Case Report

Penile ulcer as a specific clinical manifestation of Waldenstrom's macroglobulinemia*

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Abstract: Waldenstrom's macroglobulinemia is considered a lymphoma by the World Health Organization. Cutaneous lesions, particularly of a specific type, are rare occurring in 5% of patients. What draws attention in this case is the unusual cutaneous clinical manifestation and its location on the genitals, which has not been described in researched literature, therefore imposing differential diagnosis with other etiologies of genital ulcers.
Keywords: Hematologic neoplasms; Penile diseases; Skin ulcer; Waldenstrom macroglobulinemia

INTRODUCTION

Waldenstrom's macroglobulinemia (MW) is considered a lymphoma by the World Health Organization.[1] It is a malignant monoclonal gammopathy characterized by the presence of bone marrow lymphocytic infiltration and increased blood viscosity due to the high level of immunoglobulin M (Ig M). Clinical manifestation may be a consequence of blood hyperviscosity, Ig M deposition in tissues, peripheral neuropathy and primary amyloidosis. Sometimes clinical manifestations may be lymphoma specific, including tissue infiltration by neoplastic cells, but this is not very common. Several organs can be affected, such as liver, lungs, gastrointestinal tract, kidneys, skin, eyes and central nervous system. Cutaneous lesions, particularly of a specific type, are rare, occurring in 5% of patients. Here is reported a case of MW diagnosed following an unusual cutaneous manifestation. [2]

CASE REPORT

Patient, 52 years old, male, single, born in Cariri – AL, from Ameliópolis – SP, previously diabetic in use of oral hypoglycemic pills complained of penile ulcer for one month associated with unmeasured fever, asthenia and weight loss of approximately 5 kg in 4 months. On dermatological examination an extensive circular ulcer was found, 4 cm in diameter, lo-

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cated on base of penis. Its background was clean, but bleeding, and had verrucous papules around it suggesting viral warts (Figures 1 and 2). Some inguinal and axillary lymph nodes were enlarged. Laboratory investigation showed normocytic and normochromic anemia, thrombocytopenia, increased total proteins (15,99 g/dl), protein electrophoresis with monoclonal peak and IgM of 12900. The abdominal ultrasonography revealed the presence of hepatosplenomegaly. Histopathological exam showed lymphoid neoplasia with infiltrative growth composed of atypical lymphoid cells, small and drabs, with oval nuclei and nucleoli not very evident. These cells were arranged diffusely, sometimes with a tendency to follicle formation affecting the cutaneous annexa even the epidermis (Figure 3). Mitosis were often seen. Immunohistochemical reaction results with different markers were: CD5 (Leu1) positive focal; CD10/CALLA Ab-2 (56C6) positive focal; CD20 (L26) negative; CD43 (DF-T1) positive; CD45RO T-CELL (UCHL1) positive; LEUCOCYTE COMMON ANTIGEN (LCA) positive. These results are consistent with lymphoma and correlation with clinical data allows diagnosis of MW. Patient progressed rapidly to death even undergoing plasmapheresis and chemotherapy instituted by the oncology clinic.

