

Hansen's disease associated with erythromelalgia mimicking Lupus erythematosus

Shenoi Shrutakirthi Damodar, Prabhu Smitha, Balakrishnan Nirmal, Nayak Sudhir U.K., Pai Sathish Ballambat

Department of Dermatology and Venereology, Kasturba Medical College, Manipal University, Manipal, Karnataka, India

ABSTRACT

Hansen's disease, though considered to be at the verge of elimination in many countries including India, still continues to surprise patients and dermatologists alike. This is mainly due to its varying and unconventional presentations which may lead to initial misdiagnosis and prolongation of treatment. Here we describe an unusual case presenting with erythematous photosensitive facial lesions associated with erythromelalgia of the finger tips, provisionally diagnosed as SLE. A subsequent histopathology examination proved it to be Hansen's disease Borderline Tuberculoid variety. Hansen's disease can be termed as the modern great imitator, displacing the traditional great imitator, syphilis.

Key words: Erythromelalgia, Hansen's disease, histopathology, Lupus erythematosus

INTRODUCTION

Hansen's disease, though considered to be eliminated in many countries including India, still continues to surprise patients and dermatologists alike. This is because it presents in varied forms and mimics many other diseases leading to initial misdiagnosis and prolongation of treatment. In a country like India, where leprosy is still rampant, the clinician should have a high degree of suspicion and clinical acumen to pinpoint the diagnosis.

CASE REPORT

A 39-year-old male teacher presented with reddish skin lesions over the face with history of increase in redness and burning sensation on sun exposure of 25-day duration. He had also noticed redness and burning over the finger tips which was relieved by immersion of hands in cold water. In addition, he had intermittent joint pains involving knee and interphalangeal joints of 4-year duration. There were no systemic complaints such as hematuria, seizures, dyspnea or palpitation, although there was one episode of oral ulcers 10 days ago.

Examination revealed multiple well-defined, irregular erythematous mildly scaly plaques

over the forehead, cheeks, ears, V area of neck [Figure 1]. There was erythema, warmth, and mild tenderness over the bilateral finger tips [Figure 2]. There was no sensory loss over the skin lesions or thickening of peripheral nerves. Sensory and motor examination of the extremities was normal. Hot and cold water immersion tests for erythromelalgia and Raynaud's phenomenon were negative with no worsening of redness or bluish discoloration of fingers, respectively.

In view of arthritis, photosensitivity, history of oral ulcers, a clinical diagnosis of acute cutaneous lupus erythematosus with erythromelalgia was considered and patient investigated. The hemogram, chest X-ray and renal function tests, serum C3 and C4 levels were normal. Liver function tests were normal except for mildly elevated ALT. ANA was negative. USG abdomen showed grade 2 fatty infiltration of liver. Blood sugars were elevated.

Slit smear examination for lepra bacilli and direct immunofluorescence test of skin lesions were negative. Treatment was commenced with 32 mg of oral methylprednisolone per day along with subcutaneous insulin. H and E examination of skin biopsy specimen showed

Access this article online

Website: www.idoj.in

DOI: 10.4103/2229-5178.126035

Quick Response Code:



Address for

correspondence:

Dr. Smitha Prabhu,
Department of Dermatology and Venereology, Kasturba Medical College Manipal University, Manipal, India.

E-mail:
drsmithaprabhu@yahoo.com

thinning and follicular plugging of epidermis, a grenz zone with ill-defined collections of lymphocytes, macrophages, and epithelioid cells in a perineural, periadnexal, and perivascular location [Figures 3-5]. Fite stain was positive for lepra bacilli.

A revised diagnosis of borderline tuberculoid leprosy with type 1 reaction was made and antileprosy treatment as for

multibacillary regime with 3 drugs (oral pulse of Rifampicin 600 mg and clofazimine 300 mg once in 28 days, daily oral clofazimine 50 mg and dapsone 100 mg) was started along with oral steroids in tapering doses. The facial lesions and finger tip erythema resolved in 2-month time [Figure 6] and patient is on regular follow-up.

DISCUSSION

Leprosy is a chronic granulomatous disease caused by *Mycobacterium leprae* which primarily affects the peripheral nerves and skin. The diagnosis can pose a clinical challenge with its diverse manifestations resulting from granulomatous infiltrate further compounded by reactional states.^[1] Leprosy can masquerade as dermatophytosis, lupus erythematosus, sarcoidosis, psoriasis, seborrhoeic dermatitis, tuberculosis, granuloma annulare, and a myriad of other conditions.

