

Sweet's Syndrome associated with Hodgkin's Disease - Case report *

Síndrome de Sweet associada a linfoma de Hodgkin – Relato de caso

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Abstract: Sweet's syndrome is a rare cutaneous disease of unknown etiology. About 20% of the cases are associated with hematological neoplasms, and cases related with Hodgkin's disease are rare. We present the case of a 57-year old male patient who developed the syndrome concomitantly with the neoplasm. The diseases were controlled with specific treatment.

Keywords: Hematologic neoplasms; Hodgkin disease; Sweet syndrome

Resumo: A síndrome de Sweet é enfermidade cutânea rara e de etiologia pouco esclarecida. Cerca de 20% dos casos são associados a neoplasias hematológicas, sendo raros os casos relacionados à doença de Hodgkin. Relata-se caso de paciente masculino de 57 anos que desenvolveu a síndrome concomitantemente à neoplasia. As doenças foram controladas com o tratamento específico.

Palavras-chave: Doença de Hodgkin; Neoplasias hematológicas; Síndrome de Sweet

INTRODUCTION

Hodgkin's disease is a neoplasia of lymphoid origin characterized by proliferation of cells of variable morphology, usually multinucleate B lymphocytes called Reed-Sternberg cells.¹ This type of lymphoma was described by Thomas Hodgkin in 1832 and by Samuel Wilks in 1856. It most frequently affects male patients in the third and sixth decades of life.¹ Sweet's syndrome was described by Robert Douglas Sweet in 1964 and is defined by the onset of erythematous papules and nodules commonly on the face, neck and upper limbs, associated with fever and neutrophilic leukocytosis. It responds quickly and effectively to the use of systemic corticosteroids.² The syndrome may be associated with neoplasias in 20% of the cases, the most common of which is acute myeloid leukemia.³ There is also association with

myeloproliferative disorders, lymphoproliferative disorders, carcinomas and myelodysplastic syndromes. There are few case reports describing association between Hodgkin lymphoma and Sweet's syndrome in the literature. We present the case of a patient in whom the onset of cutaneous lesions was concomitant with neoplasia diagnosis.

CASE REPORT

A 57-year old male patient referred the onset of erythematous lesions on hands, feet and trunk for two months, after presenting clinical features of influenza. Simultaneously, there were oral aphthous ulcers (canker sores), fever (not measured) and progressive 14kg weight loss. The physical examination revealed 0.3 to 1.5 cm erythematous plaques, some of them

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with pseudovesiculation on the back, arms, hands, legs and feet, as well as oral ulcers (Figure 1). There was also a palpable supraclavicular lymph node, measuring around 2cm in diameter, on the left side. Laboratory exams revealed a erythrocyte sedimentation rate (ESR) of 46 mm/h and absence of leukocytosis or neutrophilia.

The clinical findings led to suspicion of Sweet's syndrome and a biopsy of papulous lesion on the back was performed. The histopathological exam showed atrophied epidermis, with straightening of the dermal-epidermal junction, papillary dermis edema and diffuse inflammatory infiltrate in the reticular dermis composed of neutrophils and some lymphocytes (Figure 2). Therapy was begun with prednisone 0.5 mg/kg/day, with initial lesion involution and posterior recurrence when the dose was decreased. As there was supraclavicular lymph node enlargement a computerized thorax scan was requested, showing mediastinal lymph node enlargement. The abdomen tomography showed celiac chain and right para-aortic lymph node enlargement. The histopathological exam of the supraclavicular lymph node provided the diagnosis of classical Hodgkin Lymphoma, with immunohistochemical pattern positive for CD15 and CD30 (Figures 3 and 4). The hematology service classified it as stage 3B Hodgkin Lymphoma, beginning the ABVD protocol (adriamycin, vinblastine, dacarbazine and bleomycin). Eight medication cycles were completed, with control of neoplastic lesions and Sweet's syndrome until the moment.

DISCUSSION

Sweet's syndrome is an acute febrile neu-



FIGURE 1: Erythematous papules and plaques on the trunk and arms

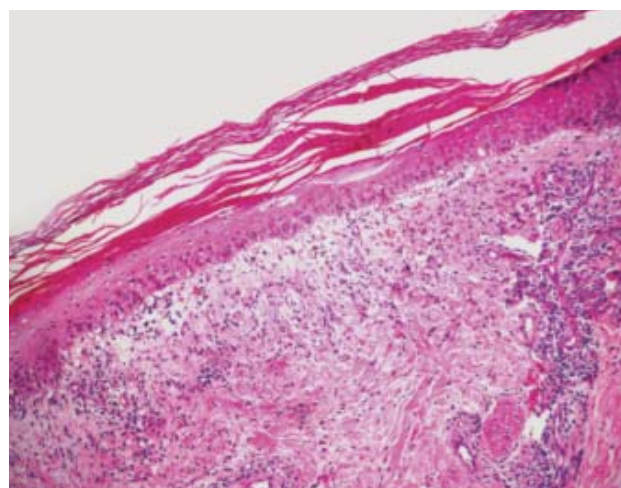


FIGURE 2: Papillary dermal edema with neutrophilic and lymphocytic infiltrate (HE -100x)

