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CASE REPORT **OPEN ACCESS**

INTERSTITIAL GRANULOMATOUS DERMATITIS ASSOCIATED WITH AUTOIMMUNE HEPATITIS: CASE REPORT

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ABSTRACT

Background: Interstitial granulomatous dermatitis (DGI) is a skin inflammatory reaction described in association with autoimmune diseases. Clinical expression is variable, ranging from linear subcutaneous and hardened cords to symmetrical violet macules until erythematous plaques on the upper part of the trunk and proximal limbs. The definitive diagnosis is given by histopathological analyze. **Main observations:** We report a rare case of a 28 years old, female pregnant, with prior diagnosis of autoimmune hepatitis that sudden and progressive appearance of erythematous-violaceous plaques, located in sun-exposed body areas. After few exams the biopsy was definitive for the diagnosis of DGI. **Conclusions:** It may be suspected that DGI was triggered in our patient by medications use to control his primary desease. Our case suggests that DGI has to be considered as a differential diagnosis to dermatoses that are present simultaneously with autoimmune disease.

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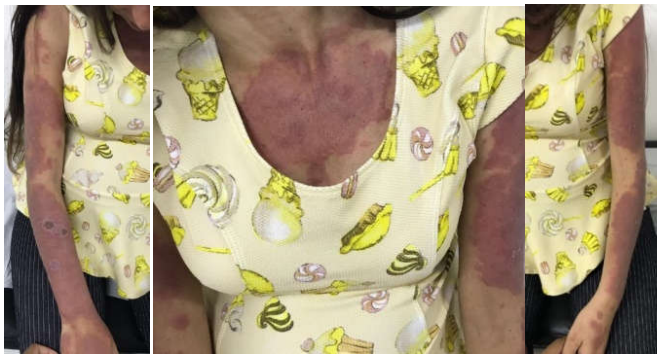
INTRODUCTION

Interstitial granulomatous dermatitis (DGI) is a skin inflammatory reaction described in association with autoimmune diseases, such as: lupus, vasculitis, arthritis rheumatoid, arthritis reactive, lymphoproliferative diseases, solid organ cancer and medications (Tomasini, 2002; Lee, 2005; Lee, 2007; Rosenbach, 2015 and Rato, 2018). There are few reports in the literature on the association of DGI with autoimmune hepatitis, the first report being described in 2007 by Lee et al. Clinical expression is variable (Rato, 2018), ranging from linear subcutaneous and hardened cords to symmetrical violet macules until erythematous plaques on the upper part of the trunk and proximal limbs (Rato, 2018; Dykman, 1965 and Peroni, 2011). The definitive diagnosis is given by histopathological analyses. There is a relation between DGI and palisade neutrophilic granulomatous dermatitis (DGNP), both are part of autoimmunity-related granulomatous dermatitis (DGRA) (Lee, 2007 and Chu, 1994). Skin lesions can have the same clinical presentation and histopathology presents specific findings that can assist in the differential diagnosis (Hawryluk, 2010).

Case Report

Female, 28 years old, with 27 weeks of gestation, sudden and progressive appearance of erythematous-violaceous plaques, slightly keratosics, with some areas of exulceration, located in sun-exposed body areas (face, upper chest, arms and legs) with mild local itching (Figure 1). Despite the nonspecific clinical presentation, skin biopsy was definitive for the diagnosis of DGI. Epidermis exhibited hyperkeratosis, orthokeratosis, acanthosis and minimal spongiosisdifuse. In the upper dermis, fibroblast proliferation, capillary proliferation, tumefied endothelial cells and perivascular infiltrates of lymphocytes, histiocytes and a few giant Langhan-type cells were found, in addition to frequently melanophages. Absence of signs of vasculitis and muciparous deposits.Fite-Faraco-stained sections did not reveal BAAR (Figure 2). An immunohistochemistry revealed CD68 positive in multinucleated cells and giant cells, predominant presence of CD3 positive T lymphocytes and CD20 positivity in some B lymphocytes (Figure 3). The diagnosis of autoimmune hepatitis was done at age 16 years old, by the presence of hypergammaglobulinemia IgG and high titers of anti-smooth

muscle antibodies. There was no positivity for anti-mitochondrial antibodies and serology for hepatitis B and C was negative.



