

# Indian Journal of Dermatology, Venereology & Leprology

## CONTENTS

<p><b>Editor</b> Uday Khopkar</p> <p><b>Associate Editors</b> Ameet Valia Sangeeta Amladi</p> <p><b>EDITORIAL BOARD MEMBERS</b> Sandipan Dhar Sanjeev Handa H. R. Jerajani Sharad Mutalik C. M. Oberai M. Ramam D. A. Satish Rajeev Sharma Shruthakirti Shenoj C. R. Srinivas D. M. Thappa S. L. Wadhwa</p> <p><b>Ex-officio Members</b> A. K. Bajaj S. Sacchidanand</p> <p><b>EDITORIAL OFFICE</b> <b>Dr. Uday Khopkar</b> Editor, IJDVL 2/7, Govt. Colony, Haji Ali, Mumbai-400034. E-mail: editor@ijdv.com</p> <p><b>PUBLISHED BY</b> <b>Medknow Publications</b> 12, Manisha Plaza, M. N. Road, Kurla (W), Mumbai-400070, India. Phone: 91-22-25032970 Fax: 91-22-25032398 E-mail: publishing@medknow.com Website: www.medknow.com</p> <p><b>Manuscript submission</b> www.journalonweb.com/ijdv</p> <p><b>Cover design courtesy</b> Sudler &amp; Hennessey</p>	<p><b>EDITORIAL</b></p> <p><b>PRESIDENTIAL ADDRESS</b></p> <p><b>REVIEW ARTICLE</b></p> <p><b>STUDIES</b></p> <p><b>CASE REPORTS</b></p>	<p><b>IJDVL at the crossroads</b></p> <p><b>A. K. Bajaj</b></p> <p><b>Serious cutaneous adverse drug reactions: Pathomechanisms and their implications to treatment</b> Arun C. Inamdar, Aparna Palit</p> <p><b>Diltiazem vs. nifedipine in chilblains: A clinical trial</b> A. K. Patra, A. L. Das, P. Ramadasan</p> <p><b>A comparative study of PUVASOL therapy in lichen planus</b> Lata Sharma, M. K. Mishra</p> <p><b>Utility of polymerase chain reaction as a diagnostic tool in cutaneous tuberculosis</b> Padmavathy L., Lakshmana Rao L., Veliath A. J.</p> <p><b>Therapeutic efficacy of intralesional triamcinolone acetonide versus intralesional triamcinolone acetonide plus lincomycin in the treatment of nodulocystic acne</b> B. B. Mahajan, Geeta Garg</p> <p><b>Ichthyosiform sarcoidosis following chemotherapy of Hodgkin's disease</b> M. P. S. Sawhney, Y. K. Sharma, V. Gera, S. Jetley</p> <p><b>Urticarial vasculitis in infancy</b> Sukhjot Kaur, Gurvinder P. Thami</p> <p><b>Koebner phenomenon in PLEVA</b> Arun C. Inamdar, Aparna Palit</p> <p><b>Familial acrogeria in a brother and sister</b> Shaikh Manzoor Ahmad, Imran Majeed</p> <p><b>Cornelia de Lange syndrome</b> K. Muhammed, B. Safia</p>	<p>_____ 203</p> <p>_____ 204</p> <p>_____ 205</p> <p>_____ 209</p> <p>_____ 212</p> <p>_____ 214</p> <p>_____ 217</p> <p>_____ 220</p> <p>_____ 223</p> <p>_____ 225</p> <p>_____ 227</p> <p>_____ 229</p>
---	--	---	---

# Indian Journal of Dermatology, Venereology & Leprology

## CONTENTS (CONTD.)

The Indian Journal of Dermatology, Venereology and Leprology is a bimonthly publication of the Indian Association of Dermatologists, Venereologists and Leprologists and published by Medknow Publications.

The Journal is indexed/listed with Health and Wellness Research Center, Health Reference Center Academic, InfoTrac One File, Expanded Academic ASAP, NIWI, INIST, Uncover, JADE (Journal Article Database), IndMed, Indian Science Abstract's and PubList.

All the rights are reserved. Apart from any fair dealing for the purposes of research or private study, or criticism or review, no part of the publication can be reproduced, stored, or transmitted, in any form or by any means, without the prior permission of the Editor, Indian Journal of Dermatology, Venereology and Leprology.

The information and opinions presented in the Journal reflect the views of the authors and not of the Indian Journal of Dermatology, Venereology and Leprology or the Editorial Board or the Indian Association of Dermatologists, Venereologists and Leprologists. Publication does not constitute endorsement by the journal.

