Volume 73, Number 2
Printed in the U.S.A.
(ISSN 0148-916X)

CASE REPORT

Erythema Nodosum Leprosum Necroticans in a Child—An Unusual Manifestation[1]

Deepika Pandhi, Shilpa Mehta, Subhav Agrawal, and Archana Singal[2]

ABSTRACT

Erythema nodosum leprosum necroticans is an uncommon manifestation of type 2 lepra reaction, encountered in lepromatous and borderline lepromatous cases of leprosy. We report an unusual clinical presentation of necrotic erythema nodosum leprosum in a 9-year-old boy with no pre-existing evidence of leprosy. The lesions resolved completely following multibacilliary multi-drug therapy for 12 months, non-steroidal anti-inflammatory drugs and corticosteroids.

RESUME

L'érythème noueux lépreux nécrotique est une complication inhabituelle des réactions lépreuses de type 2. Elle est rencontrée dans les cas de lèpre lépromateuse et lépromateuse borderline. Nous rapportons ici la présentation clinique inhabituelle, chez un garçon de 9 ans, d'un érythème noueux lépreux nécrotique, sans diagnostic préalable de lèpre. Les lésions rétrocédèrent complètement après la mise en œuvre d'un traitement incluant une polychimiothérapie contre la lèpre multibacillaire, des corticoïdes et des anti-inflammatoires non-stéroïdiens.

RESUMEN

El eritema nodoso leproso necrotizante es una manifestación poco común de las reacciones leprosas tipo 2 que aparecen con cierta frecuencia en los pacientes con lepra BL y LL. En esta comunicación se reporta un caso de esta rara complicación en un niño de 9 años de edad que no había mostrado evidencias previas de la lepra. Las lesiones se curaron completamente por tratamiento del paciente con PQT de corta duración (12 meses) acompañada de la administración de drogas antiinflamatorias no esteroidales y corticoesteroides.

Leprosy is a major health problem in India with a prevalence of 3.22/10,000, affecting all age groups from infancy to old age ([3]). Children below 15 years of age constitute about 15% of total cases of leprosy ([2]). However, it has generally been observed that in childhood, indeterminate leprosy is the most common type, followed by tuberculoid variant; borderline lepromatous and lepromatous leprosy are only occasionally encountered ([18]). Various published studies have consistently described reactions especially type 2 to be less common in children than in adults ([4, 5, 8, 9, 12, 13, 21]).

We describe a 9-year-old boy who presented primarily with erythema nodosum leprosum necroticans (ENL) and was subsequently diagnosed as having lepromatous leprosy.

Case report. A 9-year-old boy presented to dermatology outpatients with the complaint of multiple red, raised, painful lesions over the face and limbs of 4 months duration. These grouped lesions started from the chin, followed by similar eruptions over his upper and lower limbs, and subsided with scaling and bruise-like pigmentation over the next 2 to 3 weeks. However, few lesions had ruptured to discharge pus. He also had episodic fever with each crop of lesions. There was no evidence of any systemic focus of infection on history.

There was no history of light colored or

[1] Received for publication on 29 September 2004. Accepted for publication on 31 January 2005.

[2] D. Pandhi, M.D.; S. Mehta, M.D.; S. Agrawal, M.D.; A. Singal, M.D., Department of Dermatology and STD, University College of Medical Sciences and Guru, Teg Bahadur Hospital, New Delhi-110095, India.

Reprint requests to: Dr. Archana Singhal, B-16/Fi, Dilshad Garden, Delhi–11095, India.