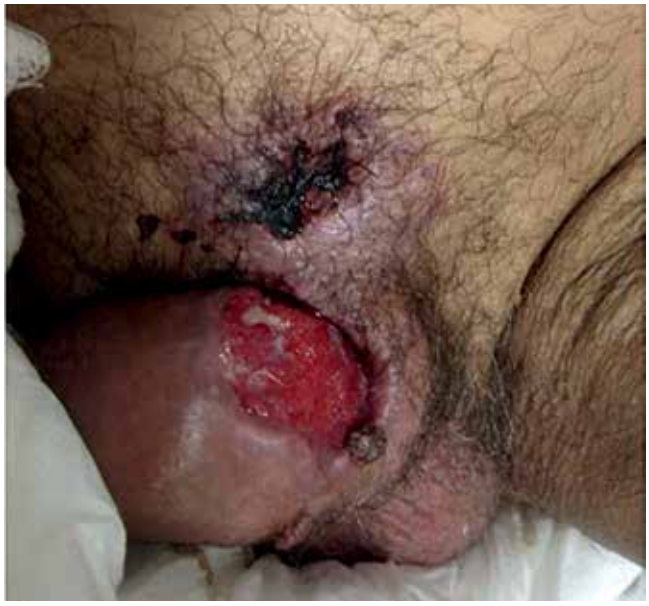

Figure 1: Lesion measuring 4 centimeters on penis base, with a clear background

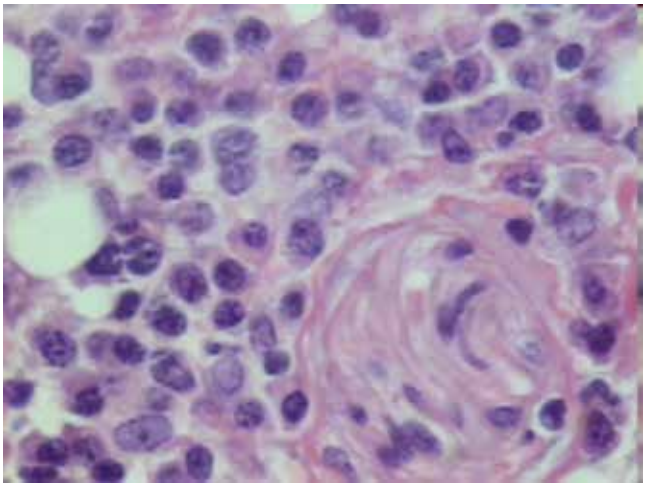

Figure 2: Anatomopathological examination of ulcer border stained with hematoxilin and eosin

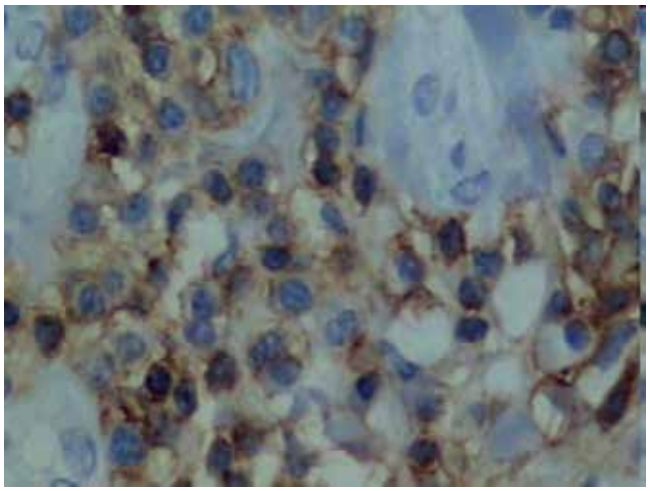

Figure 3: Immune histochemical examination of positive ulcer border

DISCUSSION

MW is a rare condition representing 2% of hematologic malignancies; the incidence rate is higher among white people, aged around 65 years, with slight predominance in males and with an average survival of 5 years. Etiology is unclear but familial case reports suggest genetic predisposition. A study of 181 patients showed 20% of them had a first-degree relative with MW of B cells or other lymphoproliferative disease. [3,4] MW has an insidious and nonspecific beginning, is usually asymptomatic and accidentally diagnosed by routine laboratory exams. Merlini et al (2003)[3] demonstrated, in 215 patients with MW disease, weakness (66%), anorexia (25%), peripheral neuropathy (24%), weight loss (17%), fever (15%), hepatomegaly (20%), splenomegaly (19%), lymphadenopathy (15%), purpura (9%) and hemorrhagic manifestations (7%). Most of these signs and symptoms were also observed in the patient here reported. Libow et al (2001) classified MW skin lesions into two categories: neoplastic (MW cutaneous) and non-neoplastic. [5] Non-neoplastic may be associated with blood hyperviscosity syndrome, cryoglobulinemia, deposit of IgM in the skin and

others factors. MW cutaneous neoplastic lesions are rare, usually occur in advanced stages of the disease and present as brownish-erythematous and infiltrated plaques. What draws attention in this case is the unusual cutaneous clinical manifestation and its location on the genitals, which has not been described in researched literature, therefore imposing differential diagnosis with other etiologies of genital ulcers. As examples we may cite oncology, especially squamous cell carcinoma, discarded by histopathological exam; infectious, as chancroid, the chancre, herpes simplex, granuloma inguinale, mycobacteria and HIV, excluded by Gram stain and culture of the secretion of injury, specific serology and histopathology; and inflammatory, such as pyoderma grangrenosum, Behçet disease and Crohn's disease, which, besides not meeting clinical criteria, had histopathological examination results also not compatible with those entities.❑

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