Journal of Pakistan Association of Dermatologists. 2020; **30(4)**: 691-694.

Case Report

Hansen's disease - A diagnostic dilemma

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Abstract Hansen's disease is a chronic granulomatous disease caused by Mycobacterium leprae. It manifests as variety of forms depending upon the immune status of the patient.[1,2] The lesions may mimic various diseases most common being cutaneous tuberculosis, subcutaneous mycosis and sarcoidosis. The neurological examination and microscopy serve as diagnostic skills in leprosy. Lepra bacilli are acid fast with 5% sulphuric acid while tubercular bacilli may resist decolourisation with 20% concentration. In this case report we have described a patient of lepromatous leprosy in whom the bacteria obtained on slit skin smears resisted decolourisation with 20% sulphuric acid in contrast to the usual 5% concentration. Since the staining patterns were consistent with tubercular bacteria, this lead to a diagnostic confusion. Further investigations were adopted which included histopathological examination and Cartridge Based Nucleic Acid Amplification Test (CBNAAT) for tubercular bacilli. Final diagnosis was made upon the histopathological examination and negative CBNAAT.

Key words
Mycobacteria leprae, acid fast bacilli, staining technique.

Introduction

Leprosy or Hansen's disease is a chronic granulomatous disease caused by Mycobacterium leprae (lepra bacilli), contagious in some cases affecting primarily the peripheral nervous system and skin but may involve other organ systems as well. It has a long incubation period of 2-7 years (usually 3-5 years). The disease exhibits a full spectrum with polar forms (multi-bacillary lepromatous leprosy and the pauci-bacillary tuberculoid leprosy) and the intermediate forms with hybrid features of polar forms.[1,2] The variety of lesions ranging from hypoanaesthetic to anaesthetic patches, papules and nodules may be the presentation of the disease making it difficult to diagnose. In such instances neurological examination and slit skin smear may be of paramount value which helps to differentiate it from other strong mimickers of the disease as well as in diagnosing the type of leprosy and thereby planning the treatment type and duration.[3] In this case report it has been observed that lepra bacilli does not always obey the rules of acid fast staining and staining techniques may sometime fail to diagnose a particular disease making it difficult to differentiate it from other Mycobacterial infections. In such circumstances a histopathological examination proves to be the standard tool.

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Case report

A sixty five year old male presented with chief complaints of multiple red raised lesions, larger lesions were studded with pustules and they were present over upper back, abdomen, right forearm and left pinna since one year. Initially he developed few asymptomatic reddish raised

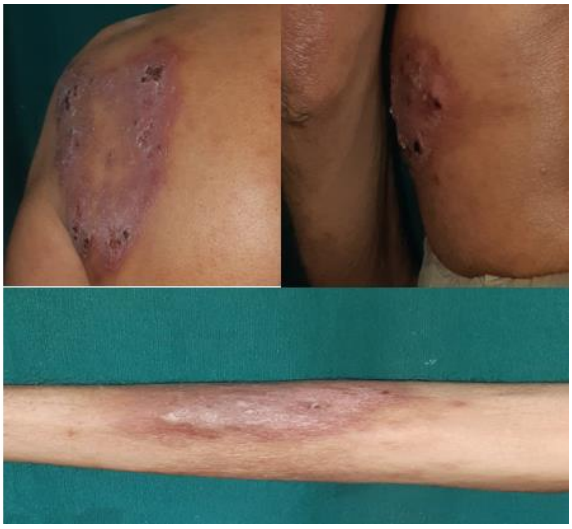

Figure 1 Clinical photographs of the patient depicting the plaques studded with discrete to coalescing pustules.

lesions over right forearm. Over a period of one month, these lesions increased in size and coalesced to form a larger lesion. Similar lesions developed on upper back, abdomen and left pinna over next 6-8 months. Patient gave a history of high grade fever following which he developed pustules in the lesions and some lesions got eroded and were associated with pain. There was no history of grittiness or burning sensation in eyes, nasal stuffiness or bloody discharge, motor or sensory weakness. Patient also denied history of any drug intake, trauma, joint pains, oral ulcerations, spontaneous blistering. There was no history of neuritic or testicular pain. Family history was non-significant. There was no history of chronic illness, either infectious or non- infectious/ metabolic.

