

Atypical Papulonecrotic Skin Manifestation by Lepromatous Leprosy

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Leprosy is a multisystemic infectious disease showing various cutaneous manifestations by the reaction between *Mycobacterium leprae* (*M. leprae*) and host immunity. We describe a 20-year-old woman with clinical multiple papulonecrotic skin eruptions resembling papulonecrotic tuberculid (PNT) and pityriasis lichenoides et varioliformis acuta (PLEVA). This kind of skin eruption is a new manifestation which has not been reported so far in lepromatous leprosy (LL). (Ann Dermatol 9:(2) 143~146, 1997)

Key Words : Lepromatous leprosy, Papulonecrotic skin eruptions

Leprosy is a chronic systemic infectious disease caused by *M. leprae* showing various clinical manifestations. The cutaneous lesions of LL are characterized by ill-defined macules, plaques and nodules. We experienced a new clinical manifestation of LL, showing multiple papulonecrotic lesions with crusts and varioliform scars, resembling PNT or PLEVA.

REPORT OF A CASE

A 20-year-old woman had multiple scattered papulonodular lesions with mild intermittent pruritus for 1 year. The lesion began as ill-defined erythematous to brownish macules on the upper and lower extremities, to which she did not pay serious attention. Since then there had been a gradual increase in the number of papulonodules. Several months later, papulonecrotic lesions were noticed in-

cidentally and were accompanied by intermittent mild pruritus, especially at nighttime. Although she was treated with topical steroids at private clinics, the necrotic lesions were persistent without recurrence, and did not show any improvement. In her family history, her father had a leprosy which had been treated since her birth. He died 1 year before her visit to our clinic. The patient's mother and sister were healthy.

Examination revealed erythematous to brownish rice- to pea-sized papules and infiltrative nodules on the face, buttock, and upper and lower extremities (Fig. 1). Some lesions of the face, lower back, buttock, and upper thigh showed necrotic papules covered with hemorrhagic crusts, leaving varioliform scars (Fig. 1, arrow). There was no sensory change.

Laboratory tests including complete a blood cell count, erythrocyte sedimentation rate, levels of electrolytes, blood urea nitrogen, creatinine, anti-nuclear antibody titers, rheumatoid factor, prothrombin and partial thromboplastin times, liver enzyme values, and urinalysis were all normal or negative. Chest radiography and electrocardiogram were normal.

A skin biopsy obtained from the nodule of the upper thigh revealed diffuse infiltration of lymphocytes, histiocytes, foamy macrophages, and eosinophils in the dermis. The specimen taken

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Fig. 1. Numerous erythematous to pigmented papulonodules on the lower extremities (A) and right buttock (B). Arrows indicate the papulonecrotic lesions among them.

from the necrotic papular lesion revealed hyperkeratosis, spongiosis, necrosis in the epidermis, and a mixed cellular infiltration of lymphocytes, neutrophils, and foamy histiocytes in the dermis (Fig. 2A). Also, vascular changes including mild perivascular infiltration of inflammatory cells, swelling of endothelial cells and narrowing of the vascular lumina were observed in the dermis (Fig. 2B). Fite staining showed innumerable acid-fast, red staining lepra bacilli in the entire dermis, especially vascular lumina (Fig. 3), and the epidermis.

Fig. 2. (A) Histological studies show the focal necrosis of the epidermis, diffuse infiltrate of inflammatory cells on the upper dermis. (B) The dermis shows a diffuse infiltrate of foamy histiocytes and vascular changes including perivascular infiltration of inflammatory cells and endothelial swelling (H & E $\times 200$).

A PCR study with a paraffin-embedded skin sample by using specific primers of *M. leprae* showed a specific positive band for leprosy. The Lepromin and Mantoux tests were negative. The Histamine test did not show a flare response in the lesional skin. Direct immunofluorescent studies did not show deposition of immunoglobulin and complement around the blood vessels.

