Ind J Dermatol Venereol Leprol 1994; 60 : 366-8

LUPUS ERYTHEMATOSUS PANNICULITIS

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An infant presented with thick erythematous plaques having central necrosis, situated over cheeks and wrists. On the basis of clinical features and histology, diagnosis of lupus erythematosus panniculitis (LEP) was made. Lesions responded well to systemic steroids. To our knowledge, cases of LEP in this age group have not been reported earlier.

Key words : Lupus erythematosus panniculitis (LEP), Infancy

Introduction

Lupus erythematosus panniculitis (LEP) is a rare disease. It is characterized clinically by deep subcutaneous nodules or plaques, with central erythematous and atrophic features, distributed on the face, scalp, proximal extremities, trunk or lower back.[1,2] It is to be differentiated from panniculitis due to other connective tissue diseases, like dermatomyositis and scleroderma and from Weber-Christian disease, protease inhibitor deficiencies, erythema nodosum, lipodystrophies and sarcoidosis.[1] Most of the reported patients are middle aged but some childhood cases (7-18 years) have been recorded previously.[1] We report a case of LEP in an infant aged 11 months.

Case Report

An 11 months male infant presented with history of erythematous nodular lesions on both cheeks, of three months duration, which ulcerated within a week and healed with scar formation within a month. Before two weeks he developed another erythematous plaque on the right forearm which also ulcerated within a week. Simultaneously he developed few maculopapular erythematous and purpuric lesions on both legs. He had moderate fever and cough for last one week. The infant was perfectly normal at birth and it was product of a full term normal delivery. There was no history of any previous significant illness. No history of similar disease in parents or any of the siblings was recorded. At the time of admission the patient had a well demarcated plaque, about 5 x 5 cm, on the dorsum of the right wrist. The margins were thickened, infiltrated and raised with marked telangiectasis. The central portion was depressed, atrophic and necrosed, with dark reddish brown colour. The consistency of the central part was firm and the lesion was slightly tender. It was not attached to the underlying muscles. There were two rounded depressed atrophic scars, 2 x 2 cm, on each cheek, with slightly raised erythematous margins. The patient's temperature was 39°C and pulse 100/min. General examination did not reveal any abnormality.

Results of laboratory studies were as follows : White blood cell count, 20,900/cu mm with neutrophils 45%, lymphocytes 54% and eosinophils 1%, platelets 400,000/cu mm, haemoglobin 11.9 gm%; ESR 80 mm/hr; total serum protein 8.5 g/dL, with albumin 53%, α_1 globulin 2.7%, α_2 globulin 20.6%, β globulin 13.6% and γ globulin 10%; Serum electrolytes - Na 139 meq/L, K 5.4 meq/L, and Cl 100 meq/L; urea 26mg/dL, GPT 21

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u/L, alkaline phosphatase 71 u/L, bilirubin 0.2 mg/dL, blood sugar 73 mg/dL. Urine and stool examination did not reveal any abnormality. Blood for L E cell phenomenon was done twice and found to be negative. Throat swab did not reveal any pathogenic organism. Mantoux test was negative. X-ray chest revealed changes of mild pneumonitis on the left base. Antinuclear factor and other immunologic studies were not available.

The biopsy specimen taken from the infiltrated margin showed slight hyperkeratosis and acanthosis with liquifaction degeneration of basal cells at some places. There was dense inflammatory infiltrate in the dermis, consisting mainly of lymphocytes. Similar infiltration was noticed in the subcutaneous tissue, specially around the blood vessels, with thickening of the blood vessel walls.

Clinical examination and laboratory tests did not reveal any evidence of SLE in the mother of the patient.

The patient was given oral cephaloridine for 7 days, fever subsided but there was no improvement in the skin lesions. Then oral prednisolone 10 mg/day was started and significant improvement was noticed. After three weeks the dose was reduced to 5 mg/day and the lesions completely healed with atrophy within next three weeks. Two months after stopping the oral steroid therapy, the patient presented with a new lesion on left wrist with similar clinical features but not accompanied with fever. He was again given prednisolone in the same dosage and the lesion healed with atrophy within four weeks. The patient is lost to follow up for the last two months.

Comments

LEP is a distinct subset of lupus erythematosus. It is a chronic recurrent inflammatory process which produces tissue necrosis and degeneration greater than that usually seen in discoid LE and SLE. LEP may occur alone prior to the onset or after years of either type of LE. The patient may have periodic flare-up of panniculitis or have long remissions with no active disease. Commonly the lesions are large painful necrotic, infiltrating, inflammatory, persistent subcutaneous masses or ulcers.[2]

Histopathologic studies in previously reported cases showed changes of discoid LE in 20% and that of poikiloderma in 25%.[3] There was liquifaction degeneration of basal layer with lymphocytic infiltration of upper dermis and focal lymphocytic panniculitis, the first one having more diagnostic value.[4] Lymphoid nodules with germinal centers were also seen in fat or dermis in 55% of the patients.[2] Secondary hyaline degeneration of basement membrane, blood vessels and adipose tissue was characteristic.[2] In our case, there was liquifaction degenration of basal cells, lymphocytic infiltration in the dermis and subcutaneous tissue with thickening of vessel walls, suggestive of LEP.

Immunofluorescent studies have shown deposits of IgM at the basement membrane in most of the reported cases and that of C3 in some patients.[3] We could not perform this study because of non-availability.

Our patient responded well to systemic steroid therapy. Chloroquine therapy was not tried because of the young age of the patient. A case with exactly similar features has been reported in a seven years old girl, who responded well to chloroquine.[1] LEP affects mainly middle age group but childhood cases under IO years[1,5,6] and between the ages of 10 and 18 years[3,5,8] have been reported. In our case the onset of the disease was at the age of 8 months. To our knowledge, there is no

previous report of LEP in infancy. Tuffanelli[9] reported LEP in twin sisters and another case with familial SLE panniculitis. Winkelmann and Peters[3] reported a case with history of SLE in mother and maternal first cousin. We could not reveal any family history of LEP of SLE in our case.

References

1. Taieb A, Hehunstre JP, Goetz J et al. Lupus erythematosus panniculitis with partial genetic deficiency of C2 and C4 in a child. Arch Dermatol 1986; 122: 576-82.
2. Winkelmann RK. Panniculitis in connective tissue disease. Arch Dermatol 1983; 119: 336-44.
3. Winkelmann RK, Peters MS. Lupus panniculitis. Dermatol update 1982; pp 135-52.
4. Sanchez NP, Peters MS, Winkelmann RK. The histopathology of lupus erythematosus panniculitis. J Am Acad Dermatol 1981; 5: 673-80.
5. Schnitzler L, Verret JL, Schubert B, et al. Le lupus erythemateux profond, Interest de l'etude ultrastructurale. Ann Dermatol Venereol 1978; 105: 153-61.
6. Koransky JS, Esterly NB. Lupus panniculitis. J Pediatr 1981; 98: 241-4.
7. Cerimele D. Contributo allo studio del lupus eritematoso profondo. Chron Dermatol 1970; 1: 295-312.
8. Winkelmann RK, Padilha-Concalves A. Connective tissue panniculitis. Arch Dermatol 1980; 116: 291-4.
9. Tuffanelli DL. Lupus erythematosus panniculitis (profundus): clinical and immunologic studies. Arch Dermatol 1971; 103: 231-42.