International Journal of Advances in Medicine
Dhillon KS et al. Int J Adv Med. 2015 May;2(2):185-187
http://www.ijmedicine.com
pISSN 2349-3925 | eISSN 2349-3933
DOI: 10.5455/2349-3933.ijam20150525

Case Report

Erythema necroticans: a rare reaction pattern in leprosy

K S Dhillon[1]*, Deepak Sharma[1], Tarunveer Singh[1], Nishi Tandon[2], Gaurav Sarin[1], Areeba Khan[1], Soni Yadav[1], Sweksha Srivastava[1]

[1]Department of Dermatology, Era's Lucknow Medical College, Lucknow, India
[2]Department of Pathology, Era's Lucknow Medical College, Lucknow, India

Received: 06 February 2015
Accepted: 13 March 2015

***Correspondence:**
Dr. K S Dhillon,
E-mail: kanwarjit29@yahoo.co.in

ABSTRACT
Leprosy reactions are rare expression of immunological perturbations that interrupt the usual chronic course and the clinical stability of patients with leprosy. They generally occur during anti-mycobacterial treatment and are characterized by the appearance of crops of brightly erythematous tender nodules or plaques. Erythema Necroticans (ENe) is an uncommon manifestation of Type 2 Erythema Nodosum Leprosum (ENL) reaction, encountered in lepromatous and borderline lepromatous cases of leprosy. Severe ENL can become vesicular or bullous and break-down and is termed Erythema Necroticans.

Keywords: Erythema Nodosum Leprosum, Erythema Necroticans, Lucio Phenomenon

INTRODUCTION

Leprosy is a major health problem in India with a prevalence of 3.22/10000, affecting all age groups from infancy to old age.[1] it is a disease of slow development that presents a wide spectrum of clinical, histo-pathological and immunological characterization. Leprosy reactions are rare and not well-known states that interrupt the usual chronic course and the clinical stability of patients with leprosy. They are expression of immunological perturbations.[2] Erythema Nodosum Leprosum (ENL) is an immune complex-mediated reaction that may complicate the course of multibacillary leprosy. It generally occurs during MDT and is characterized by the appearance of crops of brightly erythematous tender nodules or plaques. Severe ENL can become vesicular or bullous and break-down and is termed Erythema Necroticans.[3] The extra-cutaneous manifestation include neuritis, iridocyclitis, orchitis and lymphadenopathy. Fever and other constitutional symptoms are usually associated with it.[4]

CASE REPORT

A 36 year old female patient came to our OPD with complaints of multiple painful raised lesions all over the body and fever on & off for last 8 months. She was asymptomatic 8 months back when she started developing intermittent fever, which was high grade in nature. It was associated with arthralgia and constitutional symptoms. Concurrent with febrile episodes, patient developed crops of multiple red, round elevated lesions all over the body (Figure 1, 2) with predilection for lower limbs (Figure 3). The lesions were very painful and usually occurred in the evening accompanied by fever. She did not have any other chronic ailment. Bowel and bladder habits were normal. On examination, patient was febrile. Pallor was present. Multiple, tender, erythematous, nodules were present all over the body including face interspersed with skin necrosis of various sizes lesions on forearm, gluteal region, thighs and legs. Local temperature was raised. Temperature sensation was lost over lesions of the extensor surface of the left forearm. Left ulnar nerve was thickened and tender.

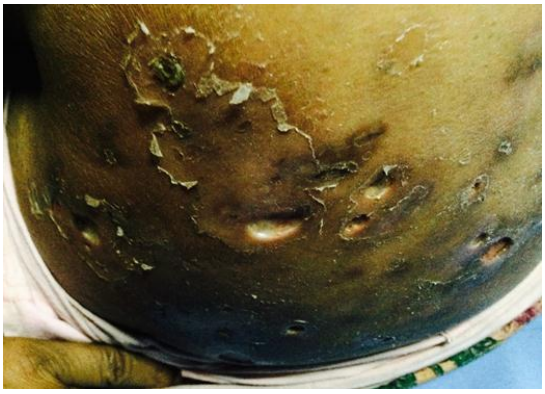

Figure 1: Ulcerative lesions over abdomen.

Both lateral popliteal and radial nerves were also thickened. Diagnosis of leprosy was confirmed by slit-skin smear (sss):5+ afb. Her HB was 7.8g/dl and her TLC was 19,000 cells/mm3 with neutrophilia. ESR and c-reactive protein were raised. All other biochemical & immunological parameters were within normal limits.

