

CASE STUDIES

Leprosy mimicking vasculitis

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Abstract

Leprosy is a chronic disease primarily involving the peripheral nerves and the skin. Painless skin patches and ulceration in hands and feet are the common clinical presentation of the disease. The present study reports a relatively unusual case of leprosy mimicking vasculitis in a 32-year-old male.

Keywords: Leprosy, vasculitic neuropathies, erythema nodosum, ulcer, mononeuritis multiplex

Introduction

Leprosy or Hansen's disease, caused by *Mycobacterium leprae* is a chronic, systemic granulomatous disease mainly involving the peripheral nerves and the skin.¹The common clinical presentations of the disease include painless skin patches with loss of sensation, thickening of skin and ulcerations in hands or feet.² Vasculitis-like presentations with skin ulcers and neuropathy are noted in rare cases. The present study deals with an interesting case of leprosy in a 32-year-old male mimicking vasculitis.

Case report

A 32-year-old male presented to the outpatient department (OPD) with the complaint of tingling numbness in the right hand for the past 2 years. His history revealed ulcer on the dorsum of right hand (Fig.1), and decreased sensation of pain and temperature in the right little finger. The condition had been diagnosed earlier as sensorimotor axonopathy and the patient was feeling better under the treatment of a neurologist using steroids and pregabalin for 2 weeks. But, he abruptly developed fever with painful nodular skin rashes on both lower limbs, which gradually progressed to upper limbs (Fig. 2). He also reported a decreased sensation in both the feet. The patient was farmer by occupation. Upon suspecting the infective etiology, the patient was prescribed with antibiotics and nonsteroidal anti-inflammatory drugs (NSAIDs).

Due to the persistence of asymmetric sensorimotor

axonopathy and nodular skin rashes, he was referred to the rheumatology OPD for vasculitis evaluation. On physical examination, the patient was noted to have bilateral ulnar nerve thickening, erythema nodosum, decreased sensation of pain and temperature in the right ulnar nerve distribution and motor weakening in the right and left peroneal nerve distribution. These findings raised the suspicion of leprosy. The patient also had leonine face (Fig. 3). Conducting a physical search revealed the presence of multiple hypopigmented macules on the back (Fig. 4). The results of clinical and laboratory investigations of the patient are provided in table 1. Skin biopsy of the patient revealed 1+ lepra bacilli on slit skin smear. The nerve conduction studies were repeated and were suggestive of asymmetric sensory motor demyelination and axonal neuropathy. Based on the clinical and lab findings and the presence of multiple hypopigmented patches and erythema nodosum macules, the diagnosis was suspected as leprosy with type 2 lepra reactions. The diagnosis was subsequently confirmed by a dermatologist. He was treated with steroids and anti-leprosy medications.

Discussion

The cardinal signs of leprosy are hypopigmented skin and enlarged peripheral nerves with loss of sensation. One of the common manifestations of these patients includes mononeuritis multiplex involving several peripheral nerves.^{1,3} On the basis of immunological reactions, leprosy has been classified into two types (type 1 and type 2).

Type 1 lepra reactions are characterized by the presence of inflammation in the existing lesions, skin infiltrates and new nodules.⁴ Type 2 lepra reactions or erythema nodosum leprosum (ENL), result in immune complex-mediated vasculitis, and are characterized by the presence of painful, bright red nodules on extremities and face.⁵ Histopathologic features of ENL include erythema nodosum, edema of papillary dermis, neutrophil infiltration and vasculitis on the

background of macrophage granuloma.^{5, 6}

Vasculitic neuropathies are peripheral nerve disorders causing inflammation mainly in the epineural arteries, subsequently leading to ischemic damage.⁷ Vasculitis neuropathy may be associated with symptoms like skin lesions, mononeuritis multiplex, tingling in the affected nerve distribution and weakness in the nerve.^{8, 9} The various