E-mail: archanasinghal@rediffmail.com

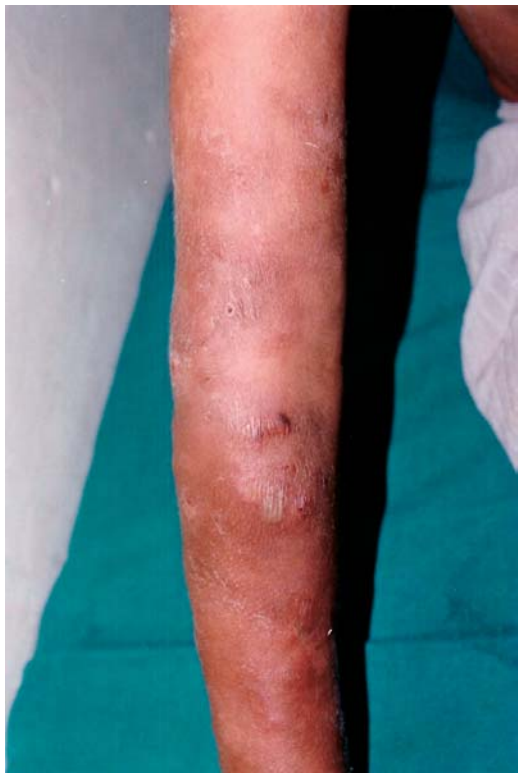

FIG. 1. Hyperpigmented infiltrated plaques with necrosis present on the extensor aspect of right upper limb.

numb lesions, spontaneous blistering or weakness in any limb. He did not have ocular, nasal, testicular, joint, or any other systemic complaints. None of the family member had leprosy, tuberculosis, or any chronic ailment.

The patient weighed 22 kg and was febrile (101°F), with multiple discrete and large submandibular lymph nodes about 0.5 to 1 cm in size, non-tender and not attached to the overlying skin. On cutaneous examination there was evidence of facial infiltration along with multiple erythematous tender nodules and plaques varying from 0.7 cm to 2 cm were seen over the cheeks, chin, and right earlobe, extensor aspect of upper and lower limbs, lower back, and buttocks (Figs. 1 and 2). Few lesions had overlying irregular ulcers with necrotic base (Fig. 2). Multiple hyperpigmented bluish macules with well to ill-defined margins were also noted on the bilateral upper and lower limbs. Bilateral ulnar, lateral popliteal, and posterior tibial nerves were symmetrically thickened and non-tender.

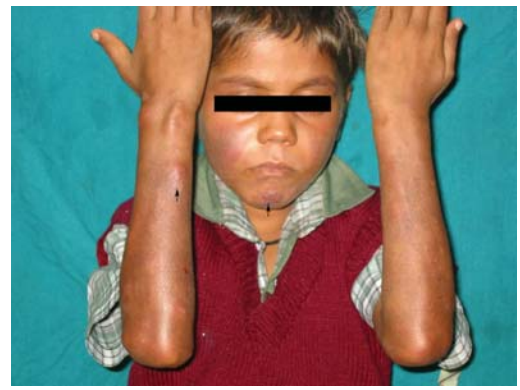

FIG. 2. Multiple erythematous to hyperpigmented nodular swellings on the face and extensor aspect of bilateral arms with few of them showing central necrosis.

There was no evidence of specific glove and stocking type of sensory loss, or any motor deficit. Systemic examination did not reveal any abnormality.

Hemogram revealed mild anemia (Hb = 10.5 gm%), neutrophilic leukocytosis (TLC = 16,000, DLC = $P_{70}L_{25}M_3E_2$), and raised erythrocyte sedimentation rate (ESR = 81 mm in the first hour). The antistreptolysin titer was less than 200 and the reports of throat swab and mantoux test were negative. Tests for activated partial thromboplastin time (aPTT), kaolin clotting time (KCT), LE cell and rheumatoid factor were negative. Other laboratory investigations including liver and kidney function test, blood sugar, urine examination, and chest x-ray were within normal limits. Slit skin smear revealed a bacteriological index of 5+ along with a morphological index of 10%. Routine histopathology section stained with hematoxylin and eosin from the nodule on the left arm demonstrated neutrophilic abscesses superimposed on a diffuse infiltrate of foamy macrophages and plasma cells, along with the evidence of leucocytoclastic vasculitis, which validated the diagnosis of lepromatous leprosy with ENL (Figs. 3 and 4). Fite's stain showed clumps of acid-fast bacilli (AFB) in the perineural cells and macrophages.

