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CASE FOR DIAGNOSIS

Widespread Nodules

Nódulos generalizados

Medical History

A 75-year-old man with a past history of systemic hypertension consulted for asymptomatic lesions that had developed on the face and trunk 1 month earlier. He also reported weight loss of 10 kg in the past year.

Physical Examination

Multiple erythematous nodules 2 to 4 cm in diameter and of firm consistency were observed on the face (leonine facies), scalp, neck, trunk, and proximal regions of the limbs (Figure 1). There was no mucosal involvement. Small, mobile inguinal lymph nodes were palpable, but there was no evidence of hepatosplenomegaly.

Histopathology

There were no notable changes in the epidermis. Except for an unaffected band of papillary dermis, a diffuse cellular infiltrate was observed around the dermal blood vessels and adnexa and penetrating between the collagen fibers (Figure 2). The cells were large with basophilic cytoplasm, containing a vesicular nucleus that was occasionally kidney-shaped, and with a small nucleolus. Frequent mitotic figures were observed (Figure 3). Immunohistochemically, the infiltrate was intensely positive for CD68 and lysozyme, with foci that were positive for myeloperoxidase, leukocyte common antigen, and CD34, and was negative for CD117 and factor VIII.

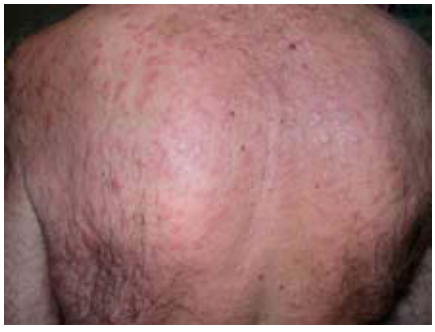

Figure 1

Additional Tests

The complete blood count revealed leukocytosis (39 600 leukocytes/μL; 56.2% neutrophils, 23.1% lymphocytes, 17.9% monocytes, 1% eosinophils, and 1.9% large unstained cells) and a low platelet count (89 000 platelets/μL). The peripheral blood smear showed hypersegmented neutrophils. Bone marrow aspiration showed massive infiltration by cells of monocytic origin, 20% to 30% monoblasts, and 65% promonocytes-monocytes. The study using fluorescent in situ hybridization revealed trisomy 8 in 70% of the cells.

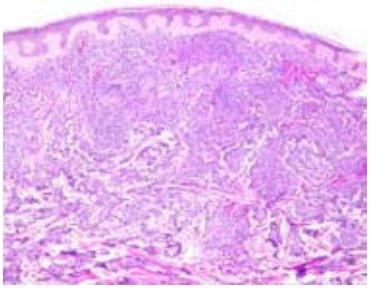

Figure 2 Hematoxylin-eosin, original magnification ×100.

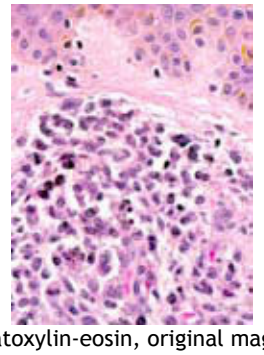

Figure 3 Hematoxylin-eosin, original magnification ×400.

What Is Your Diagnosis?

Diagnosis

Leukemia cutis in a patient with acute monocytic leukemia (M5b in the French-American-British [FAB] classification).

Clinical Course and Treatment

The first month of chemotherapy with etoposide at a dose of 50 mg/d led to partial improvement of the facial nodules.

Comment

Leukemia generally causes nonspecific reactive cutaneous lesions with no detectable tumor cells. Less than 10% of patients present lesions due to leukemic infiltration of the skin, known as leukemia cutis, and such infiltration indicates an advanced stage, with an average survival of less than 1 year. Appearance follows or occurs at the same time as hematologic diagnosis; when there is no alteration in the peripheral blood or bone marrow, the condition is reported as aleukemic leukemia cutis.[1] Treatment of leukemia cutis is the same as for the underlying leukemia, although it can be combined with radiotherapy for the skin lesions.[2]

Acute monocytic leukemia is an acute myeloid leukemia subtype, M5 in the FAB classification. This is the type of leukemia that most commonly develops leukemia cutis (10%-33%). It is subdivided into M5a and M5b according to whether more or less than 80% of the elements in the monocytic series are monoblasts.[3] Certain chromosomal abnormalities, such as trisomy 8, are common in patients with leukemia cutis.[4]

Skin manifestations of leukemia cutis are varied. The appearance of asymptomatic, firm, erythematous-violaceous papules or nodules in less than 1 month is a characteristic finding. The head (leprous appearance),[5] neck, trunk, and legs are usually affected, and areas subject to trauma and scars (due to burns or herpes virus infection) can become infiltrated. Gingival hyperplasia is not uncommon.[2-4,6]

The histologic pattern of leukemia cutis depends on the type of leukemia involved. In acute monocytic leukemia, there is a diffuse tumor cell infiltrate that invades the full thickness of the dermis and occasionally the hypodermis, with a tendency to distribute itself around blood vessels and the adnexa. It is usually denser in the upper dermis, penetrates between the collagen fibers, and destroys deep blood vessels and adnexa. The epidermis and papillary dermis are generally unaffected. The infiltrate is quite monomorphic, and is composed of large cells with basophilic cytoplasm and an oval vesicular nucleus; numerous mitoses are also observed.[2,4]

Given the high degree of clinical variability, the differential diagnosis is extensive. There are atypical presentations that simulate dermatoses that can develop in the context of leukemia, such as drug-related skin reactions, opportunistic infections (especially fungal or viral), erythema elevatum diutinum, Sweet syndrome, acute leukocytoclastic vasculitis, graft-versus-host disease, erythema nodosum, and pyoderma gangrenosum. Histopathologic confirmation is required to establish the diagnosis of leukemia cutis. Histopathologically, it can be confused with lymphoma, extramedullary hematopoiesis, anaplastic carcinoma, Ewing sarcoma, neuroblastoma, Merkel cell carcinoma, skin metastases, melanoma, and histiocytic proliferation, and therefore requires immunohistochemical analyses.[4,7]

Conflict of Interest

The authors declare that they have no conflicts of interest.

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