The Indian Journal of Dermatology, Venereology and Leprology and/or its publisher cannot be held responsible for errors or for any consequences arising from the use of the information contained in this journal. The appearance of advertising or product information in the various sections in the journal does not constitute an endorsement or approval by the journal and/or its publisher of the quality or value of the said product or of claims made for it by its manufacturer.

For advertisements, please contact the Editor

	<b>Intralesional steroid induced histological changes in the skin</b>	
	Sukhjot Kaur, Amanjeet, Gurvinder P. Thami, Harsh Mohan	_____ 232
	<b>Sparfloxacin induced toxic epidermal necrolysis</b>	
	M. Ramesh, G. Parthasarathi, B. Mohan, A. B. Harugeri	_____ 235
	<b>Fever due to levamisole</b>	
	Ramji Gupta, Sameer Gupta	_____ 237
	<b>Localized cutaneous sporotrichosis lasting for 10 years</b>	
	Sanjay K. Rathi, M. Ramam, C. Rajendran	_____ 239
<b>QUIZ</b>	S. V. Rakesh, D. M. Thappa	_____ 241
<b>RESIDENT'S PAGE</b>	<b>Sign of Nikolskiy &amp; related signs</b>	
	Deepa Sachdev	_____ 243
<b>RESEARCH METHODOLOGY</b>	<b>Declaration of Helsinki: The ethical cornerstone of human clinical research</b>	
	Gulrez Tyebkhan	_____ 245
<b>MEDICOLEGAL WINDOW</b>	<b>Drug eruptions and drug reactions</b>	
	Subodh P. Sirur	_____ 248
<b>LETTERS TO EDITOR</b>	<b>Aggravation of preexisting dermatosis with <i>Aloe vera</i></b>	
		_____ 250
	<b>Familial woolly hair in three generations</b>	_____ 250
	<b>Chronic pelvic inflammatory disease and melasma in women</b>	_____ 251
	<b>Comments on "Serological study for sexually transmitted diseases in patients attending STD clinics in Calcutta"</b>	_____ 252
<b>BOOK REVIEW</b>	<b>Colour atlas and synopsis of paediatric dermatology</b>	
	Sandipan Dhar	_____ 255
<b>ANNOUNCEMENTS</b>		_____ 255, 256,
<b>INSTRUCTIONS TO AUTHORS</b>		_____ 258

## Quiz

---

A 6-month-old male baby, born at full term pregnancy of non-consanguineous parents, was seen in the dermatology clinic with a 4 months' history of scaly lesions over the scalp and forehead and a purpuric rash over the trunk of three months' duration. The scalp lesions had been unresponsive to various topical antifungals applied by the mother over the past 1 month. His medical history was uneventful except for a recurrent ear discharge and intertriginous oozing dermatitis in the neck flexure over the past 3 months.

His physical examination revealed diffuse erythematous greasy scaly and crusted lesions over the scalp, extending onto the forehead (Figure 1). On the trunk, multiple hypopigmented as well as minute purpuric macules and papules were seen predominantly involving the anterior abdomen (Figure 2). Apart from a cleft lip, the oral cavity demonstrated gingival hyperplasia. Systemic examination revealed gross hepatosplenomegaly. The rest of the physical examination was normal.

Baseline hematological and biochemical investigations were within normal limits. Sonogram of the abdomen showed increased liver and spleen span. Histopathological examination of a biopsy

taken from the purpuric papular lesion on the abdomen showed papillary dermal edema and an infiltrate of large cells with eosinophilic cytoplasm and kidney shaped nuclei. The cells infiltrated the epidermis at a few places. The epidermis was stretched and thinned out (Figure 3). His skull roentgenogram showed well-defined lytic areas with non-sclerotic borders on the cranial vault.

### WHAT IS YOUR DIAGNOSIS?



Figure 2: Hypopigmented as well as purpuric macules and papules over the trunk



Figure 1: Papulosquamous rash of the scalp extending onto the forehead and ear



Figure 3: Photomicrograph showing upper papillary dermal edema and infiltrate of large cells with eosinophilic cytoplasm and irregular as well as kidney shaped nuclei (H&E X100)