He was started on World Health Organization (W.H.O.) recommended multibacillary (MB) multi-drug therapy (M.D.T.), which included 50 mg of dapsone daily and 300 mg of rifampicin monthly along with 50 mg of clofazimine daily. The patient was

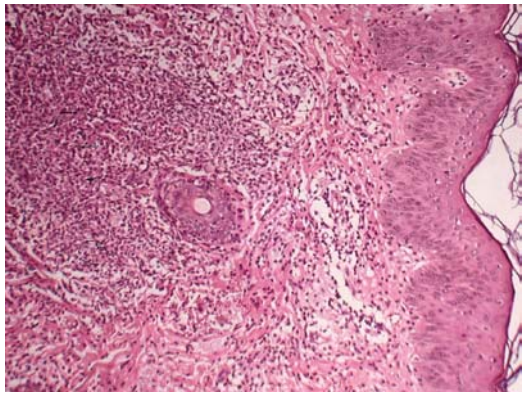

FIG. 3. Routine histopathological stained section from nodular lesion on left arm showing a diffuse infiltrate of foamy macrophages along with endothelial cell swelling, erythrocyte extravasation and polymorphonuclear infiltration (H&E ×200).

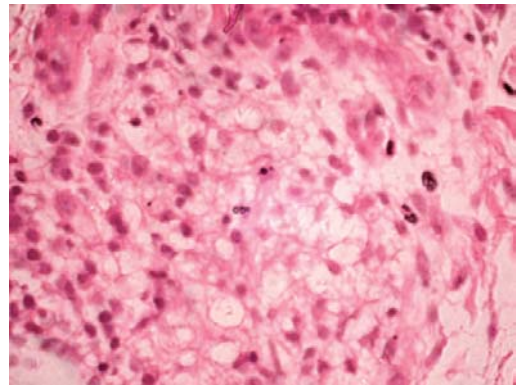

FIG. 4. Routine histopathological stained section from nodular lesion on left arm showing a diffuse infiltrate of foamy macrophages along with polymorphonuclear infiltration (H&E ×1000).

also started on diclofenac sodium 25 mg thrice daily along with 30 mg of prednisolone. The lesions subsided in 6 to 8 weeks time, with no evidence of recurrence or subsequent neural deficit on follow-up. Prednisolone was gradually tapered over a period of 6 months and diclofenac sodium was continued for another 2 months Currently, he has received MB M.D.T. for 11 months and there is no significant reduction in the nerve enlargement.

DISCUSSION

Children in leprosy endemic areas are exposed to infection by *Mycobacterium leprae*. The youngest age reported for the occurrence of leprosy is about 3 weeks ([15]). A 1978, a survey among school children in Hyderabad showed the prevalence rate to be 10 to 17 per 1000 ([8]).

Majority of the pre-pubertal children tend to have indeterminate or tuberculoid type of leprosy ([18]). The children present with asymptomatic or hypoaesthetic cutaneous lesions and, less often, with neural manifestations. A single lesion present on exposed areas of the body was reported to be more common than multiple lesions of the body. Leprosy workers have been conservative in making a diagnosis of multibacilliary or borderline leprosy in children, despite the fact that all types of leprosy can occur in any age group ([18]). The reported rate of smear positive leprosy is less than 10% in pediatric age group ([16]). This appears to be a paradoxical situation and runs counter to the concept that immune responses are poorly developed in the very young children, due to the inability of the immature lymphoid system to deal with and react with foreign antigens ([6, 19]).

Despite the high prevalence of leprosy in children, the occurrence of both type 1 and type 2 reaction especially ENL is rare as revealed by an extensive MEDLINE search. ENL has been reported in 0 to 3.1% of all the cases of childhood leprosy, which is much less than the reported incidence of 35% among all the age groups ([21]) (The Table). The largest study comprising 1028 leprosy patients has reported 25 cases of pediatric ENL, out of which only 7 patients were below 10 years of age ([5]). ENL necroticans, however, has not been reported in pediatric age group to date.