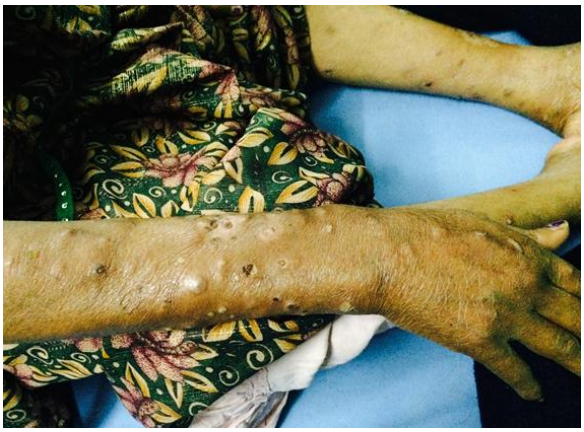

Figure 2: Nodules and ulcerations over forearms.

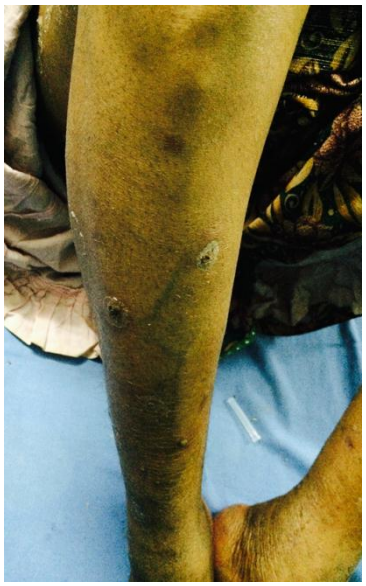

Figure 3: Nodulo-ulcerative lesions over lower leg.

Histopathologic analysis showed poorly defined epithelioid granulomas with lepra cells and numerous inflammatory cells including neutrophils, lymphocytes and rare eosinophils centred around blood vessels, nerves and adnexal structure occupying the superficial, mid and deep dermis (Figure 4). MDT therapy was exhibited along with tab prednisone 30 mg twice a day initially with gradual tapering. Dose of cap clofazimine was increased to 100 mg thrice a day. Patient responded satisfactorily and ulcers started healing after institution of therapy.

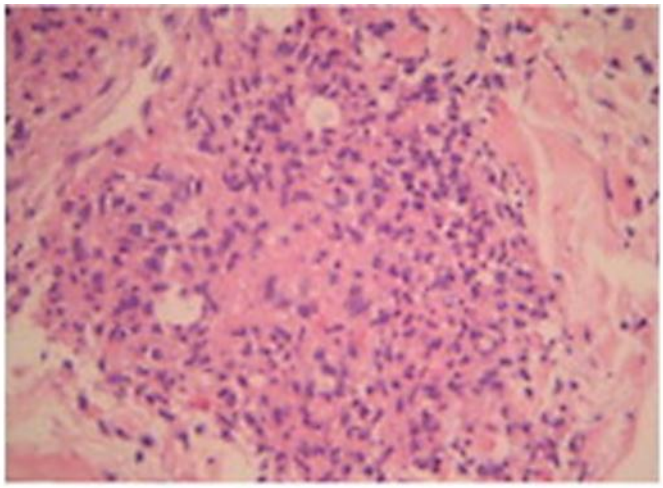

Figure 4: Dermal infiltrate (predominantly lymphocytes) with loss of adnexal structure.

DISCUSSION

Erythema Nodosum Leprosum (ENL) is an immune complex-mediated reaction that may complicate the course of multibacillary leprosy. It generally occurs during MDT and is characterized by the appearance of crops of brightly erythematous tender nodules or plaques. Severe ENL can become vesicular or bullous and break-down and is termed Erythema Necroticans. Erythema Nodosum Leprosum occurs usually during the treatment of lepromatous leprosy. However, there have been a fair number of reports of untreated lepromatous leprosy with subtle initial changes, presenting de novo as ENL, or it may present with extraordinary manifestations, often with long delay before the diagnosis of leprosy is considered.[5,6] The probable trigger factors associated with ENL reaction include surgical operations, pregnancy, parturition, lactation, menstruation, trauma, inter-current illness, vaccination, physical or mental stress and sometime therapy.[7] Tumor necrosis factor-α (TNF-α), a pro-inflammatory cytokine, plays an important role in the pathogenesis of ENL. There is activation of T-cell and macrophages, inducing production of large amounts of TNF-α.[8] Additional source of TNF-α is the severe leukocytosis and the intense neutrophilic infiltrate in ENL lesions. Plasma levels of this cytokine have been found to be high during the episode of ENL. High CRP levels along with a positive correlation between elevated

