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Case Report

Leprosy Plaques Following the Course of A Single Peripheral Nerve: A Rare Presentation

N Parajuli*

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Leprosy or Hansen's disease is a chronic granulomatous disease caused by *Mycobacterium leprae*. The skin lesions can present as few plaques in the paucibacillary type and multiple plaques and nodules in the multibacillary type. A slit skin smear is the most common procedure worldwide to make the diagnosis. Linear presentation is one of the uncommon presentations of leprosy. A case of 17-year-old male with multiple plaques arranged in a linear pattern extending from the left hand up to the elbow is presented in this case report. The presence of anaesthesia over the plaques with biopsy showing epitheloid granulomas proved the diagnosis of Hansen's disease. This case highlights the rare form of leprosy, which might prove to be an obstacle for sustaining leprosy eliminated status due to delay in diagnosis. This case report also suggests the need to explore genetic pattern of inheritance involved in the linear form of leprosy.

Keywords : Blaschko's Lines, Elimination, Hansen's, Leprosy, Linear, Multibacillary

Introduction

Leprosy is a chronic granulomatous infection caused by *Mycobacterium leprae*. It can lead to a spectrum of clinical manifestations ranging from skin lesions to disfigurement and disability. The clinical spectrum of the disease ranges from tuberculoid pole to lepromatous pole based upon the immune response of the patient to the leprosy bacilli (Franco-Paredes & Rodriguez-Morales 2016).

A case of 17-year-old male presenting with multiple plaques arranged linearly over the left upper extremity following the course of a single peripheral nerve and no skin lesion elsewhere is presented below. This case is presented here to highlight a rare presentation of leprosy and the role of tropism of leprosy bacilli to skin and nerve and distinguish it from pure neural forms.

Case Report

A 17-year-old teenager presented with multiple non-pruritic plaques over the left lateral forearm for a duration of one year. The lesion started as a single red plaque on the extensor aspect of the left wrist. Gradually over the course of one year, the patient noted a few other plaques over the extensor aspect of the dorsum of the left hand. Newer plaques followed, which were noted over the middle and upper left forearm. All of these

* Dr Niraj Parajuli, MBBS, MD, Department of Dermatology & Venereology, National Academy of Medical Sciences, Bir Hospital, Kathmandu, Nepal.

Correspondence Author : Dr Niraj Parajuli, **Email** : drnirajparajuli@gmail.com

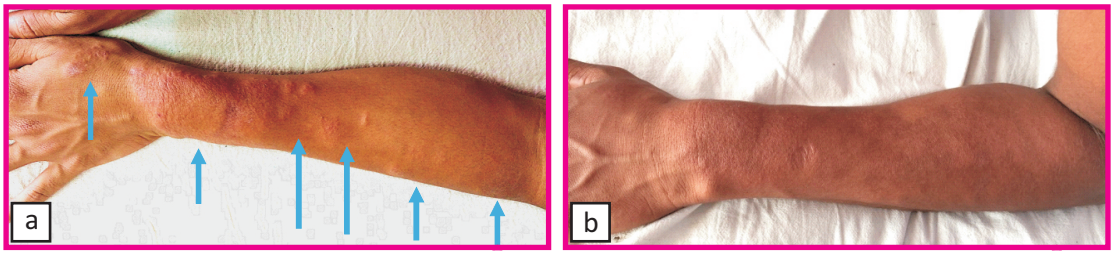

Fig. 1 (a) : A linearly arranged erythematous plaques from the postero-lateral of the left hand extending up to the elbow. There are few satellite lesions noted near the largest plaque. (b): The plaques had subsided significantly after 12 months of MDT-MB.

