described in Table 1 ranged from 2 to 16 months. Diagnosis was delayed due to the atypical presentation, and was usually established only after several medical visits. Treatment in most cases was with intramuscular pentavalent antimoni- als, given the tendency towards chronicity and the risk of deformity at the site of the lesion, although intralesional administration can also bring about complete resolution of the lesion.

The differential diagnosis of ulcerative lesions of the ear is broad, and includes acute lesions (eg, infections, Winkler disease, bites, sarcoidosis, granulomas, and neoplasms) and chronic lesions (eg, lupus vulgaris, discoid lupus, leprosy, and lymphoma). Clinical history is very important for diagnosis, which should be confirmed by microbiological studies and molecular biology techniques.

Old World CL usually heals spontaneously within 3 to 18 months, leaving a scar. Treatment is indicated in cases that involve multiple or large lesions, persist for more than 6 months (chronic CL), or affect regions at risk of functional or aesthetic compromise, such as the ear lobe. Localized forms are treated using local treatments such as intralesional MA (Case 2), the efficacy of which can be very high, albeit variable. The treatment of choice for chronic forms is parenteral pentavalent antimonials, which can be complemented with local treatments such as cryotherapy, as described in Case 1.

Rapid recognition of this disease can permit earlier treatment, decreasing the likelihood of the development of residual lesions.

References


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Erythroderma as an Initial Presentation of Langerhans Cell Histiocytosis Involving the Sinus

Eritrodermia como forma de presentación inicial de histiocitosis sinusal de células de Langerhans

To the Editor:

We report the case of a 56-year-old man with a history of chronic obstructive pulmonary disease and hypertension who was being treated with eprosartan/hydrochlorothiazide and atorvastatin/amiodipine. He presented with an eczematous, pruritic skin condition that had developed 2 months previously.

Physical examination revealed coalescing erythematous plaques on the upper limbs, face, and the upper third of the trunk, affecting approximately 25% of the body surface. There was no palmarplantar or mucosal involvement (Fig. 1). A skin biopsy of the affected area showed a superficial perivascular dermatitis with eosinophils and intraepidermal necrotic keratinocytes, suggesting drug-induced skin disease. Blood tests revealed leukocytosis (15330 × 103/L), with a predominance of neutrophils (82.2%), and increases in the erythrocyte sedimentation rate and in the levels of C-reactive protein and immunoglobulin E (> 5000 IU/mL).

After diagnosis of drug-induced skin disease antihyperensive treatment was discontinued and a tapering dosage of prednisone was prescribed, starting at 1 mg/kg/d. After 40 days of treatment the dose of corticosteroids was reduced to 10 mg/d due to the development of cushingoid habitus. Given the progression of the skin lesions to erythroderma, affecting almost 90% of the body surface, the patient was treated with acitretin (25 mg/d),

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which stabilized the symptoms and reduced the intensity of the erythroderma and pruritus. The patient’s baseline hypertension was controlled by dietary measures after a relapse induced by temporary treatment with enalapril.

Laboratory workups and monthly imaging studies revealed no differences as compared with the initial findings. Eight months later eosinophilia was detected (20.8%; 2310 × 10⁹/L). A new skin biopsy revealed no additional findings and bone marrow aspiration was normal.

Weeks later the patient noticed an enlarged right inguinal lymph node of 2 cm in diameter. A computed tomography scan of the chest and abdomen revealed considerable enlargement of the mediastinal lymph nodes (Fig. 2A) and bilateral axillary and inguinal lymph nodes of over 1 cm in diameter (Fig. 2B). No hepatosplenomegaly, mesenteric lymphadenopathy, or bone alterations were observed. Histological examination of the inguinal node revealed that the paracortex contained cellular aggregates consisting of large cells with grooved nuclei containing compact chromatin, together with a large, eosinophilic cytoplasm. Microabscesses and occasional histiocytes and plasma cells were also observed. The cells were positive for S-100/CD1a and negative for myeloperoxidase and CD117, and had a low proliferative index. A histological diagnosis of Langerhans cell histiocytosis (LCH) involving the sinus was established (Fig. 3 A-D). Based on this diagnosis, acitretin treatment was discontinued and the patient began treatment with tapering dosages of vinblastine (intravenous bolus, starting at 6 mg/m²) and oral prednisone (starting at 40 mg/m²/d) for 6 and 4 weeks, respectively. This resulted in an improvement in the patient’s baseline condition. The patient remains asymptomatic after 3 months of follow-up.

Erythroderma is defined as inflammation that affects at least 90% of the body surface. Its etiologic diagnosis is difficult. The dermatologist must be alert to the possibility of an underlying neoplasm in cases of insidious onset, treatment resistance, or weight loss, and in cases with no prior history of skin disease. Histology is nonspecific, and despite the presence of signs specific to the primary disease, etiologic diagnosis is often not achieved.

LCH is a relatively uncommon entity characterized by oligoclonal proliferation of Langerhans cells. Its incidence is unknown and it is difficult to diagnose. Cutaneous presentations of LCH resembling pyoderma gangrenosum, annular granuloma, amebiasis, eruptive xanthomas, and flat warts, among other conditions, have been described. LCH can affect one or more organs; the number of affected organs has implications for prognosis and treatment. This condition has been reported in association with solid tumors, lymphomas, and leukemias, and a common origin of LCH and hematological malignancies has been widely debated.

Several therapeutic strategies are used in the management of LCH in adult patients, depending on the number of organs involved and the clinical course. In our case, even though only lymphatic involvement was initially observed, we opted for aggressive therapy, which significantly reduces morbidity and mortality according to the Evaluation and Treatment Guidelines of the Histiocyte Society.

Erythroderma as an initial presentation of LCH, as occurred in the present case, has been described in only one series to date. The main difficulty in the present case was the length of time between the onset of erythroderma and lymphadenopathy; we established a definitive histological diagnosis of the latter through histological analyses, as all findings hitherto were nonspecific. We wish to stress the importance of repeated complementary testing and close follow-up with retesting in patients with erythrodermas with an atypical clinical course and poor therapeutic response.
Figure 3  A, Lymph nodes affected by Langerhans cell histiocytosis (hematoxylin-eosin, original magnification ×2). B, Subcapsular sinus occupied and expanded by a predominantly mononuclear cell population containing some eosinophils (hematoxylin-eosin, original magnification ×20). C and D, Higher magnification images showing the eosinophils (arrowheads in C; hematoxylin-eosin, original magnification ×40) and the grooves or folds in the elongated nuclei of the CD1a+ Langerhans cells (D; CD1a, original magnification ×10).

References


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