Figur 1. Clinical presentation: Slightly keratosic erythematous-violet plaques with areas of exulceration

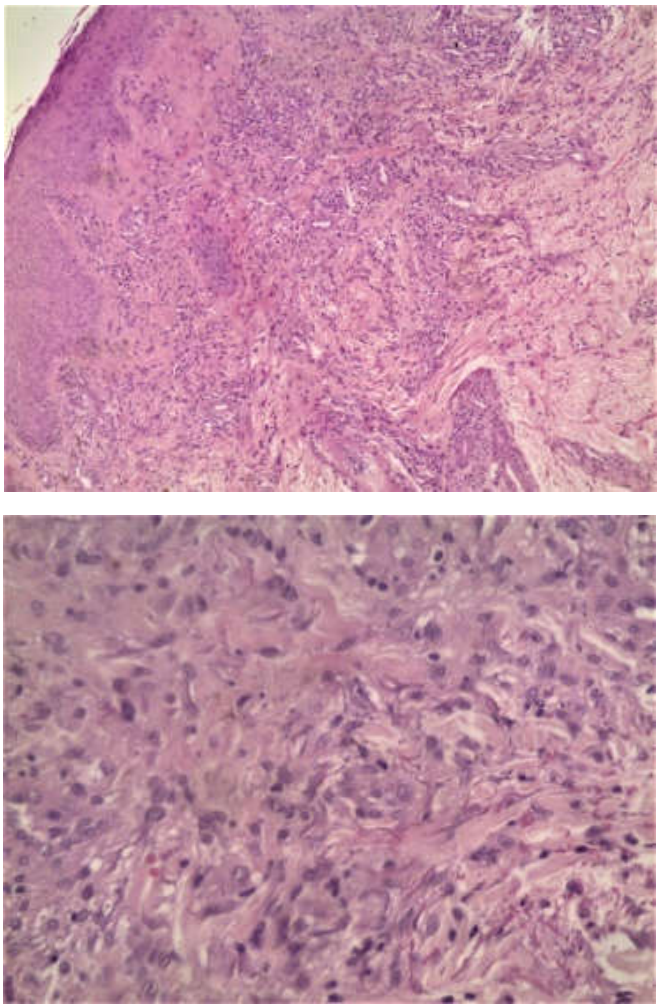


Figure 2.a, 2.b. Pathologicala) HE 10X epidermis with an irregular acanthosis and a cellular infiltrate affecting the superficial dermis and middle dermis b) HE 40X predominant presence o epithelioid pattern histiocytes, outlining granulomas, few lymphocytes

The patient underwent treatment for autoimmune hepatitis with azathioprine, but the medication was discontinued at the time of pregnancy diagnosis. During hospitalization for treatment of skin desease, the use of systemic corticosteroids was prescribed for a period of 43 days, with almost total improvement of the lesions until the time of hospital discharge.

