

Necrotizing Reaction in Lepromatous Leprosy*

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Classification of the lepra reaction is reviewed. The cutaneous eruptions in the form of vesicles, pustules, or necrosis are indicative of a severe form of the reaction. A case is reported in a woman who presented, for the first time, a lepra reaction. She had systemic symptoms, and cutaneous and mucous membrane lesions leading to palatal perforation. The rarity of lesions of this type and the difficulty in their categorization into recognized types of lepra reaction are discussed.

Reactions in leprosy are the greatest obstacles to the cure of the patient, retarding recovery in the most favourable cases and in others often bringing about death prematurely. Opinions differ as regards the definition of this condition. The panel on reaction at the 8th International Congress of Leprologists held in Rio de Janeiro in September, 1963, defined it as "An acute or subacute clinicopathological syndrome which appears during the chronic course of lepromatous leprosy with systemic symptoms and local lesions in the skin and other organs". To this, Ridley (1969) added "the histological disturbance which occurs during reactions is not associated with either the activity or the regression of the leprous granuloma".

There has so far been no generally acceptable classification of lepra reaction. Wolcott (1947) and Jopling (1959) defined the difference between "lepra reaction" and *erythema nodosum leprosum* in terms of mild and moderate forms of lepromatous reaction. The panel dealing with reaction at the Rio congress used the terms "leprosy-exacerbation" (which was the old lepra reaction) and "lepra reaction" (which was the old *erythema nodosum leprosum*). As Dharmendra subsequently remarked: "They made confusion worse confounded". Ridley (1969) proposed a classification of lepra reaction into down-grading reactions, reversal reactions, exacerbation nodules, and *erythema nodosum leprosum*.

Necrotic skin lesions appear rarely in lepra reaction. Such reactions when they occurred were variously attributed to Lucio's phenomenon (Fernandez *et al.*, 1962), cutaneous allergic vasculitis, bullous reaction (Job and Gault, 1960), a form of *erythema nodosum leprosum* (Canizares, Costello and Gigli, 1960) and lepra reaction modified by keratosis blenorrhagica (Henry, 1963).

In this paper a case is described which demonstrated unusual cutaneous, mucosal, and systemic expression of the lepra reaction. The patient, a 26-year-old married Moslem female, was admitted to the local University Hospital in August,

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1971. She had been having recurrent pustular eruptions and ulcerations all over her body for the previous 5 years. Weakness, weight loss, and intermittent high fever accompanied by pain and swelling of the joints had been present for the last 3 years. Perforation of the palate, leading to nasal regurgitation, had occurred 10 months before admission to hospital. She also complained of loss of sensation in the hands and feet.

In 1966, a few "boils", like eruptions, appeared on her face, but these subsequently healed with treatment. After 6 months, periodic crops of small painful nodules began to appear on her extremities, chest and back. In 1968 she consulted local physicians because of high fever which was accompanied by swelling of the knees, ankles, and elbow joints; treatment resulted in some alleviation of her symptoms. Two years later she experienced an episode characterized by high fever, chills, myalgia, recurrence of pain and swelling over the joints, and painful erythematous nodules and pustules. Finally, 10 months after the above episode, i.e. in August, 1971, she was admitted to our hospital with nasal regurgitation and necrotic ulcerative lesions of her extremities.

Physical examination showed a febrile, toxic and cachectic woman in moderate distress. There were many crusted lesions, the crusts being bloody and loosely attached. There were also superficial ulcers and scars on her extremities. Besides these, she had multiple necrotic lesions on her legs, arms, buttocks, shoulders and face (see Fig. 1). Fresh papulo-pustular lesions were present over the skin of the face and arms, her nose was slightly depressed, and there was a perforation in the anterior third of the palate. Her fingers were severely tapered and resembled

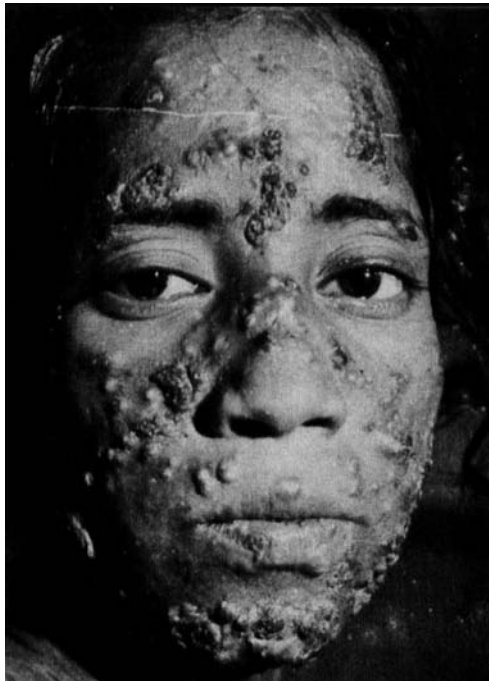


Fig. 1.