General physical examination was within normal limits except for a single non tender discrete palpable lymph node with normal consistency in left inguinal fold. BCG scar was absent. Systemic examination was within normal limits.

On cutaneous examination, lesions were in the form of multiple erythematous papules, succulent plaques and nodules over left pinna, abdomen, upper back and right forearm. Most of the plaques were studded with discrete to coalescing pustules **(Figure1)**. Desquamation was present around a few lesions. All the lesions had mildly raised temperature and tenderness was elicited only on lesion over left pinna. There was a slight loss of temperature sense over the lesions however rest of the sensations were intact.

Neurological examination revealed bilateral grade II thickened non tender ulnar nerves. Rest of the musculoskeletal and neurological examination was within normal limits.

His routine haematological, biochemical investigations, chest x-ray and ultrasound abdomen were within normal limits except for low haemoglobin. Serum ACE (Angiotensin Convertase Enzyme) levels were slightly raised. Fluid from the ulcer margins was subjected to Grams staining and ZN staining and Acid Fast Bacilli (AFB) were resistant to 20% sulphuric acid decolourisation were seen. Slit Skin Smear (SSS) from bilateral ear lobes, lateral margins of eyebrows, buttocks and lesion over forearm and abdomen were positive for AFB at 20% concentration of sulphuric acid. These findings lead to a confusion of Mycobacterium tuberculosis infection. Tissue sample was subjected for CB-NAAT (Cartridge Based Nucleic Acid Amplification Test) was thereby sent for tubercular infection and reports came out to be negative. Elliptical biopsy from lesion over abdomen on histopathological examination revealed attenuated epidermis with preserved Grenz zone. Dermis comprised of granulomas containing chiefly epitheloid cells and macrophages with very few lymphocytes and scattered plasma cells around the blood vessels and adnexal structures suggesting features of lepromatous leprosy **(Figure 2)**. Biopsy for AFB staining revealed multiple globi of bacilli with BI of 6+ **(Figure 3)**.

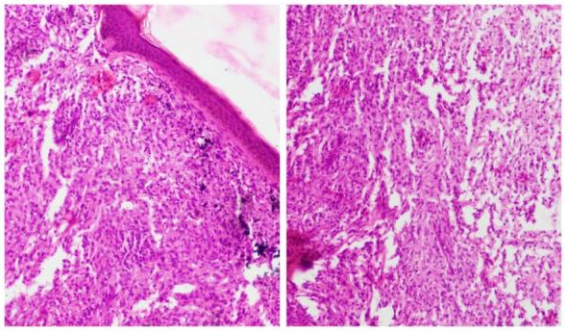

Figure 2 Dermis comprised of curvilinear granulomas containing chiefly epitheloid cells and macrophages with very few lymphocytes and scattered plasma cells around the blood vessels and adnexal structures suggesting features of lepromatous leprosy (haematoxylin and Eosin 40x).

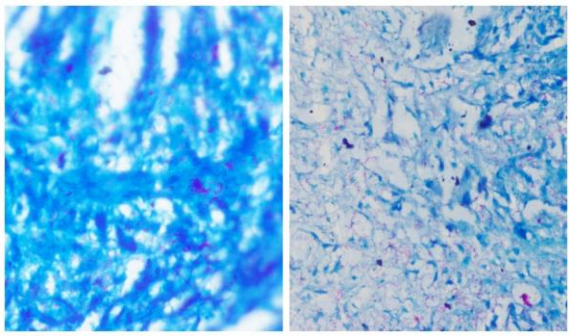

Figure 3 AFB staining revealed multiple globi of bacilli with BI of 6+

Based on investigations, a diagnosis of lepromatous leprosy was made.

