SBR 2019
36º CONGRESSO BRASILEIRO DE REUMATOLOGIA
CENTRO DE EVENTOS DO CEARÁ
04 A 07 DE SETEMBRO
FOR TALEZA 2019
PROMOÇÃO
70 anos Sociedade Brasileira de Reumatologia
REALIZAÇÃO
SCR SOCIEDADE CEARENSE DE REUMATOLOGIA

PYODERMA GANGRENOSUM-LIKE ULCERATIONS IN GRANULOMATOSIS WITH POLYANGIITIS: A CASE REPORT

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BACKGROUND

Granulomatosis with polyangiitis (GPA) is a necrotizing vasculitis that affects small e medium vessels characterized by granulomatous inflammation involving upper and lower respiratory tract. It belongs to the group of vasculitis associated with ANCA, usually associated to the cytoplasmic pattern (c-ANCA), which is directed against proteinase 3 (PR3).[1]Skin and mucosal occurs in 15-50% of the cases, and in 13% of the cases might be the initial signs. The cutaneous lesions might be of the polymorphic type, including palpable purpura, blisters, vesicles, papulo-nodular and livedo-reticularis. In addition, ulcerative lesions with elevated erythematous-violet borders mimicking pyoderma gangrenosum (PG) are reported as the rarest cutaneous presentation of GPA, representing approximately 1% of the cases, not only affecting lower limbs, but also should it affect face or other sites more rarely.[1]

CASE REPORT

A male patient, 45 years old, born in and coming from São Paulo, Brazil. He had a history of repeated upper airway infections from childhood, which was treated with antibiotics, and pulmonary tuberculosis treated in 2007. By the time of 2017, beginning a painful erythematous nodular cutaneous lesion with purulent and ulcerative secretion with erythematous-violaceous border, affecting trunk and zygomatic bilaterally, characterizing of PG. He also presented fever, night sweats and involuntary weight loss of 8 kg, as well as reduction of visual acuity. It evolved with paralysis of vocal cords and pulmonary nodulations. A biopsy of pulmonary lesions was performed and it revealed a chronic, circumscribed inflammatory process with a granulomatous pattern affecting small blood vessels, characterized by a plasmacytic infiltrate with giant cell reaction and areas of necrosis with exudate, adjacent parenchyma with extensive areas of alveolar hemorrhage, compatible with GPA. The ANCA, indirect immunofluorescence, was negative. The magnetic resonance imaging of the sinuses revealed bone erosions of the nasosinusal soft tissues. After GPA diagnosis, he underwent pulse therapy with corticosteroids and cyclophosphamide and progressed with clinical improvement and healing of cutaneous lesions.[1]

CONCLUSION

We describe a case of GPA associated with cutaneous ulcerations with PG-like features. It can be differentiated from the histology, which presents a neutrophilic inflammatory infiltrate mainly affecting lower limbs and rarely face and trunk. Suggests GPA the presence of positive c-ANCA and systemic involvement, especially with pulmonary involvement. Summarizing, the diagnosis of GPA is quite difficult, skin lesions might help in early diagnosis, implying an improvement in morbidity associated with pathology.[1]