Lepromatous Leprosy with Erythema Nodosum Leprosum Masquerading as Quadriparesis

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INTRODUCTION

Leprosy, also known as Hansen’s disease, is a chronic infectious disease caused by Mycobacterium leprae, which mainly affects the skin and peripheral nervous system with a potential to cause significant nerve damage and disability. Leprosy is caused by mycobacteria which are a group of bacteria, described to be a fungus initially, had the unique ability of acid fastness which is due to the presence of a chemical in the cell wall called mycolic acid. There are different species of mycobacteria some pathogenic, e.g., M. tuberculosis, M. leprae, M. marinum, M. scrofulaceum, etc. and some non-pathogenic e.g., M. fortuitum, Mycobacterium indicus pranii etc. Some of the mycobacteria are fast growers in vitro and some are slow growers. M. leprae is placed somewhere in between M. tuberculosis and M. avium intracellular-scrofulaceum complex.

This organism is acid fast and a rod-shaped organism. It has parallel sides and rounded ends and its morphology and size is like that of the tubercle bacillus. It occurs in abundant numbers in the LL, within lepra cells which are grouped as in bundles of cigars or as clumps of bacilli in capsular material called as globi. It has been observed that leprosy bacilli when stained with carbolfuchsin, if they appear as solid acid-fast rods, then they are viable and if bacilli stain irregularly, they are probably dead or can be degenerating. Morphological index and the bacteriological index depend on bacilli found in skin or nasal smears and are useful in determining the intensity of infection, and if the organisms are viable. Leprosy is one of the oldest diseases in the world, and yet there are many doubts and conflicting information about the origin and description of the disease.

It is an infectious disease which can lead to deformities of face, hands and feet. Leprosy results in skin lesions and deformities, mostly affecting the areas having lower temperatures or the cooler regions of the body. It is still called Kushtrog in most Indian languages, as it was in Sushrutha’s time. Gerhard Henrik Armourney Hansen discovered the pathogenic leprosy bacillus in 1871. After 1943, sulfones replaced Chaulmoogra oil in the treatment of leprosy. Guy Henry Faget described the greater effectiveness of sulfones in 1943. Patients were mistreated and received inhumane treatment everywhere. Sadly, there are reports in the Middle Ages that described mass execution of many lepers. So, from the moment the people were diagnosed with the disease, they carried the burden of isolation and discrimination for the rest of their lives.

Even though the prevalence has decreased significantly with the help of multidrug therapy (MDT), leprosy remains a public health problem in many areas and poses diagnostic and treatment challenges. Transmission occurs by nasal droplets of cases, although it could also spread by direct contact. Type 1, or reversal reactions occur mostly in borderline patients, although they can occur anywhere along the disease spectrum, and occur in 30% of patients. Reversal reactions typically present as enlargement of skin lesions, neuritis, and nerve dysfunction.
Type 2 reaction or erythema nodosum leprosum (ENL) is characterised by brightly red, raised, evanescent, tender nodules, and plaques of varying sizes. Although both reactions can cause nerve inflammation and damage, this is more likely to occur in Type 1 reactions, whereas systemic symptoms and evidence of inflammation occur more commonly in Type 2 reactions. Transmission pathways of M. leprae are not fully understood. Solid evidence exists of an increased risk for individuals living in close contact with leprosy patients, most likely through infectious aerosols, created by coughing and sneezing, but possibly also through skin to skin contact. Multidrug treatment developed in the 1980s is an effective chemotherapy and has played a critical role in the worldwide reduction of the leprosy burden, by reducing human-to-human transmission. Even though leprosy has been eliminated in most countries, in certain areas endemic leprosy persists.

According to World Health Organization (WHO), there was a considerable decrease in the number of new cases detected around the world, from 244,796 in 2009 to 208,641 in 2018. India, which has the highest leprosy burden, has reported a decrease in incidence of new cases by 15,000 over a period of past 2 years. New cases detected in India in 2018 were 120,334. The decreasing trend observed in India, which now accounts for < 60 % of global leprosy, reflects the leprosy situation at regional and global levels.

We hereby report a case of lepromatous leprosy which was misdiagnosed due to its atypical presentation. Since reactions can occur at any point during the disease, type 2 reaction may be the first manifestation of the disease and thus even more difficult for clinicians to diagnose as occurred in this case. Early diagnosis of a leprosy reaction is crucial for starting prompt treatment and relief of symptoms so as to arrest nerve damage.

