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Case Report

Misdiagnosis of Lupus Vulgaris and Systemic Sclerosis as Leprosy: Two Case Reports

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Leprosy has wide variations in its clinical presentation. There are some skin diseases which are frequently mistaken for leprosy and is often treated like it for a long period of time. As social stigma is attached to leprosy, a wrong diagnosis of leprosy can put the patient under psychological stress. We present 2 cases of lupus vulgaris and systemic sclerosis respectively who were misdiagnosed as leprosy and treated like it.

Key words : Leprosy, Lupus vulgaris, Systemic sclerosis

Introduction

Leprosy has wide variations in its clinical presentation. There are some skin diseases which are frequently mistaken for leprosy and is often treated like it for a long period of time. The rate of misdiagnosis is high in leprosy endemic countries and especially by health workers in a field situation. As social stigma is attached to leprosy, a wrong diagnosis of leprosy can put the patient under psychological stress and will result in needless treatment. Here we reported two such cases who were misdiagnosed as leprosy and treated like it.

Case 1

A 53 year old female, presented with progressive and asymptomatic non-healing skin lesion over right side of face for one year. She was diagnosed as leprosy from local hospital and was started on WHO MDT-MB and she was taking 12th blister pack. She was upset with treatment as for last one year there was no improvement. On examination a single well defined erythematous plaque of size 8x8 cm was present over right side of face extending from infraorbital rim superiorly to angle of mandible inferiorly, laterally from temporomandibular joint upto ala of nose and

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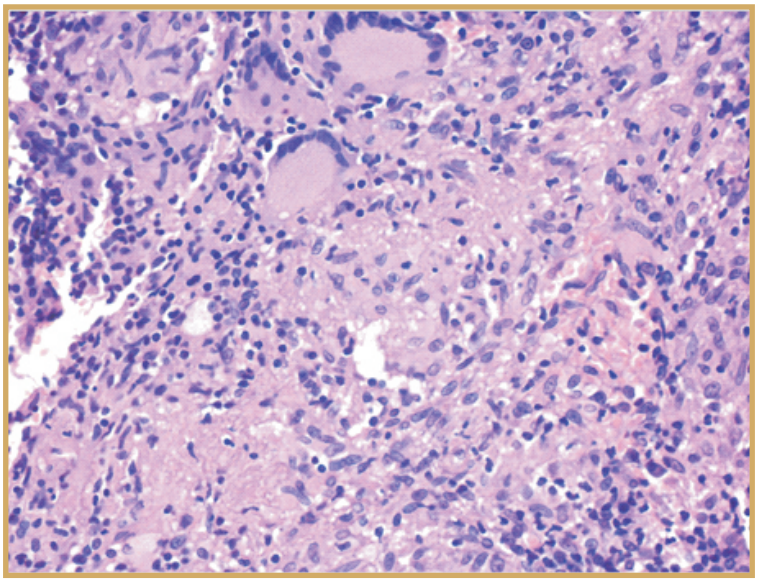

Fig 1 : Multiple confluent granulomas in dermis comprising of epithelioid cells, lymphocyte and Langhan's giant cells with focal area of caseous necrosis. (H and E stain, 40x)

philtrum medially with scarring and crusting over malar region. There was no nerve thickening or regional lymphadenopathy. Systemic examination was essentially normal. Routine hematological and biochemical investigations revealed no abnormalities. Serological test results for human immunodeficiency virus was negative. Chest radiograph didn't reveal any abnormality and purified protein derivative test (Mantoux test) was negative with 4 mm induration after 48-72 h. Slit-skin smears were negative for acidfast bacilli (AFB). Histopathological examination of the punch biopsy specimen showed hyperkeratotic epidermis and multiple confluent granulomas consisting of epithelioid cells, lymphocyte and Langhan's and foreign body giant cells, with focal area of caseous necrosis in the dermis suggestive of lupus vulgaris (Fig. 1). No AFB was seen in histopathology sections by modified Ziehl-Neelsen (ZN) and routine ZN staining. With the diagnosis of lupus vulgaris patient was started on category-1 ATT under revised national TB control programme. Cutaneous lesion started to show regression within 2 months of treatment, thereafter patient was lost to follow up.

