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Case Report Open Access

Langerhans Cell Histiocytosis in Postmenopausal Female with Intractable Pruritus: A Rare Case Report

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Abstract

Langerhans cell histiocytosis (LCH), characterised by the infiltration of one or more organs by large mononuclear cells, may affect patients of any age but in adults the features of this disease is still poorly defined. Here, we report a case of an adult with crusted erosions in seborrheic distribution, Bilateral inguinal lymphadenopathy with ulceration, which on biopsy showed features of multisystem LCH, and was confirmed by immunohistochemistry. Despite chemotherapy and high dose antihistamines our patient complained of intractable pruritus which was relieved with Narrow Band UV-B phototherapy.

Introduction

Langerhans cell histiocytosisis a clonal proliferative disease of langerhans cells that accumulate in various tissues and cause damage, express an immunophenotype positive for S100 and CD1a, and contain cytoplasmic birbeck granules. This group of disorders can involve many organ systems but affects primarily the bone, skin, lymph nodes, lungs, liver, spleen, endocrine glands and nervous system. Langerhans cell histiocytosisis commonly seen in infants and early childhood. Adult onset is rare [1]. Skin involvement occurs in 50% of adults with multisystem langerhans cell histiocytosis, characterised by pink to skin-colored papules, pustules and/or vesicles having a seborrheic dermatitis- like picture.

Keywords: Adult langerhans cell histiocytosis, Intractable pruritus, Narrow band UVB

Case Report

50 year old postmenopausal female patient presented with multiple ill-defined erosions covered with loosely adherent crusts over an erythematous base admixed with haemorrhagic crusts in between present over chest, back, scalp and abdomen with severe pruritus present throughout the day for 9 months and bilateral inguinal lymphadenopathy with ulceration covered with exudate for 5 months (Figure 1,2). The routine laboratory investigations were all within normal limits. Biopsy and immunohistochemistry was done. Stained biopsy specimens for immunohistochemistry showed cytoplasmic and nuclear reactivity for protein S100 [figure 3], and also showed reactivity to CD1a (Figure 4) which is the definite and reliable marker of langerhans cell histiocytosis (CD-1a and S-100 positive)(Figure 3,4). Skeletal survey showed osteolytic lesion in the pelvis. These findings were confirmed on bone scan (Figure 6).

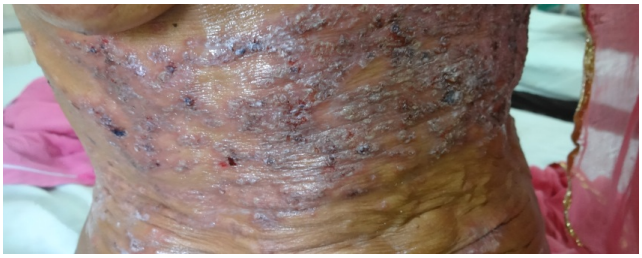

Figure 1: Crusted erosions over chest and abdomen.

MRI brain and CT chest was normal. CT abdomen and pelvis showed inguinal lymphadenopathy. The disease was staged as multisystem langerhans cell histiocytosisas there was involvement of bone, lymph node and skin.

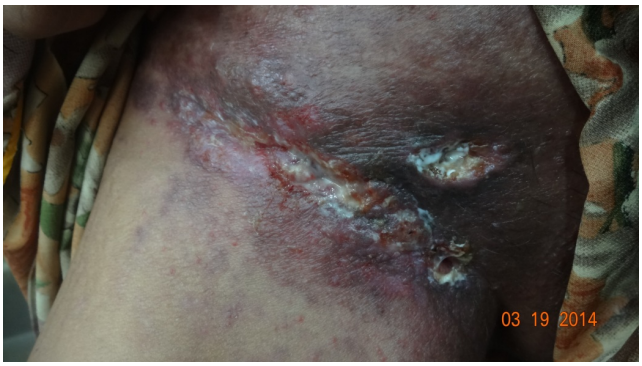

Figure 2: Inguinal lymhadenopathy with ulceration.