Diagnosis is based on the clinical signs and symptoms. In an endemic country, a patient should be diagnosed as having leprosy if he shows one of the three cardinal signs:
1) Definite loss of sensation in a hypopigmented or erythematous skin patch;
2) A thickened peripheral nerve with loss of sensation;
3) Acid-fast bacilli in slit-skin smear.
The skin lesion may be single or multiple, usually hypopigmented or sometimes erythematous macules, papules or nodules. Loss of sensations is a common feature. Thickened nerves are another important feature of leprosy. In some cases, rod-shaped, acid fast leprosy bacilli are seen which are diagnostic of the disease.

**PRESENTATION OF CASE**

A 32-year-old male patient presented to the emergency with complaint of high-grade fever (104°F on admission) on and off for 1 month. Patient was misdiagnosed as a case of dengue fever by a local practitioner 1 month back for which he took unknown medications. He also complained of few episodes of vomiting and weakness over both lower limbs in the last 21 days which gradually progressed to involve both upper limbs over a span of 7 days. Patient was not able to walk and carry out his daily routine activities. Patient also complained of few nodular and macular lesions over face 15 days back, which were thought to be a consequence of unknown medications he took from the local practitioner, but these resolved over 2 - 3 days. Patient had no history of any chronic illnesses, trauma, and bowel and bladder dysfunction. Patient was admitted under the Department of Medicine where magnetic resonance imaging (MRI) was advised which showed bulge at C3 - C4, C4 - C5 and C5 - C6 vertebrae.
Council muscle power grade: 3 out of 5) Deep tendon reflexes were intact. Atrophy of thenar and hypothenar muscles present. High stepping gait was noted. During his hospital stay, patient developed multiple erythematous, tender nodules and plaques over face, trunk and extremities. Dermatological opinion was sorted where on examination, hot and cold sensations were lost over bilateral upper and lower limbs. On nerve examination, ulnar and common peroneal nerves were thickened, and patient had severe deformities of hand and feet. Patient had bilateral partial claw hand and bilateral foot drop.

Skin smears were done which was positive for acid fast bacilli. Biopsy from the lesions revealed dermal infiltration of lymphocytes, foamy histiocytes and neutrophil infiltration with perivascular lymphocytic infiltration.

**FINAL DIAGNOSIS**

Lepromatous Leprosy with ENL

**DISCUSSION OF MANAGEMENT**

The patient was investigated for glucose-6-phosphate dehydrogenase (G6PD) which was normal and then was administered multibacillary multidrug therapy along with injection dexamethasone 8 mg and physiotherapy exercises. Patient showed dramatic improvement after which steroid was tapered gradually.

**DISCUSSION**

Leprosy is a chronic and progressive granulomatous disease affecting the skin and nerves that is caused by *Mycobacterium leprae*. Leprosy presents a spectrum of clinical presentation. It has been classified by Ridley and Jopling as tuberculoid (TT), borderline (BT, BB, BL) and lepromatous (BL) leprosy based on histological and immunological features. Two types of reactions can occur in patients with leprosy.

Type 1 or reversal reaction is a delayed type hypersensitivity reaction, which may occur in borderline leprosy cases. Type 2 reaction or erythema nodosum leprosum is a type 3 hypersensitivity reaction presenting as a sudden onset of acute inflammatory response due to the deposition of immune complexes. Type 2 lepra reactions may develop before, during or after treatment with multidrug treatment. In Type 2 reaction, the antibodies combined with *Mycobacterium leprae* antigen form immune complexes that circulate and deposit in various tissues, activate complement, and damage the tissues and presents with erythematous tender nodules or plaques. Quadriparasis is a condition characterised by weakness in all four limbs. Differential diagnosis of quadriparasis includes compressive myelopathy, transverse myelitis, brainstem encephalitis, infection like poliomyelitis and enterovirus, Guillain-Barré syndrome, diabetes, uraemia, drugs, alcohol, myasthenia gravis, Lambert-Eaton myasthenic syndrome, snake bite. In our case, the presentation was atypical wherein the patient first reported with fever which was thought to be due to dengue, and muscle weakness which was thought to be due to cervical cord compression.

The skin lesions were missed by the physician suspecting it to be drug reaction to the unknown medications and were ignored as they resolved spontaneously in a few days which can occur as a consequence of type 2 reaction. This led to a delay in the diagnosis of ENL and patient presented with deformities which could be avoided with early diagnosis and prompt treatment with antileprosy drugs and corticosteroids. Delay in diagnosis have been reported in Asia, Africa, Europe, and North America, and delays are associated with higher rates of permanent nerve damage and disability. In our case,
two types of diagnostic delays were observed, health system delay and patient delay.

**CONCLUSIONS**

Leprosy is a leading cause of preventable disability worldwide. Delay in diagnosis of patients augments the transmission of infection and allows progression of disease and more severe disability. To reduce this delay, it is important to identify factors that hinder patients from presenting to doctors, and doctors from diagnosing patients once they have presented.¹⁵

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