Case 2

A 35-year-old male presented with Raynaud's phenomenon, thickening and tightening of skin with progressive sclerodactyly for 3 years. For these complaints he was misdiagnosed as leprosy and had completed one year WHO MB-MDT course. There was no history of cough, dyspnea and dysphagia. Clinical examination revealed typical mask-like face, telangiectasia, microstomia, sclerodactyly, digital ulcerations and pits, hide bound skin on the face, trunk, hands and forearms. Chest auscultation revealed basal crepitations and other systemic examination was normal. Clinically diagnosis of systemic sclerosis was kept. Routine laboratory tests, including

complete blood count, liver, renal and urine analysis were all within normal limits. Slit-skin smears were negative for AFB. His antinuclear antibody (ANA) was 1:1280, but SSc - specific antibodies were negative. Chest radiograph showed reticular opacity in bilateral lower zone. He was advised other investigations like electrocardiogram, barium oesophagogram and pulmonary function tests and was also referred torheumatologist and pulmonologist for opinion.

Discussion

Diagnosis of leprosy is made based on its cardinal signs which are (a) hypopigmented or reddish skin lesion(s) with definite loss of sensation, (b) thickened peripheral nerve(s) with impairment of sensation in the area supplied, (c) AFB in the slitskin smear. Presence of any one of these signs has been regarded as a sufficient ground for diagnosis of leprosy (WHO 1998). Slit-skin smears are not done in the present settings (Kumar and Dogra 2009). In both reported cases there was no impairment of sensation or thickened peripheral nerve. These two cases might has been diagnosed as a case of leprosy on observing erythematous-lesion and sclerodactyly with digital pitted scars and ulcers respectively. Lupus vulgaris mimics leprosy very closely. It begins as asymptomatic, infiltrating papule and plaques. The lesions slowly spread and typical lesion is a well-demarcated, skin-colored or erythematous plaque. Healing and scarring in one area and activity in another is the hallmark of lupus vulgaris as seen in our first case. Lupus vulgaris is not uncommon in India and buttocks and extremities rather than the face are most frequently affected, while in Europe, over 80% of lesions are on the head and neck, particularly around the nose (Sehgal and Waugh 1990, Horwitz 1960). In our first case there was facial presentation which is rare in Indian scenario. Lupus vulgaris is completely curable however delayed diagnosis and treatment can be disastrous for the patients, as the disease can lead to destruction of bones and cartilage leading to permanent deformity. Mandal et al described a similar case of lupus vulgaris who was initially misdiagnosed as leprosy and was initially treated with PB-MDT (Mandal and Bandyopadhyay 2012).

Systemic sclerosis is a multisystem disorder characterized by vascular abnormalities, connective tissue sclerosis and atrophy and auto-antibodies. Acral deformity seen in systemic sclerosis can cause confusion with deformity seen in leprosy. Well-developed cases of systemic sclerosis presenting with Raynaud's phenomenon and typical cutaneous changes on the face and hands are easy to recognize. The condition may have to be distinguished from leprosy if a peripheral neuropathy is present (Lapido 1976). In our second case typical features of systemic sclerosis were present.

Tendency to over diagnose leprosy especially by field workers and general practioner is not uncommon in our country where leprosy is endemic and diagnosis is made in field condition on cardinal signs alone. It is important for health professionals, especially those in leprosy field programmes, to consider a full differential diagnosis in evaluation for leprosy. The accuracy of diagnosis should be accessed through regular supervision. If there is no response with treatment or suspected case of leprosy, patient should be referred to an appropriate center for further examination to prevent misdiagnosis and wrong management.

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