Discussion

Leprosy presents with a variety of lesions ranging from macular lesions to ulcers. The presentation depends upon the immune-genetics and current immune status of the patient. Patient may have a single hypo-anaesthetic patch or plaque to multiple ill defined erythematous plaques and nodules which may be ulcerated. The patient may be in reactions depending on course of disease and immune status it may be type 1 reaction (upregulation of immunity) in response to treatment or type 2 reaction (usually downgrading) in lepromatous leprosy during these phases patient may be febrile, lesions may be erythematous, hot, oedematous and tender with other constitutional symptoms.[4,5]

The differential diagnosis includes sarcoidosis, subcutaneous mycosis, cutaneous tuberculosis. In such circumstances neurological examination comes into play. Leprosy is a primary disease of peripheral nervous system affecting Schwann cells, thereby leading to loss of sensations over the affected regions.[6] The peripheral nerves may be enlarged and tender due to ongoing inflammatory process. This usually occurs at the cooler parts since bacilli prefers low temperature for multiplication. Besides clinical examination, microbiological examination of bacteria obtained by slit skin smear examination aids the diagnosis. In SSS bacilli residing deep in dermis are obtained by small slits (incisions) made over the skin. The slides are smeared and are subjected to modified Zeihl Neelson staining which involves decolourisation with 5% sulphuric acid. Lepra bacilli are acid fast and are stained bright red with a bluish background of counter stain. This acid fast nature owes to the mycolic acid present in the cell wall of bacilli.[7] The organisms in Mycobacteria genera are acid fast with tuberculosis bacilli being most acid fast resisting decolourisation at 20% concentration. Since lesions of leprosy may mimic cutaneous lesions of tuberculosis as well, a possibility of lupus vulgaris should be ruled out.[8]

Lupus vulgaris is a cutaneous manifestation of tuberculosis characterised by erythematous papules, plaques which may be annular extending centrifugally with occasional ulcers with undermined edges, centre may be atrophic. The lesions are usually painful but can be painless as well. The smears taken from lesions may reveal acid fast bacilli resistant to 20% sulphuric acid. Histopathological examination is of utmost value in diagnosis which may reveal epidermal reaction and caseating granulomas

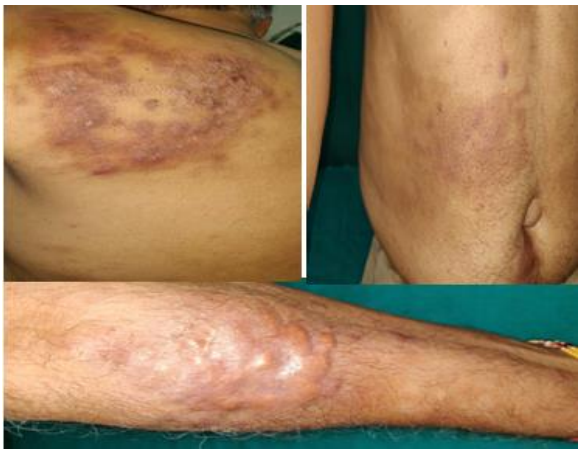

Figure 4 Clinical photographs depicting resolution of lesions following treatment with MBMDT.

which forms the diagnosis of cutaneous tuberculosis.[9]

In our case SSS obtained from bilateral ear lobes, lateral parts of eyebrows, buttocks and lesions over forearm and abdomen yielded acid fast bacilli in all the locations resisting decolourisation with 20% sulphuric acid and so the report consistent with of tubercle bacilli was sent by microbiology department. This lead to a confusion in diagnosis since neurological examination was almost within normal limits so the possibility of cutaneous tuberculosis was to be ruled out. Tissue biopsy for CB-NAAT was thereby sent which came out to be negative for tuberculosis.[10] The final diagnosis was made on the basis of histopathological examination which came out to be consistent with lepromatous leprosy and patient was started on Adult Multibacillary Multidrug Treatment (MBMDT [A]) pack and followed up in leprosy clinics and a positive response to treatment was seen with healing of lesions **(Figure 4)**.

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