Table 1: Results of clinical and laboratory investigations

Parameters evaluated	Results
Hemoglobin (g/dL)	12
Total WBC count (cells/cumm)	17770
Neutrophils (%)	86
Lymphocytes (%)	9
Eosinophils (%)	4
Monocytes (%)	-
ESR (Westrgren's method) (mm/hour)	108
Platelet count (lakhs/ cu.mm)	5.52
Urine routine	
Sugar	Trace
Protein	Nil
WBC	Plenty
RBC	Nil
Serum creatinine (mg/dl)	0.7
CRP (mg/L)	51.65
ANA (IF)	Negative
C3 (mg/dl)	235
ANCA (immunofluorescence)	Negative

Fig. 1: Painless ulcer on the right hand



Fig. 2: Skin rashes on lower limb



Fig. 3: Leonine face



Fig. 4: Hypopigmented patches on the back



laboratory features of non-specific systemic manifestations include leukocytosis, thrombocytosis, elevated levels of ESR, C-reactive proteins (CRP), and normocytic and normochromic anemia.¹⁰

In the present patient, leprosy was suspected due to the occurrence of painless ulcer and erythema nodosum. A careful search for hypopigmented patches suspected the clinical diagnosis of leprosy. The finding was further corroborated by split skin smear and the diagnosis was confirmed by a dermatologist. Macular hypopigmented lesions are generally localized on the face, ears, elbows, knees and buttocks.¹¹ A 2009 study by Kumar and Dogra has reported that the presence of hypopigmented or erythematous patches, accompanied by definite sensory loss, is the prominent initial clinical sign noted in newly diagnosed leprosy patients.¹² Even though paresthesia was present in the hands and feet of the patient, ulcers were painless. Ulcers are uncommon presentation in leprosy and occur due to loss of sensation. Their occurrence in ENL is characterized by the presence of painless nodules in areas of repeated trauma like arms, legs and face.¹³ Vasculitic neuropathies are mostly associated with severe pain due to the effective transfer of nerve growth factors to the sensory axons, and hyperesthesia and painful ulcers.¹⁴ However, the elevated levels of CRP, ESR, platelets, WBCs and skin lesions signified the possible diagnosis of vasculitis. But, reduced sensation and absence of painful ulcers with skin lesions (erythema nodosum and hypopigmented patches) supported the differential diagnosis as leprosy with type 2 lepra reactions.

The presence of mononeuritis multiplex in the current patient indicated the presence of both leprosy and vasculitic neuropathy. The nerve conduction studies of the patient revealed asymmetric sensory motor demyelination and axonal neuropathy. Asymmetric patterns of sensory nerve impairment associated with few axonal and demyelinating lesions and demyelinating lesions with secondary axonal degeneration in motor nerve are common nerve conduction findings in leprosy neuropathy.¹⁵ Whereas, axonopathy alone is a feature of vasculitis.¹⁶

The possibility of other inflammatory diseases like lupus and primary vasculitis (indicated by the increased levels of ESR and CRP) were excluded due to the absence of antinuclear antibodies (ANA and ANCA) and other clinical signs of the disease. Eventually, the detection of lepra bacilli on slit skin smear confirmed the differential diagnosis of leprosy in the current patient.

The study by Ovid *et al.*, conducted on 31 subjects with neural leprosy, has reported the presence of positive CRP in around 35% of the subjects.¹⁷ Leal *et al.* (2006), have also reported significantly higher levels of CRP and ESR in subjects with lepromatous leprosy compared to the control.¹⁸ In addition, elevated platelet count has been reported in ENL patients. The study by Rea *et al.* (2002) conducted on 11 patients with ENL have reported a significant increase in the mean platelet count from $235 \times 10^3/\text{mm}^3$ to $322 \times 10^3/\text{mm}^3$.¹⁹

Literature evidence on Hansen's disease resembling

vasculitis is limited. Sampaio *et al.* (2011) have reported the case of an 86-year-old lady, whose differential diagnosis included primary vasculitis and diffuse connective tissue disease. The diagnosis was finally confirmed as leprosy.²⁰ Misra *et al.* (2014) have reported a case of lepra reactions with Lucio cutaneous vasculitis in a 20-year-old female.²¹

In conclusion, the current case suggests that the probable diagnosis of leprosy should be considered in patients with mononeuritis multiplex (nerve thickening), with reduced temperature and pain, with or without erythema nodosum. A careful search for hypopigmented patches should be carried out in such patients

Competing interests

The authors declare that they have no competing interests.

Citation

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