

Case Report

Visible disability as a presenting feature of leprosy: a rare but real concern in post-elimination phase in India

Vasudha A. Belgaumkar, Ravindranath B. Chavan*,
Nitika S. Deshmukh, Dhanshri D. Gangode

Department of Dermatology, Venereology and Leprosy, B. J. Government Medical College, Pune, Maharashtra, India

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*Correspondence:

Dr. Ravindranath B. Chavan,

E-mail: drravindranathchavan@gmail.com

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ABSTRACT

Leprosy (Hansen's disease) is a chronic infectious disease caused by *Mycobacterium leprae*, affecting mainly peripheral nerves and skin. Delayed diagnosis contributes to sensory-motor impairment leading to deformity and disability, thereby imposing a major impact on the physical, social, economic and psychological domains of the quality of life of affected individuals. Deformities and disabilities are largely preventable complications associated with tremendous social stigma and discrimination, assuming public health significance. India declared elimination of leprosy more than a decade and half ago in 2005. Hence, grade 2 disability is expected to be a rare manifestation in the post-elimination era. However, to the contrary, the proportion of new leprosy patients with visible disability recorded in India exceeds the global rate. Herein we report a middle-aged female presenting with saddle nose, bilateral fixed ulnar and mobile median claw hands (grade 2 disability) with multiple trophic ulcers. This case emphasizes that clinical suspicion after careful examination of skin and neural manifestations is of utmost importance for the early diagnosis of leprosy. Timely self-reporting and management with WHO-multi drug therapy (MDT) remains the key to minimize the risk of deformity and disability. Above all, it is essential to create awareness amongst the community regarding prompt recognition of symptoms and easy access to appropriate care along with intensified efforts to address the associated stigma.

Keywords: Leprosy, Impairment, Disability, Diagnostic delay

INTRODUCTION

Leprosy is a chronic granulomatous infection caused by *M. leprae* mainly affecting peripheral nerves and skin. Despite declaring elimination in 2005, India is one of the countries with the highest leprosy burden with more than 1,14,451 new leprosy patients being detected every year, including 2,761 (24.38%) incident cases with a visible (grade 2) disability (G2D). This is significantly higher compared to the global grade 2 disability rate 5.8% (11,323 new G2D cases) as per WHO weekly epidemiological record (September 2020).¹ Leprosy has a lengthy incubation period with clinical features appearing within 3-7 years. The cutaneous and neural

manifestations are determined by the host immune response, characterizing leprosy as a spectral disease between two poles, tuberculoid to lepromatous.² Diagnosis is mainly based on clinical findings which are graded according to Ridley and Jopling classification.³ Cutaneous lesions usually presenting as hypoesthetic, hypopigmented or erythematous patches or plaques and neurological symptoms may occasionally be inconspicuous and are likely to be missed by both the patient and health care provider, leading to delay in diagnosis. One of the major factors responsible for stigmatization of leprosy patients is their identification in society due to visible disabilities.

CASE REPORT

A 40 years old married female, domestic help by occupation, visited the skin outpatient department with multiple wounds over hands and feet with pus discharge since, 5 months. On dermatological examination, she had a large well-defined erythematous infiltrated plaque with irregular borders over bridge of nose extending to bilateral malar areas, left lower eyelid and upper lip. Saddle nose deformity and lateral madarosis (bilateral) were prominent (Figure 1). There were multiple hypopigmented hypoesthetic patches over back, buttocks and lower limbs. Extremities showed bilateral fixed ulnar and mobile median claw hands with multiple trophic ulcers over knuckles, dorsum of right foot and both soles (Figure 2).



Figure 1: Well-defined erythematous infiltrated plaque with irregular borders over bridge of nose extending to malar areas, upper lip; single oval well-defined hypopigmented patch below left eye; saddle nose and lateral madarosis present.

Sensory examination showed hypoesthesia over bilateral upper limbs in ulnar distribution and over palms and soles. Motor examination revealed reduced muscle power (grade 4) in all lumbricals, interossei, abductor pollicis brevis, extensor hallucis longus, peronei longus and brevis with positive card test in both hands. Bilateral ulnar, radial cutaneous, common peroneal and posterior tibial nerves were thickened but non-tender.

Routine investigations were within normal limits except anemia (hemoglobin 9 gm/dl). Radiological investigations revealed resorption of distal phalanges of index, middle and little finger of left hand and index finger of right hand with flexion deformity and periarticular osteopenia.

Histopathological examination of the infiltrated plaque showed thinned out epidermis, and perivascular, periadnexal and perineural collection of foamy

histiocytes and few lymphocytes in dermis (Figure 3a). Ziehl-Neelson stain was positive for acid fast bacilli (Figure 3b).



Figure 2: Multiple hypopigmented hypoesthetic patches over back and gluteal region; multiple trophic ulcers over knuckles, dorsum of right foot and bilateral soles; fixed ulnar and mobile median clawing of bilateral hands.