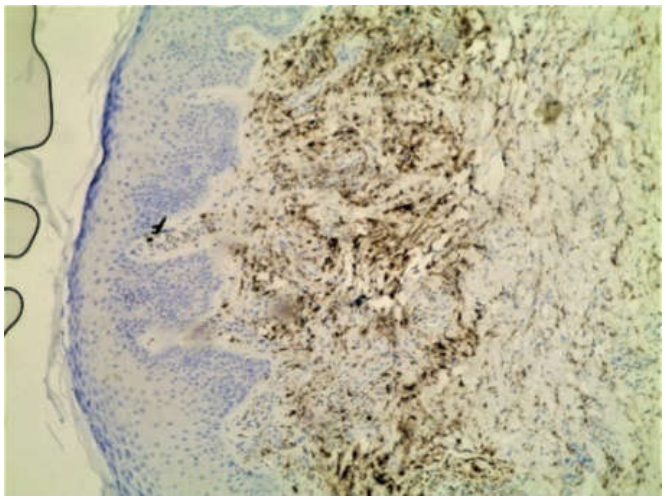


Figure 3. Immunohistochemistry: CD68

DISCUSSION

DGI is a chronic inflammation of the skin that was first described by Ackerman in 1993. At this initial report the clinical lesion consisted of the presence of hard subcutaneous and linear cords in the anterior thorax. After this report, other diverse clinical presentations were described, such as: nodules, macules and plaques (Chu, 2011). In our case report the patient developed slightly keratotic violet erythematous plaques on the face, upper chest, arms and legs. The diagnosis of autoimmune hepatitis is characterized by the presence of hepatitis on histological examination, a compatible clinical picture, laboratory findings (increase in serum globulins and the presence of one or more characteristic autoantibodies) and exclusion of chronic liver diseases, including viral hepatitis and Wilson's Desease (Lee, 2007; Albert, 2002 and Correia, 2013). The patient in our report had negative serology for hepatitis B and C. However have positivity for specific antibody such as the presence of smooth anti-muscle in high titers, absence of anti-mitochondrial antibody and IgG hypergammaglobulinemia confirming the diagnostic. The cause of DGI remains unknown. Even so, the association with autoimmune diseases makes probable pathogenesis mediated by the immune complex (Verneuil, 2001). In our case, skin lesions arose after discontinuation of Azathioprine therapy during pregnancy. This fact may support the hypothesis that the deposition of immunes complex are related the pathogenesis of DGI.

The definitive diagnosis is histopathological and is characterized by the presence of a dermal histiocytic infiltrate with histiocytes frequently arranged around degenerate collagen (Peroni, 2011 and Szepetiuk, 2012), rare eosinophils and neutrophils can be observed, (Altaykan, 2004) there is usually no vasculitis and mucin deposition is minimal or nonexistent (Peroni, 2011 and Altaykan, 2004). In some cases, differentiating between DGI and DGNP is difficult, since both conditions correspond to the group of granulomatous autoimmunity-related dermatitis (DGRA) (Lee, 2007 and Correia, 2013). In the histopathological evaluation, inflammatory cell infiltration is usually scarce to moderate in DGI, while at DGNP there is a dense histiocytic and neutrophilic interstitial infiltrate (Hawryluk, 2010). Furthermore, vasculitis may occur in DGNP, while in DGI it is generally absent (Peroni, 2011 and Altaykan, 2004).

Histopathology of this report showed dermal histiocytic infiltrate, absence of signs of vasculitis and muciparous deposits, so it is compatible with the diagnosis of DGI. Immunohistochemistry showed CD68 positivity for histiocytes and epithelioid cells, in addition to a predominance of CD3 positive T lymphocytes. Findings compatible with the two cases reported in the literature by Lee et al 2007 and Szepetiuk et al 2012. Before beginning the treatment for DGI, it is necessary verify with the pacient is not using some medications that can trigger of the disease, such as: TNF inhibitors, ACE inhibitors and diuretics (Szepetiuk, 2012). If there is no report of use of previous medications, treatment should be directed to control the primary disease in order to control the DGI. There are reports of the use of several drugs, among them topical corticosteroids, (Peroni, 2011; Busquets-Perez, 2006 and Warycha, 2008) systemic corticosteroids, [7]non-steroidal anti-inflammatory drugs (Busquets-Perez, 2003) and other specific drugs for each related primary disease. In this report, the use of medications that trigger the disease was discarded and the proposed therapy was systemic corticosteroids, orally, with good therapeutic response.

Conclusions

Our case suggests that Interstitial granulomatous dermatitis (DGI) should be considered as an important differential diagnosis of palisade neutrophilic granulomatous dermatitis (DGNP) and also of skin rashes in patients with autoimmune diseases. Finally, further studies are needed to identify immunological mechanisms related to the pathogenesis of the disease.

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