

Case Report

Multisystem Langerhans Cell Histiocytosis in Adult

Anubhav Garg, Pramod Kumar¹

From the Department of Dermatology, Venereology and Leprosy, RNT Medical College, Udaipur, Rajasthan, ¹Department of Dermatology, Venereology and Leprosy, Kasturba Medical College, Mangalore, Karnataka, India

Abstract

Langerhans cell histiocytosis (LCH), is a rare disorder, clinically presents with heterogeneous manifestations, and has an unpredictable outcome. Commonly seen in infancy or early childhood, the disorder is characterized by proliferation of abnormal and clonal Langerhans cell in skin, bone, lymph nodes, lungs, liver, spleen, and bone marrow. Occurrence of LCH in adults is rare. Here, we report the case of an adult with acute onset of polymorphic eruptions all over the body, which on biopsy showed features of multisystem LCH, and was confirmed by immunohistochemistry. Although multisystem LCH has a poor prognosis, our patient responded well to chemotherapy.

Key Words: Adult LCH, histiocytic infiltrate, langerhans cell histiocytosis

Introduction

Langerhans cell histiocytosis (LCH) is a rare, clinically polymorphous group of disorders, presenting with heterogeneous clinical manifestations and an unpredictable outcome.^[1] LCH is characterized by proliferation of abnormal and clonal Langerhans cells, in one or more organs like skin, bone, lymph nodes, lungs, liver, spleen, and bone marrow. Occurrence of LCH in adults is rare and is commonly seen in infants and early childhood.^[1,2] The disorder is difficult to diagnose in adults and once diagnosed, the multisystem LCH has a poor prognosis.

Case Report

A 26-year-old man, presented with a one-week history of polymorphous eruptions over face, trunk, and extremities, associated with painful sores in the mouth as well as fever. He had no history of any systemic illness. The lesions started around the mouth with greasy scaling, which gradually progressed to involve scalp, neck, trunk, and extremities, including genitals.

On general physical examination, jaundice, and inguinal lymphadenopathy were present. His vital statistics were stable. Systemic examination was within normal limits, except hepatosplenomegaly.

On cutaneous examination, multiple skin colored to yellowish papules and pustules were present over the scalp, retro-auricular area, face, neck, trunk, extremities, and genitalia, which were associated with facial edema [Figures 1 and 2]. Diffuse greasy scaling was present over the beard area and retro-auricular region. Oral mucosa showed multiple vesicles, pustules, and hemorrhagic crusting of lips [Figure 3].

Laboratory data showed: Hb-16.1 gm/dl; Total leukocyte count (TLC)-33,600/cu.mm; Neutrophil 54%, Lymphocyte 25%, Eosinophil 21%; total bilirubin-5.9 mg/dl;

direct bilirubin-3.5 mg/dl; serum glutamic oxaloacetic transaminase (SGOT)-179 U/L; serum glutamic pyruvic transaminase (SGPT)-269 U/L; alkaline phosphatase (ALP)-167 U/L; renal function test- within normal limits. USG of the abdomen showed fatty changes in the liver with hepatosplenomegaly.

Biopsy from the skin, liver, and bone marrow showed dense infiltrate of histiocytes with longitudinal nuclear groove and numerous eosinophils and lymphocytes [Figures 4 and 5]. Occasional mitoses were also seen in the histiocytic infiltrate. Immunohistochemistry was positive for S-100 [Figure 6].

With the history, clinical findings, and investigation, the diagnosis of multisystem LCH was made. The patient was started on chemotherapy under Medical Oncology Department. He was given Vinblastine and Etoposide weekly for 6 weeks with daily Prednisolone, followed by maintenance therapy of 12 cycles with same drugs at intervals of 3 weeks and daily 6-Mercaptopurine. The patient responded well and is on continuous follow-up till date.

Discussion

Lichtenstein, in 1953, coined the term Histiocytosis-X 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 LCH. LCH is a clonal neoplastic disorder,^[1-3] and its pathogenesis is unknown. Whether the infiltrating cells are truly neoplastic or reactive in nature is still a matter of debate.

Address for correspondence: Dr. Anubhav Garg, 71- A, Madhuban, Hospital Road, Udaipur-313001, Rajasthan, India. E-mail: dranubhav_garg@rediffmail.com

Access this article online	
Quick Response Code: 	Website: www.e-ijd.org
	DOI: 10.4103/0019-5154.92683



Figure 1: Scaling with crusting present over face, mimicking seborrheic dermatitis