THE TABLE. *Studies showing incidence of erythema nodosum leprosum in pediatric age group.*

S No.	Author	Year	No. of patients	ENL
1	I. Kaur, *et al*. (12)	1991	132	1
2	V. N. Sehgal, *et al*. (19)	1993	161	Not specified
3	P. V. Prasad (17)	1998	66	Nil
4	A. Selvesekar, *et al*. (1)	1999	794	1
5	X. S. Chen, *et al*. (5)	2000	1028	25
6	H. C. Leu, *et al*. (13)	2000		Nil
7	S. Jain (8)	2002	306	5
8	K. D. Burman, *et al*. (4)	2003	20	Not specified

Hypersenstivity responses are known to play an important role in the occurrence of reactions. Reactional episodes and disability are less frequently seen in the younger children, due to the poorly developed immunological mechanisms in early childhood ([6]). Therefore, development of disabilities as a consequence of reactions is, more often encountered in adults and in older children ([5, 19]).

Jain, *et al*. reported the incidence of neuritis to be 24.2% in the cohort of pediatric leprosy patients, and emphasized the importance of appropriate use of steroids to prevent deformities ([8]). Surprisingly, none of these studies on childhood leprosy have described the use of systemic steroids in reactions, with the exception of the study conducted by Thirugananam, *et al*., who reported one child who developed primary focus of tuberculosis after having received one and a half years steroid therapy ([5, 22, 23]). Digeorge, *et al*. have described numerous side effects of systemic corticosteroids in pediatric conditions, with a special mention of HPA axis suppression, growth failure and retarded bone formation, especially when they are used over a longer period of time ([7]). Thus, though we should be aggressive in using steroids in reactions to prevent sequele, the potential serious adverse effects should be screened for.

Lucio's phenomena is characterized by the development of painful, tender, and purpuric macules, particularly on the extremities, with necrotic center which finally develop a black eschar which heals in a few days to leave superficial atrophic scars ([10]). This phenomena is typically seen in patients with Lucio's leprosy, which usually manifests as shiny thickened skin with loss of body hair (but not scalp hair) and widespread sensory loss. Unlike polar lepromatous (LL) there are no skin or ocular lesions, motor palsies, and finger contractions ([11]). Though this phenomena is characterized histopathologically by leucocytoclastic vasculitis, as seen in the present case, the usual clinical presentation of Lucio's leprosy was not observed and thus, the possibility was not considered.

Family contacts with leprosy in the pediatric age group are documented to be a significant factor in contracting the disease ([8]). Though no such history was found in the present case, the importance of contact screening strategies cannot be undermined.

(i) The various factors associated with the prevalence of ENL include older age group, bacillary index of more than 4, multiple enlarged nerves (>5), the presence of nodules and infiltration, more than 1 year of untreated disease and the presence of the anti-PGL-1 antibodies ([14]). Our patient had facial infiltration, high bacillary index, and multiple thickened nerves though duration of the disease could not be ascertained.

(ii) In the present case, history of fever with the eruption of ENL necroticans lesions along with thickened nerves pointed towards a diagnosis of lepromatous leprosy. Characteristic histopathology, demonstration of acid-fast bacilli and response to therapy further confirmed the diagnosis and excluded other possible causes of erythema nodosum.

(iii) To the best of our knowledge, this report of a nine-year-old boy describes the first case of necrotic ENL along with lepromatous leprosy in a child.