The sudden development of erythematous plaques on the face associated with photosensitivity and fingertip erythema without sensory loss or nerve thickening in our patient suggested an initial diagnosis of lupus erythematosus, but ANA and lupus



Figure 1: Erythematous plaques over the face at presentation



Figure 2: Erythema over finger tips at presentation

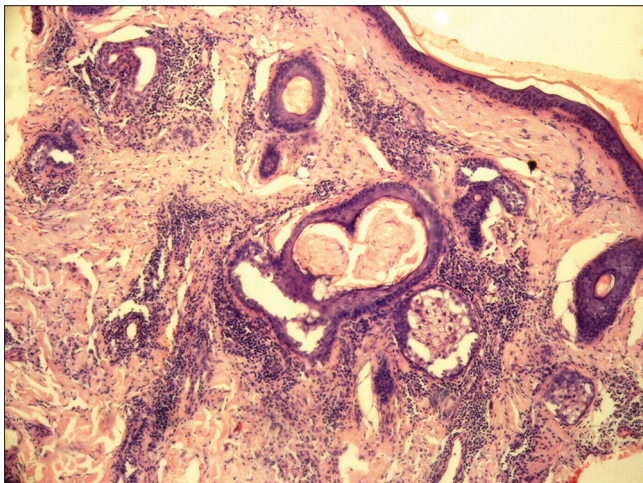


Figure 3: Histopathological specimen – H and E, ×10 stain showing attenuated epidermis with a grenz zone and dermis containing ill-defined collections of lymphocytes, macrophages and epithelioid cells in a perineural, periadnexal and perivascular location

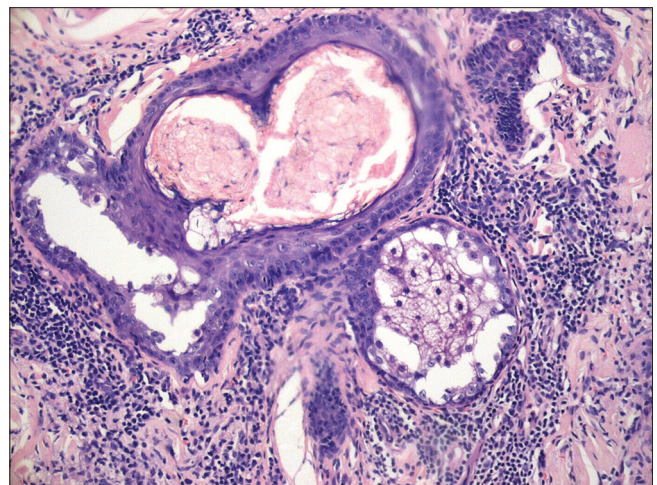


Figure 4: Histopathological specimen – H and E, ×40 showing periappendageal concentration of inflammatory cells consisting of lymphocytes, epithelioid cells and macrophages

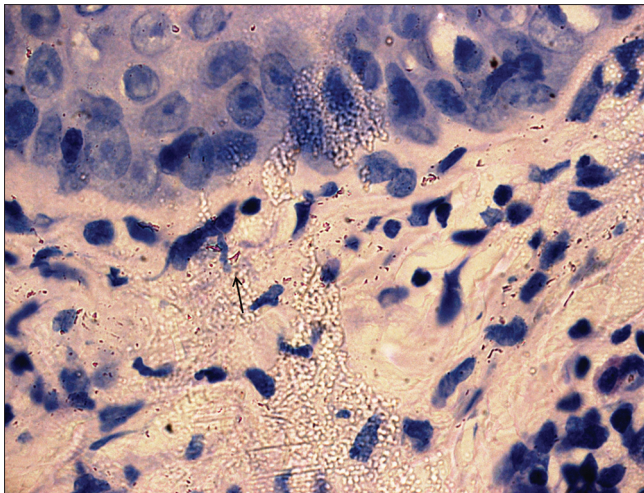


Figure 5: Fite stain showing acid-fast bacilli (marked with arrow)

band tests were negative. Some patients with leprosy do not notice their lesions until they become inflamed during a reaction^[1] as in this case. It is well known that leprosy lesions on the face do not show sensory loss due to the rich innervation and intermingling of nerves.

