Manifestación cutánea de una enfermedad sistémica

Skin manifestation of a systemic disease

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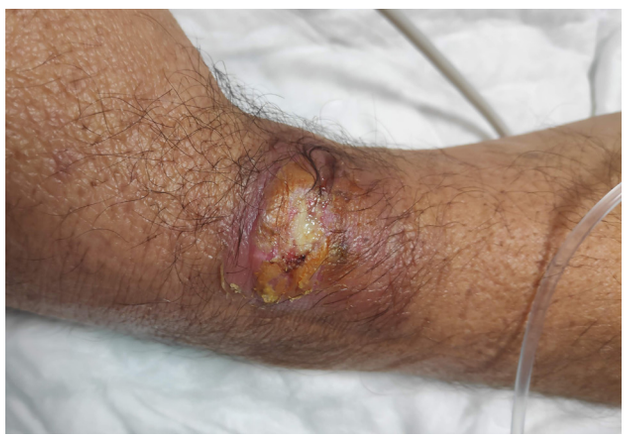
CLINICAL CASE

We present a case of a 51-year-old man who presented with a lesion on the back of his left hand, accompanied by ongoing suppurative exudate for one week. After receiving amoxicillin/clavulanic acid treatment without improvement, he visited the emergency room. At this point, he exhibited symptoms such as anorexia, asthenia, a feverish feeling, and a new ulcerated lesion on the nasal pyramid, along with unintentional weight loss.

Laboratory analysis revealed severe pancytopenia (hemoglobin 5.5 g/dL, leukocytes 670/uL, platelets 20,000/uL) and a reduced reticulocyte count, with elevated CRP and sedimentation velocity. Blood smear exhibited anisocytosis. Tests for HIV, hepatitis, VDRL, CMV, EBV, Leptospira, Parvovirus, Rickettsia, Borrelia, Brucella, and Coxiella infections were negative, including blood cultures and myeloculture. Abdominal CT scan indicated hepatosplenomegaly at the upper limit of normal without other abnormalities.

The patient initiated antibiotic therapy and received a blood transfusion, which yielded lower-than-expected results. Subsequently, a myelogram with bone biopsy revealed bone marrow infiltration by 80% hairy cell leukocytes, with immunophenotyping consistent with HCL, and positivity for BRAF V600E mutation.

He was referred to oncology and initiated vemurafenib because during hospitalization tested positive for SARS-CoV-2 with good tolerance. Currently, he is undergoing treatment with noticeable improvement in cutaneous lesions.



Although skin changes in pancytopenic patients are uncommon, they should prompt consideration of hematological neoplasms.

The new guidelines for this disease recommend that patients should be in a watch-and-wait strategy, with treatment initiated only if the patient develops symptoms, as was the case with our patient. Purine analogs therapy (using either cladribine or pentostatin) remains the standard treatment. Additionally, recent evidence suggests that adding rituximab enhances therapeutic activity and may improve the duration of complete response. In special cases, such SARS-Cov2 infections, alternative options should be considered[1]. Although the disease follows a remitting and relapsing pattern of response to sequential treatments[2], early diagnosis and treatment initiation help to prevent complications, particularly infections.

DISCUSSION

Hairy cell leukemia is a rare indolent lymphoproliferative neoplasm of mature B cells affecting the bone marrow and spleen[1-5]. The peripheral blood involvement is less common. A defining feature of this hematolymphoid tumor is the presence of medium-sized mature lymphocytes characterized by 'hairy' projections, typically found in the bone marrow and spleen[1,3,4]. BRAF V600E are useful in distinguishing HCL from diseases that mimic this condition, which is very relevant because the mimic diseases tend to have a poorer prognosis and typically show limited response to purine analogues, which are usually the standard treatment for HCL.[2]

This condition predominantly affects middle-aged males and is four to five times more frequent in men than women[1,2,5]. The majority of patients are asymptomatic at diagnosis and the disease is usually found incidentally during routine blood cell counts[5]. Approximately 10% of patients experiencing skin lesions[4,5]. Skin symptoms may be specific to HCL or due to autoimmune reactions, infections and secondary neoplastic or drug-induced reasons.[5]

CONFLICT OF INTEREST
The authors declare that they have no conflict of interests.

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This research had no funding sources.

ETHICAL ASPECTS
All participants submitted a consent form to be included in this study.

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