Combination treatment with anti-leprotic drugs including dapsone, clofazimine, and rifampicin showed rapid remittance of the papulonodular and

Fig. 3. Fite stain reveals the presence of innumerable acid-fast, red staining bacilli in the vascular lumina and the dermis ($\times 1,000$).

papulonecrotic lesions from 2 months after treatment, and left dark pigmented, slightly atrophic scars at about 5 months. Fite staining of biopsy samples from the previous lesions revealed diminution of the bacilli. There was no recurrence of papulonecrotic eruptions for the next 1-year of follow-up.

DISCUSSION

There is no other infectious dermatoses as leprosy, in which the clinical picture is so complicated as to cause difficulty in diagnosis. It was assumed that the clinical manifestations resulted from the host response to the presence of *M. leprae*, causing classification into two polar types, the tuberculoid (TT) type of high host resistance, and the lepromatous (LL) type of low host resistance¹. Usually the onset of leprosy is insidious, and the disease is not noticed until the appearance of cutaneous eruptions. The most common cutaneous eruption is an ill-defined hypopigmented macule of indeterminate leprosy, which evolve into different clinical patterns by the immunological state of the host². The clinical features of LL consist of wide spread, symmetrical, ill-defined macules, papulonodules and plaques. There are several unusual expressions of multibacillary leprosy that may cause confusion in diagnosis and pose therapeutic problems; localized lepromatous or borderline lepromatous disease showing a single nodule or a localized area of nodules or papules; histoid leprosy showing firm shiny papules or nodules; spontaneous skin ulceration in patients

with severe, long-standing, untreated LL; Lucio leprosy showing diffuse shiny infiltration³. Hyperkeratotic and verrucous skin lesions were rarely reported in India⁴. But papulonecrotic lesions observed in the present case have not been reported so far from our review of dermatological literature. Clinically, both PNT or PLEVA show multiple necrotic papules with resulting scarring; PNT shows a symmetrical acral distribution, while PLEVA shows a more widespread distribution. PNT is regarded as an Arthus reaction followed by a delayed hypersensitivity response to mycobacteria, which may be responsible for the vascular damage resulting in a subacute or chronic vasculitis⁵. Iden et al⁶ reported that many cases of Takayasu's arteritis were associated with tuberculosis, suggesting the close relationship between tuberculosis infection and vasculitis. Wilson-Jones et al⁷ regarded PNT as a distinctive form of immunological host response to hematogenous bacterial antigens. A similar mechanism was known to be responsible for inducing septic vasculitis in meningococcal and gonococcal septicemia, and *Pseudomonas* infection. The etiology of PLEVA was unknown. Some regarded it as an immune reaction, such as a hypersensitivity to microorganisms of viral and bacterial infections⁸⁻¹¹, while some regarded it as a lymphoproliferative process like lymphomatoid papulosis^{12,13}. The relationship between PNT or PLEVA and infectious organisms suggest that papulonecrotic eruptions of our case may be associated with lepra bacilli. Interestingly, necrotic papules of the present case showed infiltration of acid-fast bacilli within the vascular lumina in the Fite stain. Thus, it is presumed that infiltration of *M. leprae* into vessels ensue vascular changes with skin eruptions resembling PNT or PLEVA.

The possibility of the coincidental presence of PNT or PLEVA could be ruled-out by the following findings; papulonecrotic lesions showed infiltration of the lepra bacilli and anti-leprosy medication resulted in complete resolution of the lesions. Histological findings were commensurate with the clinical findings, but characteristic findings of PLEVA, such as exocytosis of lymphocytes and red cells were not found in this case.

Vascular changes in LL include vasculitis of erythema nodosum leprosum and Lucio's phenomenon¹⁴⁻¹⁷ and granulomatous reaction by colonization of bacilli in the blood vessels^{18,19}. However

there were no reports to show that necrotic variform papules in LL is caused by leprosy infiltration

In conclusion, leprosy should be considered as a differential diagnosis of papulonecrotic skin lesions, suggestive of PNT or PLEVA.

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