin but that presented distinct histopathological characteristics.[2]

By contrast, other authors, such as Ringrose *et al*.,[4] believed that GDML was a non-diabetic variant of LN. Later, however, it was shown that many of the GDML patients were in fact pre-diabetics or presented change in their glucose curves, proving that both co-morbidities can be concomitant.[1]

The etiology, of both LN and GDML, is still rather unknown. It is believed that changes in the small blood vessels of the skin and hypodermis are important in their pathogeneses, and that habits, like smoking, can worsen vascular damage or even precipitate the clinical manifestation of the disease. The amount of smoking and the beginning of the smoking habit are important risk factors for the development of GDML. Other factors include venous stasis, genetic predisposition, and trauma.[4]

Clinically, GDML can also be confused with LN. The lesions are normally bilateral and symmetric on the anterior surface of the lower and upper limbs, slightly yellow, of a firm consistency, of a translucent and shiny surface, with a discretely atrophic center and subtly raised edges.[1,2,3]

From the histopathological point of view, GDML joins the following aspects that differentiate it from LN. First, the intense participation of the hypodermis can be observed in specific cases, together with a larger quantity of plasmocytes in the site; a minimal degree of necrobiosis; an area of hyalinization of collagen; and the absence or lack of mucin deposits. By contrast, in the LM, one can almost always observe the presence of "palisading granulomas" with necrobiosis.[1,2,3]

The treatment for LN and GDML is difficult to determine, as strong topical corticosteroids, systemic corticosteroids, pentoxifylline, puva therapy, mycophenolate mofetil, chloroquine, sulfone, and anti-TNFs can all be used. It is important to highlight that one of the main long-term risks of NL is its transformation in squamous cell carcinoma.[5]

The present case reported on a middle-aged, non-diabetic female patient who had reported the appearance of lesions on her limbs three years three years before. The clinical condition of this case was very similar to that of LN, and the diagnosis of the disease was only able to be confirmed by performing an anatomopathologic exam that was compatible with GDML.

It is important to note that, due to its rarity and its morphological similarity to LN, GDML is subject to being clinically underdiagnosed, and a biopsy should always be performed to confirm a proper histopathological diagnosis. ❑

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How to cite this article: *Oliveira ARFM, Almeidinha YD, Oliveira Filho J, Michalany AO. Granulomatosis disciformis in non-diabetic patient. An Bras Dermatol. 2017;92(3):438-9.*

Erythematous, vesicular, and circinate lesions in a 78-year-old female – benign familial pemphigus*

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DOI: http://dx.doi.org/10.1590/abd1806-4841.20176711

Dear Editor,

Hailey-Hailey disease, also called benign familial pemphigus, is an autosomal dominant disorder caused by mutations in the ATP2C1 gene. Positive family history is detected in two thirds of all cases. The prevalence is 1:50,000 and the incidence of sporadic mutations might be as high as 26%.[1,2] We present a case of a patient who developed benign familial pemphigus in the seventh decade of life without any medical family history.

A 78-year-old white woman with hypertension and dyslipidemia presented with erythematous plaques in the groin with satellite pustules diagnosed as candidal intertrigo. The lesions had presented for 4 months and showed no improvement under topical and oral antifungal treatment (Figure 1). The patient reported severe itching. In two weeks' time, a new perineal erythema appeared with linear erosions and eczematous, circinate lesions with some flaccid vesicles involving her neck, back, and upper and lower extremities (Figure 2). The mucosal membranes were not involved. Blood test

Received on 04.03.2016
Approved by the Advisory Board and accepted for publication on 16.12.2016
* Work performed at Hospital Universitari Sagrat Cor – Grupo Quirónsalud – Barcelona, Spain.
Financial support: None.
Conflict of interests: None.

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