TNF-α and CRP levels in the serum of ENL patients has been reported.[9] Three types of ENL are identified: single acute ENL, multiple acute ENL (repeated discrete episodes) and chronic ENL (continuous episodes). 92% of ENL reactions are usually chronic and relapsing with unpredictable clinical course.[10] Ulcers are an unusual presentation of leprosy. These occur as a result of loss of sensation (peripheral), ENL (rarely), or due to Lucio's phenomenon.[11-13] Besides the classic presentation of erythema nodosum, severe ENL very rarely presents as ulcerative skin lesions (erythema necroticans).[13,14] Lucio phenomenon is a similar entity as Erythema necroticans but there is no fever or constitutional symptoms. Multiple drug therapy (MB-MDT) for borderline lepromatous leprosy should preferably be taken as per the recommendations of WHO. In addition, treatment with prednisolone should be instituted for 12 weeks course. The WHO has recommended the anti-inflammatory clofazimine for chronic and severe ENL and as steroid-sparing agent.[15] However, it takes 4–6 weeks for the effect of clofazimine to be clinically detectable. Thus, it is not useful for the management of acute ENL. Thalidomide is now considered the drug of choice for ENL. It was approved by the FDA in 1998 in the acute treatment of moderate to severe ENL and preventing new episodes. The search for an effective alternatives, led to experiencing second-line drugs such as pentoxifyline[16], mycophenolate mofetil[17], azathioprine and infliximab[18] with inconsistent results.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Burman KD, Rijall A, Agrawal S, Agarwalla A, Verma KK. Childhood leprosy in eastern Nepal: a hospital- based study. Indian J Lepr. 2003;75(1):47-52.
2. Sehgal VN, Srivastava G, Sundharam JA. Immunology of reactions in leprosy. International Journal of Dermatology.1988;27(3):157–62.
3. Jopling WH, Mc Dougall, AC. Leprosy reactions. In: Handbook of leprosy, 5th ed. CBS Publishers, New Delhi.1996;82–91.
4. Cuevas J, Rodríguez-Peralto JL, Carrillo R, Contreras F. Erythema nodosum leprosum: reactional leprosy. Semin Cutan Med Surg. 2007;26(2):126-30.
5. Zannatun N, Fazle RM, Monzurul HC, Ziaus S, A. Billal A, Nazmul HAM. Steroid dependent ulcerated erythema nodosum leprosum 16 years after successful treatment of leprosy. J Med. 2009;10(1):31–3.
6. Mc Dougall AC, Archibald GC. Lepromatous leprosy presenting with swelling of the legs. Br. Med J. 1977;1(6052):23–24.
7. Verma KK, Pandhi RK. Necrotic erythema nodosum leprosum; presenting manifestation of lepromatous leprosy. Int. J. Lepr.1993;61(2):293-4.
8. Kaushal KV, Srivastava P, Anil M, Verma K. Role of azathioprine in preventing recurrences in a patient of recurrent erythema nodosum leprosum. Leprosy Review. 2006;77(3):225-9.
9. Foss NT, De Oliveira EB, Silva CL. Correlation between TNF production, increase of plasma C-reactive protein level and suppression of T lymphocyte response to concanavalin A during erythema nodosum leprosum. Int J Lepr other Mycobact Dis. 1993;61(2): 218–26.
10. Pocaterra L, Jain S, Reddy R, Muzaffarullah S,Torres O,Suneetha S. Clinical course of erythema nodosum leprosum: an 11-year cohort study in hyderabad, india. Am J Trop Med Hyg. 2006;74(5):868–79.
11. Britton WJ, Lockwood DN. Tropical Infectious Diseases: Principles, Pathogens and Practice Leprosy. Lancet 2004;363(9416):1209-19.
12. Kasper D, Fauci A, Hauser S,Longo D,Jameson J, Loscalzo J. Harrisons Principles of Internal Medicine. McGraq Hill.16th edn. New York. 2004;1:966-72.
13. Davis SV, Shenoi SD, Balachandran C, Pai SB. A fatal case of erythema necroticans. Indian J Lepr. 2002;74(2):145-9.
14. Dixit VB. Diffuse lepromatosis with recurring ulcerative erythema nodosum leprosum. Indian J Lepr. 1992;64(1):112-3.
15. Pannikar V. The return of thalidomide: new uses and renewed concerns. Lepr Rev. 2003;74(3):286–8.
16. Sales AM, De Matos HJ, Nery JA, Duppre NC, Sampaio EP, Sarno EN. Double-blind trial of the efficacy of pentoxifyline vs thalidomide for the treatment of type II reaction in leprosy. Braz J Med Biol Res. 2007;40(2):243-8.
17. Kalyan B, Raghubir B. Management of erythema nodosum leprosum by mycophenolate mofetil. Indian J. Dermatol. 2008;53(3):142–3.
18. Williame RF, Annigie JJ,Wouter FMG.Treatment of recurrent erythema nodosum leprosum with infliximab. N Engl J Med. 2006;355(7):739.

DOI: 10.5455/2349-3933.ijam20150525
Cite this article as: Dhillon KS, Sharma D, Singh T, Tandon N, Sarin G, Khan A, Yadav S, Srivastava S. Erythema necroticans: a rare reaction pattern in leprosy. Int J Adv Med 2015;2:185-7.