**Diagnosis:** Langerhans cell histiocytosis (Letterer-Siwe disease)

## DISCUSSION

This case illustrates the characteristic clinical, histological and radiological features of Langerhans cell histiocytosis (LCH), a reactive condition in which cells with the phenotype of Langerhans cells infiltrate in various tissues and cause damage to the tissues. These infiltrating cells are named as 'LCH cells'.<sup>1,2</sup> Langerhans cell histiocytosis (LCH) has replaced the older term 'histiocytosis-X', originally coined by Lichtenstein in 1953 to link three related clinical entities of unknown origin (Letterer-Siwe disease, Hand-Schüller-Christian disease, and eosinophilic granulomas).<sup>3</sup> The unifying feature of these conditions is the presence of rod shaped cytoplasmic structures inside the 'LCH cell' that are indistinguishable from the Birbeck granules of epidermal Langerhans cells.<sup>4</sup>

The etiology of LCH remains elusive. Viral, immunological and neoplastic pathogenic mechanisms have been considered, but none has been proved. In view of the spontaneous regression in some cases, it is believed that LCH is a non-neoplastic disease.<sup>2</sup>

Letterer-Siwe disease (LSD) is the acute disseminated multisystemic form of LCH.<sup>5</sup> It most commonly presents in the first year of life and cutaneous involvement is an early feature. The characteristic presentation is a seborrheic dermatitis-like rash of the scalp with erythematous greasy scaling. Discrete yellow-brown scaly papules, often purpuric (representing a poor prognostic sign), may occur on the trunk. Vesicles and pustules may occur, simulating eczema, miliaria, scabies and varicella. Gingival and oral mucosal ulcerations are also commonly seen. Painful ulcerated inguinal, retroauricular, and external auditory canal nodules may occur in some patients. Temporal bone involvement may masquerade as chronic otitis media, causing a delay in the diagnosis as in our case. Marked hepatomegaly is a frequent complication and is a bad prognostic sign, particularly when accompanied by jaundice.<sup>6</sup> Splenomegaly is less frequent. Classically, the course of LSD is rapid and fatal.<sup>2</sup>

The characteristic histological appearance of the Letterer-Siwe type of LCH is a patchy lichenoid infiltrate of large histiocytes that have a large kidney shaped, indented or bilobed nucleus and abundant homogeneous pink cytoplasm. The histiocytes frequently infiltrate the epidermis, sometimes causing a pagetoid pattern. Three kinds of histological reactions have been described in LCH, proliferative, granulomatous and xanthomatous, but only the first two are commonly seen.<sup>7</sup> If facilities are available, marker studies for S-100 and CD1a as well as electron microscopy may be done.

The management of LCH depends on the age of the patient, extent of the disease and location of the disease.<sup>2</sup> The best option for children with only skin involvement is observation. Topical nitrogen mustard (20%) may be an effective treatment. The use of systemic glucocorticoids or anti-mitotic drugs should be considered for resistant cases. Systemic chemotherapy with prednisolone and vinca alkaloids is indicated for multi-system disease. Etoposide, cyclosporine and IFN- $\gamma$  have also been reported to be beneficial in systemic disease.

## REFERENCES

1. Chu AC. Histiocytosis. In: Champion RH, Burton JL, Burns DA, Breathnach DA, editors. *Rook/Wilkinson/Ebling Textbook of dermatology*. 6th ed. Oxford: Blackwell Science; 1998. p. 2311-36.
2. Caputo R. Langerhans cell histiocytosis. In: Freedberg IM, Essen AZ, Wolff K, Austen KF, Goldsmith LA, Katz SI, et al, editors. *Fitzpatrick's Dermatology in general medicine*. 5th ed. New York: McGraw-Hill; 1999. p. 1882-92.
3. Caldemeyer KS, Parks ET, Mirowski GW. Langerhans cell histiocytosis. *J Am Acad Dermatol* 2001;44:509-11.
4. Jerajani HR, Amladi ST. Skin tumors and lymphoproliferative disorders. In: Valia RG, Valia AR, editors. *IADVL Textbook and atlas of dermatology*. 2nd ed. Mumbai: Bhalani Publishing House; 2001. p. 1149-244.
5. Gianotti F, Caputo R. Histiocytic syndromes: A review. *J Am Acad Dermatol* 1985;13:383-404.
6. Belaich S. Langerhans cell histiocytosis. *Dermatology* 1994;189(Suppl. 2):2-7.
7. Burgdorf WHC. Histiocytosis. In: Elder D, Elenitsas R, Jaworsky C, Johnson Jr B, editors. *Lever's Histopathology of the skin*. 8th ed. Philadelphia: Lippincott-Raven; 1997. p. 591-616.

**S. V. Rakesh, D. M. Thappa**

Department of Dermatology & STD, JIPMER,  
Pondicherry-605006, India. E-mail: dmthappa@vsnl.net