Case Report

Leprosy- Atypical Presentation with Reaction

Khan MMI

DOI: https://doi.org/10.3329/jafmc.v19i1.68450

Abstract

Leprosy is a chronic granulomatous disease affecting mainly the skin and nerves caused by the obligate intracellular pathogen Mycobacterium leprae. Patient not always present with typical skin lesion with loss of sensation or muscle weakness rather may present with atypical skin lesion with swelling of the affected area like cellulites or lymphangitis as reactional state. A 25-years-old male patient presented with linear erythematous verrucous plaque along with multiple nodules and pain and swelling of left upper extremity. Local examination revealed sensory loss on inner aspect of left hand and finger and ulner nerve found enlarged, thickened and tender. Slit skin smear showed numerous lepra bacilli. Skin biopsy for histopathology showed collection of foamy histocytes forming granuloma within the upper dermis and wade fite staining, moderate number of lepra bacilli was seen. Patient was diagnosed as a case of borderline leprosy with type 1 lepra reaction. Patient was getting treatment with MDT (multi drug therapy) and systemic steroid and responded very well. His skin lesions are almost subsided and weakness of hand was recovering gradually.

Key words: Leprosy, Type 1 Lepra reaction, Slit skin smear, Skin biopsy, Prednisolone, MDT.

Introduction

Leprosy is a chronic infectious disease caused by *Mycobacterium leprae*, an acid-fast, rod-shaped bacillus. The disease mainly affects the skin, the peripheral nerves, mucosa of the upper respiratory tract and the eyes but may also involve lymphoid tissue, testicles, muscles and bones. About 214,783 new cases detected worldwide in 2016, essentially unchanged for the last 4 years. More than 80% of all new cases are detected in only 3 countries- India, Brazil and Indonesia.[1] Diverse clinical presentations, the very low rate of disease progression and issues with misdiagnosis all create clinical challenges. Despite the global use of multidrug therapy, up to 30% to 50% of all leprosy patients experience some type of reactional episode that may result in a permanent neurological deficit or disability.[2] Leprosy is curable and early diagnosis and treatment can prevent disability. Here one case is reported who has admitted in CMH Dhaka in April 2022. The main objective of publishing this case report is to generate awareness amongst the medicos regarding the atypical presentation, early diagnosis and treatment of leprosy to prevent deformity and disability.

Case Report

A 25 years old boat seaman from Bangladesh Navy (L/Cpl) presented to Department of Dermatology, CMH Dhaka on 10th April 2022 as a transferred case from BNS Potenga with the complains of swelling of left hand and multiple reddish elevated skin lesions for last 20 days. He also presented with linear elevated rough mild scaly skin lesion extending from left arm to mid forearm. This linear lesion initially started as erythematous non itchy skin lesion in 2019. For this, he was consulted with Dermatologist at BNS Mongla and civil doctor multiple times without any improvement. For last 2 years, the skin lesion gradually became elevated linearly and extended from arm to upper part of forearm. For last 20 days the skin lesion became more reddish and elevated, dry, rough and subsequently developed multiple painful nodules on the left forearm, hands including little finger along with pain and swelling of left hand. Initially, sporotrichosis, swimming pool granuloma, eosinophilic fasciitis, cellulitis and leprosy were kept in differential diagnosis as patient didn't give any sensation loss or weakness of affected limb. On inquiry, he complains of dysasthesia on medial aspect of left forearm near wrist joint and hand. He had no history of allergy, no history of contact of any allergens or irritants. He had no past history of thyroid disorder, malignancy, diabetes, hypertension, bronchial asthma or any rheumatologic diseases.