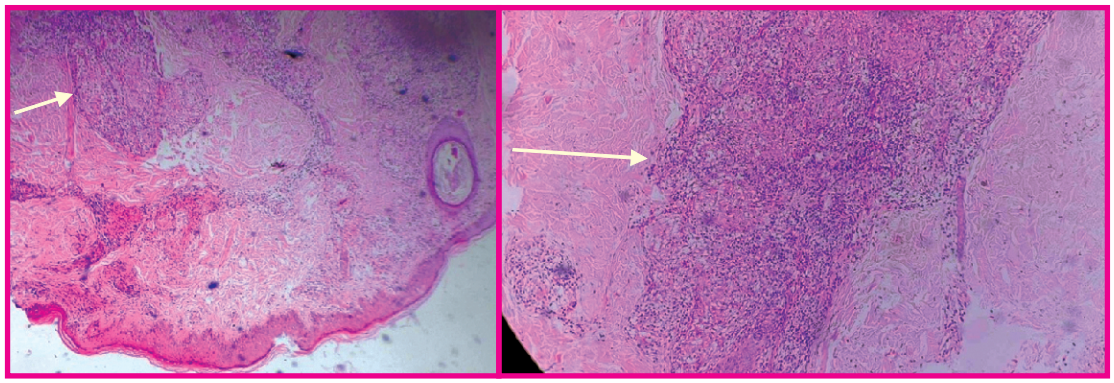

Fig. 2(a) : Figure showing multiple epitheloid cell granulomas in the dermis (H & E stain 40x magnification). (b) : Higher magnification showing few granulomas with peripheral rimming of lymphocytes in upper and deep dermis (H & E stain, 100x magnification) .

plaques were distributed in a linear fashion from extensor hands up to the elbow. There was no history of joint pain, fever or jaundice. There was no history of Hansen's disease in family members.

On examination, there were multiple linearly arranged erythematous plaques of different shapes and sizes over the extensor aspect of the left forearm, with the largest being 6cm x 3cm. Sensation to touch, pain and temperature were lost over all of the skin lesions (Fig. 1a).

Examination of peripheral nerves showed non-tender enlargement of the left ulnar and left superficial radial nerve. Motor examinations were grossly within the normal limit. Slit skin smear from bilateral earlobes and lesions and stained with modified Ziehl-Neelson (ZN) stain was negative for any acid-fast bacilli. An incisional biopsy from the lesion showed numerous granulomas consisting of epitheloid cell aggregates with peripheral rimming of lymphocytes in the upper and deep dermis (Fig. 2a & 2b). However, no acid-fast bacilli were seen.

The patient was diagnosed as a case of multi-bacillary leprosy based on the World Health Organization classification, and multi-drug therapy multibacillary type (MDT-MB) was started.

After two months of MDT-MB, a follow-up showed a significant decrease in erythema and induration over the lesions. A follow-up at 12 months of treatment showed complete clearance of the lesions (Fig. 1b). Patient was appropriately counselled and advised for follow-up if needed.

Discussion

Leprosy, a disease caused by *Mycobacterium leprae*, has tropism towards peripheral nerves and the skin. In pure neural leprosy, skin along the distribution of the affected nerve is either anaesthetic or hypo-anesthetic but as a rule classical skin lesion should not be present (Rao & Suneetha 2016). The microorganism's tropism and host immunity determines the clinical characteristics of leprosy (Fischer 2017).

It has been shown that cases of pure neuritic leprosy may develop skin lesion over the course of time. It is caused by an asymmetric invasion of *Mycobacterium leprae* in peripheral nerve fibers, an interaction that causes an inflammatory response at the axonal level resulting in peripheral nerve trunk inflammation and damaging sensory fibers (Rao & Suneetha 2016). Here, the plaques followed the path of left radial nerve. In this particular case, the patient did not have any symptoms pertaining to nerve involvement prior to skin eruption, suggesting that both of the processess were either simultaneous in onset or an early skin involvement.

Sensory loss in the nerve path with or without clear nerve thickening may be the only manifestation characterizing the primary neural form (Lastória & de Abreu, 2014) but no skin lesions.

Radial nerve is the largest nerve in the upper limb, with fibers originating from the brachial plexus. It divides into deep and superficial branches proximal to the elbow. The superficial pure sensory branch runs at the radial side of the forearm and dorsum of the hands. It innervates the posterior forearm and dorsa of hands in a continuous linear fashion (Bumbasirevic et al 2016). Here, in our case, the skin lesions were following the nerve course of radial nerve from the elbow distally.

An argument of genetic inheritance has even been put forth for leprosy presenting in linear fashion following the lines of Blaschko (Chetan & Shailesh 2019, Liegeon et al 2015, Vijaikumar et al 2001). Blaschko lines, which represent a classic pattern of cutaneous mosaicism, could be present in both congenital and acquired skin disorders (Molho-Pessach & Schaffer 2011). Genetic studies have also made a significant contribution to understanding the pathophysiology and susceptibility of leprosy (Cambri & Mira 2018). It has been shown that genetics play a role not only in the development of disease but also in the different clinical forms, reactions and even in recurrences (Sauer et al 2015, Uaska Sartori et al 2020).

Linear leprosy is one of the rare presentations of leprosy. Due to its unusual presentation, it may also lead to delay in diagnosis and timely management. Early recognition and prompt treatment may help in interruption of transmission and disability prevention which is crucial in maintaining leprosy eliminated status of the country. These rare forms of leprosy should be diagnosed early and treatment should be initiated.

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