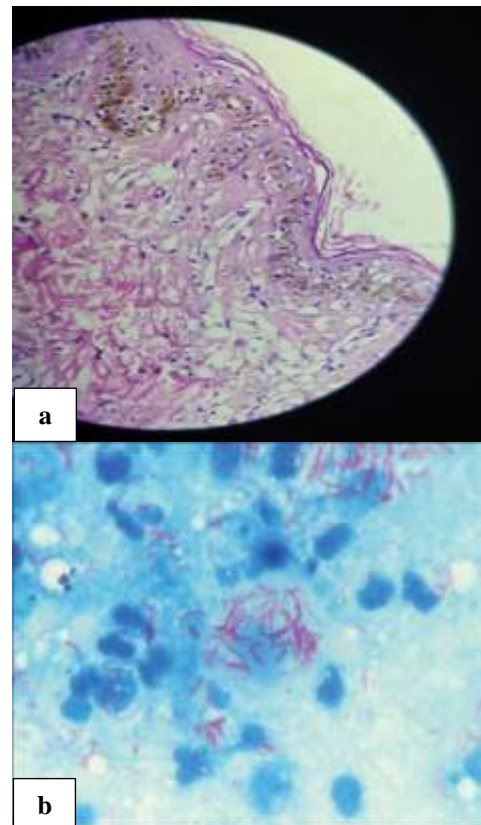


Figure 3: (a) Histopathological examination (H&E, 40X) of the infiltrated plaque showed thinned out epidermis, and perivascular, periadnexal and perineural collection of foamy histiocytes and few lymphocytes in dermis; (b) Ziehl-Neelson stain was positive for acid fast Bacilli.

Clinical examination and other investigations confirmed the diagnosis of borderline lepromatous Hansen's disease with secondary deformity (bilateral claw hands) with trophic ulcers and sensory-motor deficit. Patient was started on MDT for 12 months as per the NLEP guidelines.^{3,4} Rifampicin and clofazimine without dapsone until her anemia improves, intravenous antibiotics for her ulcers and hematinics. Physiotherapy and daily cleaning and dressing of the ulcers was done.

DISCUSSION

Among communicable diseases, leprosy is one of the foremost causes of peripheral neuropathy and preventable deformity and disability. In the year 2016, India reported 63% of the world's new leprosy cases; about 40% of the world's new G2D among new leprosy cases and an increasing trend (3.1% to 4.6%) of new cases with G2D in the period 2008–2015. Patients mainly in rural settings neglect the subtle symptoms which further delays the diagnosis and treatment.⁵ Peripheral neuropathy in leprosy leads to functional impairment resulting in inability to perform an activity considered normal for a human being of the same age, gender and culture defined as disability. Deformity is the visible anatomical alteration in the form, shape or appearance of the body. Deformities are classified as: specific, occurring due to the disease process (like loss of eyebrows, other facial deformities), those resulting from paralysis of some muscle due to peripheral nerve trunk damage (like claw-hand, foot-drop, lagophthalmos), or sensory–autonomic deficit induced injuries or infection to hands or feet (like scar contractures of fingers, mutilation of hands and feet, corneal ulceration).⁶ Disability associated with deformity is termed as visible disability.

Our case presented with paralytic (claw hand), as well as specific (madarosis, saddle nose) and sensory-autonomic (trophic ulcers) deformities. Timely initiation of treatment with MDT is essential for prevention of deformities and disabilities. The number of cases with G2D at the time of diagnosis directly reflects the delay in the early detection; the level of leprosy awareness in the community; the capacity of the health system to recognize leprosy early and to some extent the reach of services. Physical disability in leprosy is defined by the WHO in three categories (Table 1). Previous research on determinants of disability risk and delay in diagnosis among adult leprosy cases has demonstrated that the main risk factors for presenting with G2D/G1D at the time of diagnosis were multi-bacillary leprosy, patient delay of more than 3 months, health care provider delay of more than 1 month, daily wage laborer and age more than 60 years.⁷ Our patient had not approached the health-care system until she presented to us with irreversible grade 2 disability of hands and feet with previously undiagnosed multi-bacillary leprosy (indicative of patient delay). It is likely that early diagnosis and management would have either averted this complication altogether or at least allowed the hand function to be salvaged.

Recently, the government of India has rechristened the prevention of disability (POD) as DPMP (disability prevention and medical rehabilitation). It includes all activities aimed at prevention and care for disabilities and deformities. Previous studies in India and across the world have also reported varying proportion of disabilities among new leprosy cases.^{8,9}

Table 1: WHO disability grading (WHO 2014).

Hands and feet	
Grade '0'	No disability found
Grade '1'	No visible damage (loss of sensation)
Grade '2'	Visible damage (disability, wounds (ulcer), deformity due to muscle weakness, (such as foot drop, claw hand, loss /partial resorption of fingers / toes, etc.)
Eyes	
Grade '0'	No eye problem due to leprosy, no evidence of visual loss.
Grade '1'	No grade 1 for Eyes
Grade '2'	Inability to close eyes, obvious redness, visual impairment, blindness.

CONCLUSION

Clinical suspicion after careful examination of skin and neural manifestations is of utmost importance for the diagnosis of leprosy. Early self-reporting and management with MDT is the key to minimize the risk of deformity and disability. Above all, it is essential to create awareness amongst the community regarding prompt recognition of symptoms and access to appropriate management along with intensified efforts to address the associated stigma.

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