trophilic dermatosis, characterized by fever, neutrophilia, erythematous and painful cutaneous lesions, a dense infiltrate of mature neutrophils mainly in the upper dermis and a dramatic response to systemic corticosteroid therapy. The dermatosis is characterized by the onset of erythematous papules and nodules with a pseudovesiculation aspect, asymmetrical distribution mainly on the face, neck and upper limbs, possibly induced by trauma that usually resolves without leaving scars. Extracutaneous manifestations are less common.^{1,2}

Sweet's syndrome has three clinical forms: classical, drug induced and associated with malignancies.³ The form associated with malignancy occurs in approximately 20% of patients and is related to hema-

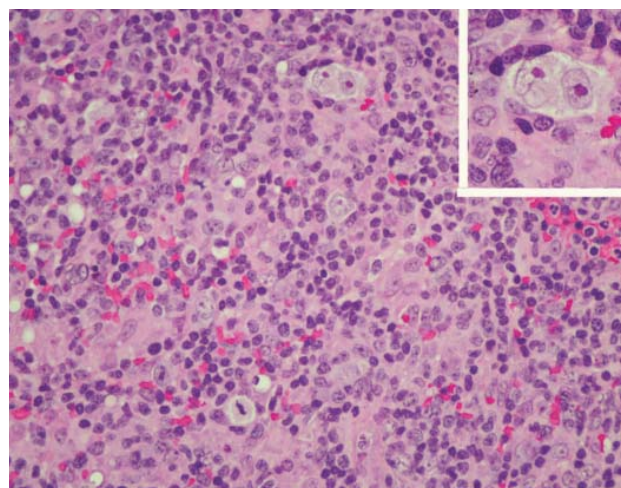


FIGURE 3: Histopathological exam of lymphoid tissue stained by H-E showing diffuse lymphocytic infiltrate, some pleomorphic plasmacytes and red blood cells (increased 400X). On the right, detail of binucleate Reed-Sternberg cell

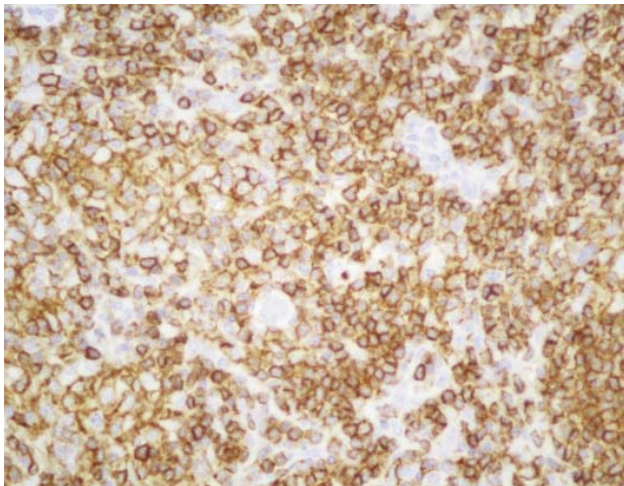


FIGURE 4: Photomicrograph showing immunohistochemical pattern positive for CD30 antigen (increased 400x)

tologic neoplasias (mainly acute myeloid leukemia) and solid tumors (breast, genitourinary tract and gastrointestinal tract). It affects men and women equally and is less frequently preceded by upper respiratory tract infection.^{3,5} When there is recurrence of cutaneous lesions, this may represent neoplasia recurrence as well. They are more diffusely distributed,

affect lower limbs more frequently and may have blisters, ulcerations and/or mime pyoderma gangrenosum lesions.^{6,7} The oral mucosa is more often associated with blood disorder. Anemia, neutropenia or absence of neutrophilia and an abnormal platelet count may occur. Myelodysplastic diseases such as polycythemia vera have already been reported as associated with the syndrome.⁸ Although uncommon, Sweet's syndrome usually precedes hematologic neoplasia.⁹ Cutaneous lesions respond to the use of systemic corticosteroids but there is more recurrence when the dose is decreased and the dermatosis may heal with successful treatment of the neoplasia.

The association of Hodgkin's disease with Sweet's syndrome is rare and very few cases have been published so far.^{10,11} Our patient presented fever, a characteristic cutaneous rash preceded by infection of the upper respiratory tract, compatible histopathological findings, ESR > 20mm/h and excellent response to corticosteroid therapy, fulfilling the diagnostic criteria for Sweet's syndrome. As a particularity of the case, there were oral canker sores, recurrence of cutaneous and mucosa lesions with reduction of the corticosteroid dose, lesions on lower limbs, anemia and absence of neutrophilia, which have been reported as evidence of subjacent malignancy.¹² □

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