Clinically patient was ill looking and anxious. Vitals were normal. On examination, integumentary system revealed there is well circumscribed erythematous rough, mild scaly linear elevated plaque extended from upper part of left arm to forearm, back of the hand up to little finger. Multiple erythematous nodules were also present on the forearm along with swelling and tenderness of left forearm and hand. Hypotrichosis was present over the lesions. Nail and mucous membrane were normal. Nervous system examination revealed temperature and touch sensation was diminished to absent on inner aspect

Lt Col Mohammad Mofizul Islam Khan, MBBS, DDV, FCPS, Classified Specialist in Dermatology & Venereology, CMH Ghatail (*E-mail:* shameem_101166@yahoo.com)

of the lower part of forearm, inner aspect of the hand and ring finger and half of the ring finger. Motor function was intact. Peripheral nerve examination revealed ulnar nerve was thickened, enlarged and tender on affected limb. Great auricular nerve was visibly enlarge and thickened. Other peripheral nerve examination was normal. Lymph node was not enlarged. Basing on the history and clinical finding he was diagnosed as Borderline Leprosy with Type 1 Lepra reaction.

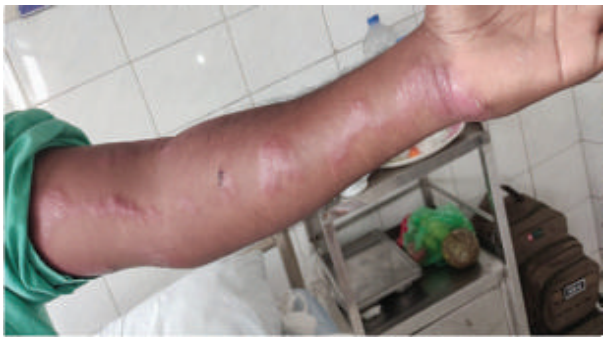

Figure-1: Swelling of the affected hand of the patient like cellulitis.

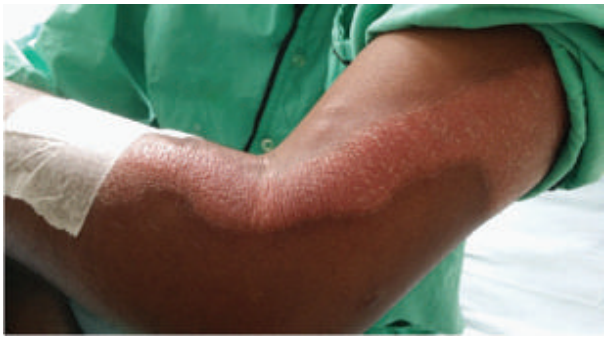

Figure-2: Scaly Linear Plaque of the case.

He was extensively investigated to confirm the diagnosis, to rule out other possibilities. His CBC showed HB- 15.10 gm/dl, WBC- 11,190, neutrophil- 84%, and Platelet- 244000. RBS, liver function test, serum creatinine and electrolyte were normal. Urine R/E showed numerous pus cell, serum albumin was 45mg/dl, A:G ratio was 2.3:1. Lipid profile was high (Serum cholesterol- 204 mg/dl, triglycerides- 192 mg/dl), CRP was positive (32.0 mg/dl). ANCA, ANA and anti-ds DNA were negative. X-ray of the left hand showed normal. MRI of left wrist joint was normal except subcutaneous soft tissue swelling with edema. Duplex study of the left upper limb vessels was normal. Slit skin smear from ear lobe and lesional skin showed numerous lepra bacilli. Skin biopsy for histopathology from lesional skin showed collection of foamy histocytes forming granuloma within the upper dermis. On the wade fite staining moderate number of lepra bacilli was seen within the macrophages arranged in cigar bundle. On PAS staining, no fungal element is seen. Therefore, he was confirmed the case as multibacillary leprosy with type 1 lepra reaction.

Figure-3: Resolution of the skin lesions after 2 months of treatment.

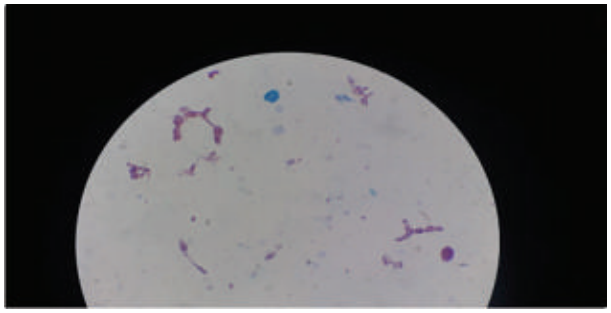

Figure-4: Slit skin smear showing numerous lepra bacilli.