"sharpened pencils". Some flattening of the thenar and hypothenar eminences due to muscular wasting was noted. The ulnar nerves were tender and visibly enlarged. Areas of anaesthesia to heat and cold, pain, and light touch were present on her hands and feet. She had minimal pitting oedema on her ankles and there was generalized lymphadenopathy. The liver was palpable two fingers' breadth below the right costal margin.

Blood examination showed a mild leukocytosis and hypochromic microcytic anaemia. The results of urinalysis were normal. Slit examination of skin smears revealed numerous acid-fast bacilli; the smears were taken from both infiltrated and reactive lesions. The biopsy specimen of a reactive skin lesion of 72 hours' duration revealed histopathologically infiltrative lepromatous leprosy with sub-epidermal bulla and intense angitis of blood vessels of all sizes. The latex-fixation test for rheumatoid factor and L.E.-cell tests were negative.

The patient was treated with small doses of dapsone and intramuscular injections of antimony in the form of Fantorin (stibophen, B.P.). The systemic symptoms subsided and the ulcerations began to heal.

Discussion

The cutaneous expression of the lepra reaction is determined by the sub-type of lepromatous leprosy, the location, the degree to which cutaneous vessels are involved, the duration of the reaction, and the degree to which the reaction has been modified by therapy (Jopling, 1959; Waters and Ridley, 1963; Moschella, 1967). Severe reactions characterized by vesicles, pustules and necrosis remain a source of considerable difficulty in categorization (Waters and Ridley, 1963). These do not fully conform to any accepted classification (Ridley, 1969; Moschella, 1967), which is further documented by the present case.

Necrotic skin lesions may be seen in patients manifesting Lucio's phenomenon (*erythema necroticans*), severe *erythema nodosum leprosum* and a lepra reaction simulating the allergic vasculitis of Reuter (Moschella, 1967). The Lucio phenomenon is known to be a rarity outside South and Central America. Perusal of the literature, however, reveals that it has more world-wide distribution than hitherto believed (Moschella, 1967). The cutaneous lesions of the lepra reaction, resembling the cutaneous allergic vasculitis of Reuter, are numerous, superficial and more polymorphous than those of *erythema nodosum leprosum*. There is little justification for classifying the present case on one or the other. Moschella (1967) described a patient who had severe *erythema nodosum leprosum* with necrosis of her reactive skin lesions. Earlier, such patients were reported as having "formes escarrotiques" and *erythema nodosum leprosum*. Furthermore, pustulation and ulceration are known to occur in severe lepra reaction and in "progressive reaction" as defined by Cochrane and Davey (1964). In severe *erythema nodosum leprosum*, suppuration, with or without ulceration, has been reported by many workers including Wolcott (1947), Jopling (1959) and Ridley (1969). To represent such dual manifestations, a new name, *Erythema nodosum necroticans*—was introduced at the Rio Congress.

Fresh involvement of mucous membrane may produce eye and nose symptoms for the first time during the reaction (Dharmendra, 1967). In our case eye symptoms were absent, but the nasal tone in her voice and regurgitation, due to the perforation of the palate, made the patient visit the hospital. Swollen joints and nephritis are less common manifestations of *erythema nodosum leprosum*.

according to Ridley (1969). In our patient, in addition to the skin and mucosal lesions, constitutional symptoms and polyarthritis were the distressing problems.

That leprosy can appear for the first time in the form of the lepra reaction is well documented. In endemic areas, because of the healthy appearance of the patients with lepromatous leprosy, they are usually diagnosed late (Cochrane, 1964). This was also true in the present case.

The use of systemic corticosteroids in the lepra reaction of all types is inadvisable unless the reaction is very severe and uncontrollable (Jopling and Cochrane, 1957). Even though the reaction was very severe in our case, we resorted to antimony treatment in the form of Fantorin (stibophen) injections. This was effective in controlling the reactional state.

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