“Refractory Intertrigo” in an Elderly Woman

Sir,

Langerhans cell histiocytosis (LCH) is a rare neoplastic disorder characterized by clonal proliferation and infiltration of CD 1a and CD 207 (Langerin) positive dendritic cells in various organs.

An 80-year-old presented with a non-healing, itchy rash in both groins. Lesions had been present for 2 years, having been previously been treated by several physicians with various topical anti-fungal and steroidal agents as intertrigo, without any relief. Her general health condition was satisfactory and she denied having any comorbidities like diabetes or hypertension. Cutaneous examination revealed erythematous, fissured and scaly plaques with few satellite lesions in the both inguinal areas [Figure 1]. No other body site was involved. No lymphadenopathy and hepatosplenomegaly were found on systemic examination.

A skin biopsy taken from the plaque revealed dense infiltrates of mononuclear cells with pale, kidney shaped nuclei infiltrating the papillary dermis and epidermis [Figure 2a and b]. The infiltrate also included lymphocytes, neutrophils, and few eosinophils which partly obscured the neoplastic cell population [Figure 2c and d]. A panel of immunohistochemistry markers including CD1a, S100, CD3, CD20, CD30, EMA, CD68, and Langerin showed clusters of CD1a and Langerin positive cells in the papillary dermis and epidermis [Figure 2e and f]. A repeat biopsy from the lesion showed similar features along with additional superinfection by bacteria and candida. A diagnosis of cutaneous Langerhans cell histiocytosis was made based on the typical cytomorphology and immunophenotype of the neoplastic cell population. On further inquiry, the patient complained of increased thirst for last few months (drinking 6 liters of water per day). Routine hematological tests, ultrasonography of lymph nodes, CT scan of thorax and abdomen were normal, but cranial MRI demonstrated infundibulo-hypophysis with damage to neuro-hypophysis compatible with an infiltration by langerhans cell histiocytes. Fluid deprivation test showed no increase of urine osmolarity while serum osmolarity remained slightly elevated confirming the diagnosis of diabetes insipidus (DI). Bone scan revealed pathological enhancement in distal femoral and left tibial areas indicating bone marrow involvement of LCH. There were no respiratory symptoms from the start and spirometry was normal. The patient was started on desmopressin 0.1 mg/ml HS nasal spray with radiation therapy for bone lesions and topical steroids for skin lesions with good response at 8 months of regular follow-up.

The definitive diagnosis of LCH is based on histological and immunophenotypical evaluation of lesional tissue. Presence of characteristic pleomorphic histiocytic cells which are positive for CD1a and Langerin stain confirms the diagnosis. Since the expression of Langerin confirms the presence of Birbeck granules, the demonstration of these granules by electron microscopy is no longer required for a definitive diagnosis. CD1a may also be positive in indeterminate cells and S100 may show positive staining in melanocytes, nerve sheath cells, myoepithelial cells, adipocytes, and chondrocytes. Thus, these markers are not specific for diagnosis of LCH. Further classification of the disease is based on involvement of high risk organs, namely, hematopoetic system, liver, spleen, and lungs.

After a diagnosis of LCH is made, further investigations with regard to internal organ involvement are guided by the criteria laid down by the histiocyte

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society in April 2009. The skin lesions in LCH typically include papules, pustules and/or vesicles with yellowish crusts and erythema which classically occur in intertriginous areas, as in our patient, or on the scalp. This manifestation may be mistaken for Candidial intertrigo or dermatophytosis with a consequent delay in diagnosis and treatment. In our patient, a superinfection with candida was seen in PAS stained section, a potential pitfall especially when the clinical presentation is similar to candidial intertrigo.

Although LCH has been classically described in pediatric age group, a small percentage of patients are elderly, who may have cutaneous involvement for many years before the onset of systemic involvement. Diabetes insipidus (DI) is the most common endocrine manifestation of LCH. It results from involvement of the posterior pituitary gland and infundibulum by infiltrating Langerhans cells. MRI is the most sensitive tool for detection. In our patient, the skin biopsy finding of LCH prompted the clinician to probe into further systemic investigation leading to diagnosis of LCH with symptomatic DI in the form of polydipsia. These patients require lifelong supplementation of desmopressin acetate.

Treatment of cutaneous LCH includes topical and systemic steroids, oral isotretinoin, thalidomide, nitrogen mustard, radiation, excimer laser, resection, interferon-alpha, and chemotherapy. Systemic involvement is treated with systemic steroids, vincristine or vinblastine based chemotherapy or radiation therapy. With the help of this article, we seek to emphasize LCH as a possible diagnosis in any case of long standing and difficult to treat intertrigo. Skin involvement of LCH may be the first clinical sign of systemic disease and a skin biopsy with additional immunohistochemistry for langerin is mandatory to reach specific diagnosis.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

References
1. Aoki M, Aoki R, Akinomoto M, Hara K. Primary cutaneous langerhans cell histiocytosis in an adult. Am J Dermatopathol 1998;20:281-4.
2. Minkov M, Grois N, McClain K, Nanduri V, Rodriguez-Galindo C, et al. Histiocyte society evaluation and treatment guidelines. Histiocyte society [cited 2009 April]. Available from: https://histiocytesociety.org>document.doc?id=290.

3. Hu JC, Ra S, Gutierrez MA. Cutaneous langerhans cell histiocytosis in an elderly woman. Dermatol Online J 2010;16:6.

4. Stefanato CM, Andersen WK, Calonje E, Swain FA, Borghi S, et al. Langerhans cell histiocytosis in the elderly: A report of three cases. J Am Acad Dermatol 1998;39:375-8.

5. Goncalves CF, Morais MO, de Cassia Goncalves Alencar R, Bastista AC, Mendonca EF. Solitary langerhans cell histiocytosis in an adult: Case report and literature review. BMC Res Notes 2016;9:19.

6. Girschikofsky M, Arico M, Castillo D, Chu A, Doberauer C, et al. Management of adult patients with langerhans cell histiocytosis: Recommendations from an expert panel on behalf of euro-histio-net. Orphanet J Rare Dis 2013; 8:72.