Patient was started on injection vinblastine 6 mg/m^2 weekly along with tablet prednisolone 40 mg once daily for 6 weeks. Following which patient was evaluated for response, there was significant

reduction in size of lymph nodes with healing of ulcer but residual skin lesions with severe pruritus still persisted.

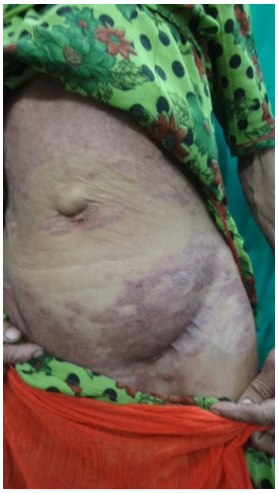

Figure 3: Ulcer healed after treatment.

Hence, it was decided to continue the same regimen for another 6 weeks. Pruritus did not subside with high doses of antihistamines, benzodiazepenes, selective serotonin reuptake inhibitors (paroxetine 25 mg) and tricyclic anti-depressants (doxepin 50 mg).

Therefore, Narrow Band UVB (NBUVB) was started with an initial dose of 280 mj/cm^2, thrice weekly with 20% increment at each sitting. After one week of therapy there was marked improvement in the pruritus which got completely relieved after 9-10 sessions. Maximum dose of 1200 mj/cm^2 was achieved and this dose was maintained for 3 weeks and then slowly tapered in next 5 weeks.

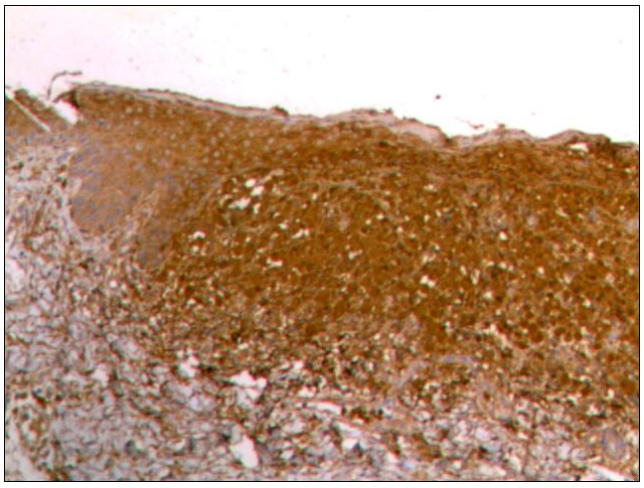

Figure 4: Immunohistochemical study, S100 positivity (10x).

After completion of 12 weeks of therapy patient was lost to follow up and she reported after 2 months with new skin lesions and pruritus. The patient was labelled refractory to treatment and put on palliative chemotherapy comprising of injection vinblastine 6 mg/m^2 , tablet prednisolone 40 mg daily for 5 days every 3 weeks, tablet 6 mercaptopurine 50 mg daily along with Narrow Band UVB(NBUVB) for which the same regime.

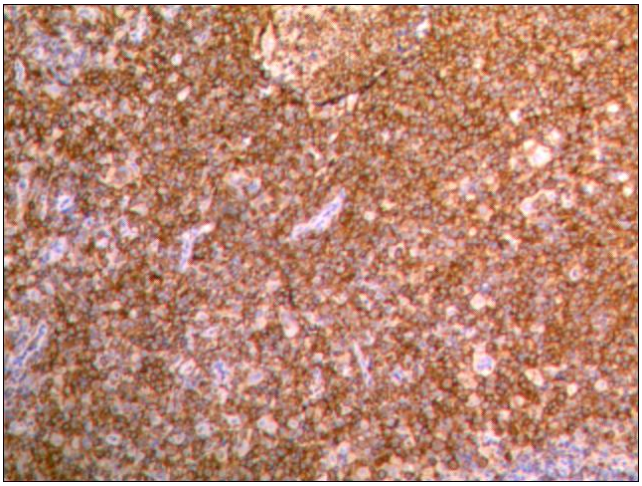

Figure 5: Immunohistochemical study, CD1a positivity (10x).