After adequate education and counseling he was started treatment of multibacillary leprosy with Rifampicin, Dapsone and Clofazimine along with high dose oral Prednisolone for reaction on 19 April 2022.

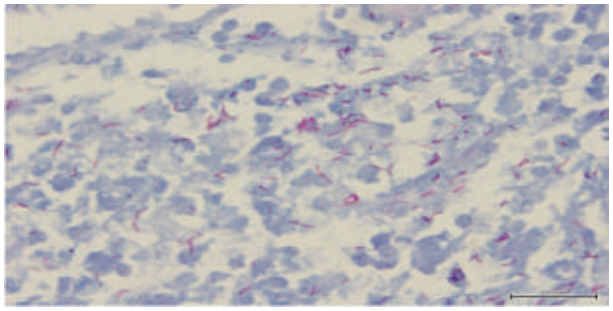

Figure-5: Skin biopsy for histopathology from lesional skin showing moderate number of lepra bacilli within the macrophages.

Discussion

Leprosy or Hansen's disease is a chronic disabling disease caused by *Mycobacterium leprae*, mainly affecting the skin and peripheral nerves. The causative agent was first identified in 1873 by the Norwegian physician, Armuer Hansen.[3]

It is most prevalent in tropical countries. The infection is more common in environments where the people are of low economic status with inadequqte housing, poor sanitation, poor nutrition and lack of education. In Bangladesh, the district and areas which are still endemic are as follows: Bandarban, Dinajpur, Rangpur, Nilfamari, Rangamati, Khagrachari, Lalmanirhat, Gaibandha districts and Dhaka and Chattogram metro.

Incubation period in tuberculoid leprosy is up to 5 years and for lepromatous leprosy may be 20 years or longer (Average 2-5 years). Mode of transmission is usually by Droplet infection from nasal secretions of active multibacillary cases, by shedding organism into the environment from ulcerated skin lesions and prolong close contact. Cardinal signs of leprosy includes skin lesions with anesthesia, positive skin smear and enlarged nerves with definite loss of sensation and muscle weakness.[4]

The Ridley and Jopling classification (on the basis of clinical, bacteriological, immunological and histo- pathology):
(I) Tuberculoid Leprosy (TT)
(II) Borderline Tuberculoid (BT)
(III) Borderline (BB)
(IV) Borderline Lepromatous (BL)
(V) Lepromatous Leprosy (LL)

WHO classifications (on the basis of BI/MI) are Paucibacillary and Multibacillary leprosy.

Table-I: Spectrum of Host-Parasite relationship in Hansen's disease

	High Resistance		Unstabbble Resistance		No Resistance
	Tuberculoid TT	Borderline Tuberculoid BT	Borderline BB	Borderline Lepromatous BL	Lepromatous LL
Lesions	One to three	Few	Few or many Asymmetrical	Many	Numerous and Symmetrical
Smear for bacilli	0	1+	2 +	3+	4+
Lepromin test	3+	2+	+	±	0
Histology	Epithelioid cells decreasing Nerve destruction, granuloma	sarcoidlike		Increasing hbistiocytes, foam cells Granuloma, xanthoma-like	

Atypical presentation of MB leprosy includes single plaque, single nodule on the face, erythema multiform like lesion, lympahdenopathy misreading as lymphoma, long standing leg ulcer, verrucous lesions of lepromatous leprosy, histoid leprosy, lucio leprosy and zosteriform segmental dermatomal leprosy.[5]

Changes in mucous membrane include nasal stuffiness, ulceration, perforation of nasal septum and saddle nose deformity, Perforation of the palate, Hoarseness of voice. Changes in eyes are corneal opacity, avascular keratitis, interstitial keratitis, pannus formation and acute corneal leproma. Changes in peripheral nerve includes anesthesia, paralysis, loss of sweating and vasomotor dysfunction. Changes in bone include osteoporosis of the phalanges, absorption of the phalangeal shaft of fingers, toes and metatarsals. Except gastrointestinal tract, lungs and brain, virtually every organ can contain leprosy bacilli. Liver, spleen, bone marrow, lymph nodes and testicles are commonly infected. Testicular atrophy and gynaecomastia may be present.