Figure 2: Yellow-brown papules with pustules present over chest



Figure 3: Ulcers present over lips

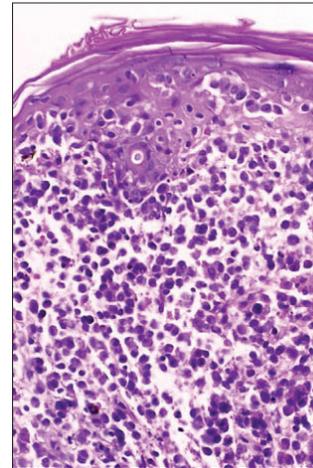


Figure 4: Diffuse histiocytic infiltrate, H and E staining $\times 10$

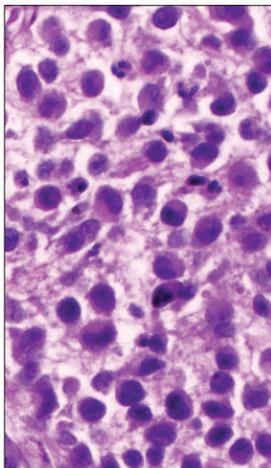


Figure 5: Histiocytic infiltrate, H and E staining $\times 100$

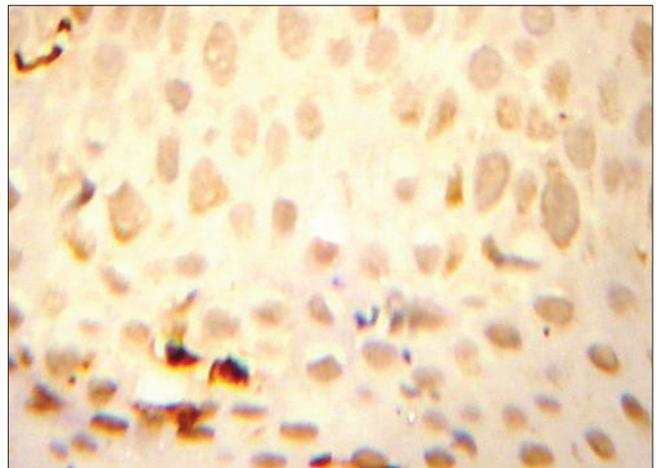


Figure 6: S-100 positive $\times 100$

The disease affects young children aged 1-4 years, but can occur at any age. The incidence is approximately 2-5 per million per year in infants and children and is even rarer in adults.^[1,2]

The Writing Group of Histiocytic Society (1987) has recently defined the criteria for diagnosis of Langerhans Cell Histiocytosis.^[4] The group proposed three levels of certainty in the diagnosis of LCH based on clinical features,

histopathology, and immunohistochemistry.

The most characteristic dermatological presentation is with scalp involvement. The scalp is erythematous with greasy scales, appearing much like seborrheic dermatitis. On the trunk, the lesions are discrete, yellow-brown scaly papules, often showing areas of purpura. Ulceration in the flexures, groins, and the perianal area is the common presentation in adults. The bone marrow involvement can be occult, or pancytopenia may occur. This presents as purpura. Lymph nodes are characteristically involved, especially cervical nodes. Solitary bone involvement with LCH is common and often goes undiagnosed. The most common sites are the bones of calvarium. Diabetes insipidus is the most common endocrinal abnormality associated with LCH caused by the infiltration of pituitary gland by Langerhans cells.^[5]

Histologically, there is a dense dermal infiltrate of Langerhans cell, recognized by abundant, amphophilic cytoplasm and round or kidney bean-shaped nucleus.^[5]

Immunohistochemistry is helpful in confirming the diagnosis of LCH. Infiltrating cells are S-100, CD1a, CD4, and HLA-DR positive. Electron microscopy will detect the presence of Birbeck granules.^[6]

A patient with isolated bone LCH lesions has the best prognosis. By contrast, 20% of the patients with multisystem involvement have a progressive disease course despite treatment.^[2] The prognosis is directly related to the age of onset, numbers of organ involved, and the extent of organ dysfunction. The prognosis of the disease depends on systemic rather than cutaneous involvement.^[5]

Treatment of LCH is controversial. In multisystem LCH, systemic chemotherapy is indicated. Etoposide as a single drug is better than other drugs tested, eg, vinka alkaloid, methotrexate, 6-MP.^[7]

Our patient responded well to the combination therapy of vinblastine, etoposide, and oral corticosteroids. He is currently on follow-up.

References

1. Lichtenstein L. Histiocytosis X; Integration of eosinophilic granuloma of bone, Letterer-Siwe disease and Hand-Schuller-Christian disease, as related manifestation of single nosologic entity. *Arch Pathol* 1953;56:84-102.
2. Howarth DM, Gilchrist GS, Mullan BP, Wiseman GA, Edmonson JH, Schomberg PJ. Langerhans cell histiocytosis: diagnosis, natural history, management and outcome. *Cancer* 1999;85:2278-90.
3. Munn S, Chu AC. Langerhans cell histiocytosis of the skin. *Haematol Oncol Clin North Am* 1998;12:269-86.
4. Chu T, Angio GJ, Farara BE. The writing group of the histiocytic society: Histiocytosis syndromes in children. *Lancet* 1987;1:208-9.
5. Vollum DI. Letterer-Siwe disease in adult. *Clin Dermatol* 1979;4:395-406.
6. Burgdorf WH. The histiocytosis. *Lever's Histopathology of the Skin*. 9th ed. Lippincott Williams and Wilkins, Philadelphia, 2005.p. 681-703.
7. Broadbent V, Pritchard J, Yeoman E. Etoposide (VP16) in the treatment of multisystem langerhans cell histiocytosis. *Med Pediatr Oncol* 1989;17:97-100.

How to cite this article: Garg A, Kumar P. Multisystem langerhans cell histiocytosis in adult. *Indian J Dermatol* 2012;57:58-60.

Received: July, 2010. **Accepted:** November, 2011.

Source of Support: Nil, **Conflict of Interest:** Nil.

Staying in touch with the journal

1) Table of Contents (TOC) email alert

Receive an email alert containing the TOC when a new complete issue of the journal is made available online. To register for TOC alerts go to www.e-ijd.org/signup.asp.

2) RSS feeds

Really Simple Syndication (RSS) helps you to get alerts on new publication right on your desktop without going to the journal's website. You need a software (e.g. RSSReader, Feed Demon, FeedReader, My Yahoo!, NewsGator and NewzCrawler) to get advantage of this tool. RSS feeds can also be read through FireFox or Microsoft Outlook 2007. Once any of these small (and mostly free) software is installed, add www.e-ijd.org/rssfeed.asp as one of the feeds.