As before was followed. After completion of 3 months of palliative protocol, the disease was static and only a mild mucositis had developed. The patient is still taking treatment as per this protocol.

Figure 6: Showing Bone scan

Discussion

Histiocytes are derived from circulating monocytes and thus share a common bone marrow progenitor cell, the neutrophil/macrophage colony forming unit. Aberrant cytokine production by these cells results in their proliferation and contributes to the pathological sequelae of langerhans cell histiocytosis including fibrosis, bone resorption, fever, and necrosis.

Histiocytosis-X term was coined by Lichtenstein, in 1953, to describe a group of disorders (Hand-Schuller-Christian disease, Letterer-Siwe disease, and Eosinophilic Granuloma) characterized by infiltration of involved tissue with large number of abnormal histiocytes [1]. Subsequently, these histiocytes were found to be similar to langerhans cell normally present in the skin, and therefore termed as langerhans cell histiocytosis. It is a clonal neoplastic disorder [2], whose pathogenesis is unknown.

The Writing Group of Histiocytic Society (1987) has recently defined the criteria for diagnosis of langerhans cell histiocytosis, [3] based on clinical feature, histopathology and immunohistochemistry. Langerhans cell histiocytosis is a rare disease and generally affects children. Three to four cases per million occur annually in children under 15 years of age, with a male: female ratio of 2:1. In adults, the condition is even rarer, with an annual incidence of 1 in 560,000 [4]. In our case the patient was an adult postmenopausal female with langerhans cell histiocytosis which is rarely seen.

Skin lesions are common and are often the presenting manifestation of langerhans cell histiocytosis. Skin lesions present as translucent, scaly crusted papules, vesicles and pustules mimicking seborrheic dermatitis over scalp, trunk and intertriginous areas. In our case, patient presented with crusted erosions over chest, back, scalp and abdomen for 9 months and bilateral inguinal lymphadenopathy with ulceration for 5 months. Patient had severe intractable pruritus which is not a feature of langerhans cell histiocytosis as per literature.

Langerhans cell histiocytosis may involve any organ system, but the frequency and the extent of the disease is age dependent. According to several retrospective studies done on neonates and children under the age of 4, 51 to 71 percent of children with langerhans cell histiocytosis present with multiorgan disease. A recent retrospective study showed multisystem involvement in 68.9% of adults [5].

The Histiocyte Society has established guidelines to assist in the diagnosis and study of langerhans cell histiocytosis [6]. The initial evaluation consists of a complete physical examination, inclusive of height and weight measurements, in addition to laboratory studies including hematological assays, liver function tests, complete skeletal radiographic survey, chest radiography, CT or MRI of the brain with endocrine evaluation.

Treatment depends on the extent and severity of disease. Steroids have been used either topically for skin lesions or systemically for more invasive disease and chemotherapeutic agents are indicated for multisystem involvement, mainly the vinca alkaloids, especially vinblastine. The patient's response to chemotherapy during the 6 week induction phase is the single best prognostic indicator according to recent therapeutic trials [6]. The first 6 weeks of therapy have an 88-91 percent survival rate in patients who respond to chemotherapy, which drops to 17-34 percent in patients who fail to respond. It has been advocated that non-responders be identified early so that more aggressive therapy may be employed [6]. In our case, patient initially responded after 12 weeks of therapy, was lost to follow up for 2 months and reported back with recurrence. Therefore, the patient was put on palliative treatment comprising of vinblastine, 6 mercaptopurine and prednisolone. Narrow Band UVB (NBUVB) by its inhibitory action on NK cells and altering cytokine secretion at the site of inflammation has role in alleviating pruritus.

To the best of our knowledge, this is the first case of langerhans cell histiocytosis in a postmenopausal female with severe intractable pruritus which responded to Narrow Band UVB (NBUVB).

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