Reaction in Leprosy: During the usually chronic course of leprosy, acute episodes (reactions) may occur. Any type of leprosy, except an early indeterminate form, may undergo a sudden inflammatory phase of exacerbation. Sometimes, the reaction may be ushered in by immunological changes following effective chemotherapy and reduction in the bacillary load.[6] Reactions may also occur spontaneously or may be precipitated by intercurrent infections (viral malaria etc), anaemia, mental and / or physical stress, puberty, pregnancy, parturition or surgical interventions.

Two kinds of hypersensitivity are believed to underlie to be wildering clinical manifestations that may appear during reactions.

Type 1 lepra reaction is an example of type IV hypersensitivity (allergic) reaction (Coombs and Gell). The terms "reversal" or "upgrading" are used for the reaction following an increase in the cell–mediated Immunity (CMI) resulting in a shift towards the tuberculoid pole, and the term down grading is applied to the reaction associated with a decrease in CMI resulting in a shift towards the lepromatous pole. Reversal reaction commonly follows treatment, drowngrading reaction only occurs in a patient who is not receiving adequate treatment. Type 1 lepra reaction is considered severe if the pain and tenderness in the nerves is severe, if paralysis or anaesthesia threatens to follow the neuritis and if the skin is so severely inflamed that it is likely to ulcerate.

Type 2 lepra reaction (Erythema nodosum leprosum) is humoral hypersensitivity and it is an example of type III hypersensitivity (allergic) reaction (Coombs and Gell). It tends to occur later during the course of treatment when skin lesions appear quiescent and most of the bacilli in the skin are granular. However, a patient may be in reaction (type 1or 2) when first seen. During reactions inflamed skin lesions and nerves may be extremely painful and tender. Acute neuritis may cripple patients with borderline leprosy overnight, while acute iritis may rapidly result in blindness. In patients with borderline-lepromatous leprosy Type 1 and Type 2 lepra reactions may occur simultaneously.

Diagnosis is based on examination of the skin and peripheral nerves, identifying the infectious organism in slit skin smears and skin biopsy.[7] Newer diagnostic tests are Serologic test to detect antibodies against *M. leprae* antigen Phenolic Glycolipid-1 (PGL-1), Detection of *M. Leprae* by polymerase chain reaction (PCR) and Fluorescent leprosy antibody absorption test (FLA-ABS). Slit skin smear from ear lobe and lesional skin may show no bacilli to numerous lepra bacilli from tuberculoid to lepromatous leprosy. Skin biopsy from lesional skin shows dermal tuberculoid granulomas with no grenz zone in tuberculoid leprosy and in lepromatous leprosy, granuloma composed of foamy histocytes to lipid-laden histocytes (lepra cells or foam cells of Virchow) forming granuloma within the upper dermis separated from the epidermis by a well defined grenz zone. Acid-fast bacilli are typically abundant in lepromatous leprosy and appear as round clumps (globi).[8]

Antileprotic multidrug therapy is the mainstay of treatment.[9] The WHO recommendation for paucibacillary disease is 600mg of rifampicin under supervision once monthly for 6 months and 100mg/day of dapsone for 6 months, unsupervised. Multibacillary patients are treated with rifampicin 600mg and clofazimine 300mg, once monthly under supervision, with dapsone 100mg/day and clofazimine 50mg/day for 12 months. Type 1 reactions are usually managed with systemic corticosteroids. Prednisolone is given orally, starting at a dose of 40-60mg daily. Once the reaction is controlled, the prednisolone may need to be tapered slowly over months to years. Thalidomide is uniquely effective against ENL and is the treatment of choice.[10] Systemic corticosteroids are also effective in type 2 reactions but long-term use may lead to complications. Clofazimine in higher doses, upto 300mg/day, is effective in ENL and may be used alone or to reduce corticosteroid or thalidomide doses. Supportive measures include moral support and reassurance and proper attention to diet and general health.[11]