Our patient had simultaneous appearance of skin lesions and erythema and burning sensation of the finger tips resembling erythromelalgia. Erythromelalgia, also known as red neuralgia, acromelalgia or Mitchell's disease is a functional vascular disease like Raynaud's syndrome and acrocyanosis characterized by paroxysmal burning pain and erythema over the extremities and may be precipitated by emotional stress, injury, pressure, and various other stimuli.^[2] Primary erythromelalgia is idiopathic in nature and is more common in males. Secondary erythromelalgia is associated with hypertension, rheumatoid arthritis, lupus erythematosus, thromboangiitis obliterans, gout, and myeloproliferative disorders such as polycythemia rubra vera and thrombocythemia. Other causes include drugs like calcium channel blockers, misoprostone, bromocriptine, topical isopropanolol, mushroom and mercury poisoning, and certain pox virus infections (Erythromelalgia-related Pox Viruses – ERPV)^[3] and even diabetes mellitus, although pathophysiological mechanism in primary erythromelalgia is thought to be shunting of blood through arteriovenous anastomosis in skin and episodic blockade and inflammation of blood vessels. Lately, there are reports on pathological changes in peripheral nervous system due to mutations in gene SCN 9A on chromosome 2q which encodes voltage-gated sodium channels expressed in nociceptive dorsal root ganglion neurons leading to their hyper excitability in a temperature-dependent fashion, as also changes in sympathetic ganglion neurons.^[4] A small fiber neuropathy leading to disturbance of neural control of vascular muscle tone and increased episodic blood flow to the affected area has been demonstrated in most patients and erythromelalgia is considered to be a prototype of painful neuropathies.^[5] In leprosy a damage to vascular innervation



Figure 6: Two months post-treatment with antileprosy therapy showing resolution of facial plaques

due to sympathetic nerve damage causes loss of vascular tone and stasis of capillary blood.^[1] Endarteritis leading to ischemia of nerves may be responsible for the sensory, motor, and autonomic disturbances.^[6] Impairment of vasomotor reflexes has been demonstrated in leprosy even before the appearance of skin lesions.^[7,8]

Leprosy can have a myriad of clinical presentations and a high index of suspicion is needed especially in diagnosing atypical cases.

Acute intermittent porphyria has been described as “the little imitator” and syphilis as “the great imitator”, but perhaps leprosy is “the greatest imitator” of all.

In our case we postulate that the finger tip erythema simulating erythromelalgia is a consequence of autonomic dysfunction. Hence leprosy has to be included as one of the causes of secondary erythromelalgia. So far, there are no published case reports on this association, though there are a few reports on erythromelalgia of the earlobes,^[9] wherein leprosy has to be ruled out, especially in endemic areas.

ACKNOWLEDGMENT

Dr. Manna Valiathan, Professor, Department of Pathology, Kasturba Medical College, Manipal University, for reporting the histopathology findings.

REFERENCES

1. Brycesson AD, Pfaltzgraff RE. Leprosy, 2nd ed. Churchill – Livingstone, Edinburgh; 1984. p. 25.
2. Heidrich H. Functional vascular diseases: Raynaud's syndrome, acrocyanosis and erythro melalgia. *Vasa* 2010;39:33-41.
3. Zheng ZM, Specter S, Zhang JH, Friedman H, Zhu WP. Further

- characterization of the biological and pathogenic properties of erythromelalgia-related pox viruses. *J Gen Virol* 1992;73:2011-9.
4. Drenth JP, te Morsche RH, Guillet G, Taieb A, Kirby RL, Jansen JB. SCN9A mutations define primary erythromelalgia as a neuropathic disorder of voltage gated sodium channels. *J Invest Dermatol* 2005;124:1333-8.
 5. Davis MD, Sandroni P, Rooke TW, Low PA. Erythromelalgia: Vasculopathy, neuropathy, or both? a prospective study of vascular and neurophysiologic studies in erythromelalgia. *Arch Dermatol* 2003;139:1337-43.
 6. Carayon A. Dharmendra. Vascular changes in leprosy. In: *Textbook of Leprosy*, vol. 2., Samant and Co; 1985. p. 853-71.
 7. Illarramendi X, Bühner-Sékula S, Sales AM, Bakker MI, Oliveira A, Nery JA, *et al.* High prevalence of vasomotor reflex impairment in newly diagnosed leprosy patients. *Eur J Clin Invest* 2005;35:658-65.
 8. Beck JS, Abbot NC, Samson PD, Butlin CR, Grange JM, Cree IA, *et al.* Impairment of vasomotor reflexes in the fingertips of leprosy patients. *J Neurol Neurosurg Psychiatry* 1991;54:965-71.
 9. Berk DR, Eisen AZ. Erythromelalgia of the ears: An unusual variant and response to therapy. *J Drugs Dermatol* 2008;7:285-7.

Cite this article as: Damodar SS, Smitha P, Nirmal B, Sudhir NU, Ballambat PS. Hansen's disease associated with erythromelalgia mimicking Lupus erythematosus. *Indian Dermatol Online J* 2014;5:59-62.

Source of Support: Nil, **Conflict of Interest:** None declared.