REFERENCES

1. Anonymous. Ministry of Health & Family, Welfare Govt. of India (1982) *Annual Report* 1981–82, New Delhi.
2. Anonymous. Update on leprosy elimination in SEAR (November 2003). Regional Health Forum **7(2)**.
3. Burman, K. D., Ryall, A., Agarwal, S., Agarwalla, A. Childhood leprosy in eastern Nepal: A hospital based study. Indian J. Lepr. **75** (2003) 47–52.
4. Chenm X, S,, Wen Zhong Li, Chang Jiang, and Gan-Yun Yi. Leprosy in children: a retrospective study in China. J. Trop. Pediatric. **46** (2000): 207–211.
5. Cruishank, R., Duguid, J. P., Marmian, B. P., and Swain, R. H. A. Natural and acquired immunity. In: *Medical Microbiology*, 2nd edn. FLAS Ed, 1973. p. 134.
6. Digeorge, A. M., and Levine, L. S. Cushing syndrome. In: *Nelson's Textbook of Pediatrics*, 15th edn. Bangalore: Prism books pvt. Ltd., 1996. pp. 1623–1624.
7. Jain, S., Reddy, R. G., Normani, S., Lockwood, D. N. J. Childhood leprosy in an urban clinic, Hyderabad, India; clinical presentation and the role of household contacts. Lepr. Rev. **73** (2002) 248–253
8. Jayam, S., Sekhar, K., Ramasekhar, K., and Ganapathi, R. Childhood leprosy: a study in an urban slum. Indian Pediatr. **15** (1978) 375–377.

9. JOPLING, W. H., and MCDOUGALL, A. C. Leprosy reactions. In: *Handbook of Leprosy*, 5th edn. New Delhi: CBS publishers, 1996. pp. 82–91.
10. JOPLING, W. H., and MCDOUGALL, A. C. The disease. In: *Handbook of Leprosy*, 5th edn. New Delhi: CBS publishers, 1996. pp. 10–53.
11. KAUR, I., KAUR, S., SHARMA, V. K., and KUMAR, B. Childhood leprosy in Northern India. Pediatric. Dermatol. **8** (1991) 21–24.
12. LEU, H. C., CHEN, J. C., CHAO, J. Y., CHAO, P., ET AL. Epidemiology of leprosy in Taiwan; its pattern in children. Int. J. Lepr. Other Mycobact. Dis. **68** (2000) 57–62.
13. MANANDHAR, R., LE MASTER, J. W., and ROCHE, P. W. Risk factors for ENL. Int. J. Lepr. Other Mycobact. Dis. **67** (1999) 270–278.
14. NOORDEN, S. K. The epidemiology of leprosy. In: *Leprosy*, 2nd edn. Edinburgh: Churchill Livingstone, 1994. pp. 29–48.
15. NOUSSITOU, F. M., and SANSARRICA, H. *Leprosy in Children*. Geneva: World Health Organization, 1976. pp. 11–28.
16. PRASAD, P. V. Childhood leprosy in an endemic area. Indian J. Pediatrics. **65** (1997) 751–754.
17. SAMUEL, N. M., SAMUEL, S., and ABIGA, R. B. Borderline leprosy in a 3-year-old child. Indian J. Lepr. **57** (1985) 628–631.
18. SEHGAL, V. N., and CHAUDHARY, A. Leprosy in children: a prospective study. Int. J. Dermatol. **32** (1993) 194–197.
19. SEHGAL, V. N., REGE, V. L., MASCARENHAS, M. F., AND REYS, M. The prevalence and pattern of leprosy in a school survey. Int. J. Lepr. Other Mycobact. Dis. **45** (1977) 360–363.
20. SELVESKAR, A., GEETHA, J., NISHA, K., and MANIMOZHI, N. Childhood leprosy in an endemic area. **70** (1999) 21–27.
21. SHARMA, P., KAR, H. K., MISRA, R. S., MUKHERJEE, A., KAUR, H., MUKHERJEE, R., AND RANI, R. Reactional states and neuritis in multibacilliary leprosy patients following M.D.T. with/without immunotherapy with *Mycobacterium w* anti-leprosy vaccine. Lepr. Rev. **71** (2000) 193–205.
22. THIRUGANANAM, T., and RAJAN, M. A. Borderline reactions treated with clofazimine and corticosteroids. Indian J. Lepr. **57** (1985) 164–171.