Without proper treatment a wide range of complications may occur. Complications due to massive invasion of tissues by *M. leprae* include rhinitis, laryngitis, iritis, testicular atrophy, myopathy, lymphadenopathy, gynaecomastia, leonine facies, saddle nose and palatal perforation. Complication also occur due to reactions type-1 Lepra reaction (reversal or downgrading reaction) and type-II Lepra reaction (ENL). Complications due to immune suppression are intercurrent infection, nephritis and amyloidosis.[12] Complications due to nerve damage include anaesthesia, paralysis and loss of sweating and vasomotor dysfunction. Secondary complications following anesthesia, paralysis and autonomic dysfunction are tissue necrosis, plantar ulceration, malum performance pedis, secondary bacterial infection and cellulites.

BCG vaccination alone provides about 34-80% protection against *M. leprae* infection.[13] Prevention also depends on treating active multibacillary patients and examining exposed persons on an annual basis to detect early evidence of infection. Prophylactic antibiotic regimens have been used in such exposed patients and demonstrate a reduction in new Hansen's disease cases by more than 50% in the first 2 years.[14]

Conclusion

Leprosy is the most ancient disease in the history of mankind. Though the disease is most ancient, its effective treatment with Multi Drug Therapy (MDT) was introduced only in about 20 years. Effective use of MDT has made it possible to eliminate leprosy as a public health problem in the vast majority of the countries of the world including Bangladesh. Prevention of leprosy by improvement of living conditions, reduction of overcrowding and poverty are a major public health challenge. Vaccination with BCG is partially effective in preventing leprosy. Leprosy patients should be treated with patience, perseverance and understanding. Besides the medical treatment, leprosy patient needs moral support and reassurance so that he can gain self confidence and self respect. Early detection of Leprosy and treatment by MDT are the most important steps in preventing deformity and disability.

References

1. William D. James MD, in Andrews' Diseases of the Skin: Clinical Dermatology. Hensen's Disease, 12th ed; 2016:331-42.

2. Ridley DS, Jopling WH. Classfication of leprosy according to immunity: A five-group system. Int J Lepr Other Mycobact Dis. 1966; 34:255-73.

3. Vineetha M, Seena P, Sobhana KK et al. Atypical manifestations of leprosy- A case series. Indian J Lepr. 2016; 88(1):1-6.

4. Jindal R, Shirazi N, Jindal R et al. Uncommon clinical presentations of leprosy: Apropos of three cases. Lepr Rev. 2016; 87(2):246-51.

5. Flageul B. Diagnosis and treatment of leprous neuropathy in practice. Rev Neurol (Paris). 2012; 168(12):960-6.

6. Strobel M, Ndiaye B, Marchand JP et al. Leprosy tests: Diagnostic problems (apropos of 2 cases. Acta Leprol. 1981; 83:11-9.

7. Chowdhary KN, Rao R, Priya P et al. Cutaneous sarcoidosis misdiagnosed as leprosy. Report of two cases and review of literature. Indian J Lepr. 2016; 88(3):177-83.

8. Das S, Roy AK, Kar C, Giri PP. Atypical presentation of leprosy: A report of two cases. Indian J Dermatol. 2007; 52:198.

9. Sharma VK. Leprosy: Classification and clinical features. IADVL Text book of Dermatology. 3rd ed, 2016:1578-601.

10. Bentekan L, Hassam B. Atypical presentations of leprosy: Apropos of 2 cases. Acta Leprologica. 1997; 10:195-8.

11. Suzuki Y, Takigawa M, Ishii N. A case of suspected multibacillary leprosy. Nihon Hansenbyo Gakkai Zasshi. 2003; 72:287-90.

12. Vinitha G, Asha T, Thankappan TP et al. Hansen's Disease "Clinically Atypical Presentation with Atypical Histopathology". Indian Journal of Leprosy. 2019; 91(2):159-64.

13. WHO. "Global leprosy update, 2014: Need for early case detection". The Weekly Epidemiological Record. 2015; 90(36):389–400.

14. Clapasson A and Canata S. "Laboratory investigations," in Leprosy: A Practical Guide. Nunzi E and Massone C. 1st Ed, 2012:55–82.