Case Report

Hansen's Arthritis: An Overlooked Entity

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Abstract:

Leprosy, a chronic granulomatous infection caused by Mycobacterium leprae, classically presents with cutaneous and neurological manifestations. Musculoskeletal involvement though third most common is under diagnosed and underreported. This case report aimed at presenting a clinical scenario of rheumatological manifestations of leprosy which are often confused with primary rheumatic disorder. Diagnosis is often delayed if typical cutaneous menifestations of leprosy are absent

Introduction

Leprosy, a chronic granulomatous infection caused by Mycobacterium leprae, classically presents with cutaneous and neurological manifestations. Musculoskeletal involvement though third most common is underdiagnosed and underreported. It may manifest in the form of Charcot's arthropathy, acute symmetrical polyarthritis or swollen hands and feet syndrome during lepra reactions, insidious-onset chronic symmetrical polyarthritis mimicking RA or as isolated tenosynovitis or tenosynovitis associated with arthritis or neuropathy. At times, articular involvement may be the sole presenting manifestation even without cutaneous lesions. Other rheumatological manifestations occasionally reported are enthesitis, sacroiliitis, cryoglobulinaemic vasculitis and DM. Delay in diagnosis and management may be detrimental and may result in deformities and loss of function. This case report aimed at presenting a clinical scenario of rheumatological manifestations of leprosy which are often confused with primary rheumatic disorder. Diagnosis is often delayed if typical cutaneous menifestations of leprosy are absent.

Case Report

58 yrs male patient, textile industry worker recently detected case of type 2 diabetes mellitus came with complaints of pain and swelling over both hands and feet associated with marked limitation of movements of hands and feet since 15 days. There was also history of pain and swelling over bilateral knee joints . He also complained of tingling numbness in glove and stocking distribution simultaneously associated with generalised weakness. There was no family history of neuropathy. But he had positive family history of hansen's disease .

On examination he was found to have extensor tenosynovitis of hands, arthritis of wrists, metacarpo-phalangeal (MCP) and proximal interphalangeal (PIP) joints(figure 1) , knee joints bilaterally , swelling of hands

Figure 1: Photograph showing swelling of small joints of hands with extensor tenosynovitis at wrists

and tender swollen feet (figure2) There were no skin lesions. Bilateral ulnar nerves were thickened. There was

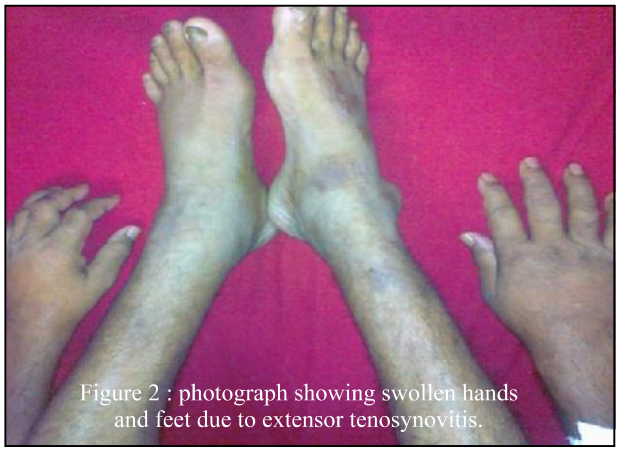

Figure 2 : photograph showing swollen hands and feet due to extensor tenosynovitis.

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hyposthesia in glove and stocking distribution Rest of the nervous and systemic examination was normal.

During hospital stay patient developed nasal cellulitis. 20 cc pus was drained from lesion. His complete haemogram, renal function tests, liver function tests. His urinalysis was normal except for microalbuminuria. He was found to have raised inflammatory parameters

(erythrocyte sedimentation rate 55 mm in the first hour, C-reactive protein 3.5 mg/dl; normal <0.6 mg/dl). RF, ANA were negative. NCS showed symmetric sensory motor polyneuropathy in both upper and lower limbs . Right sural nerve biopsy (figure 3)

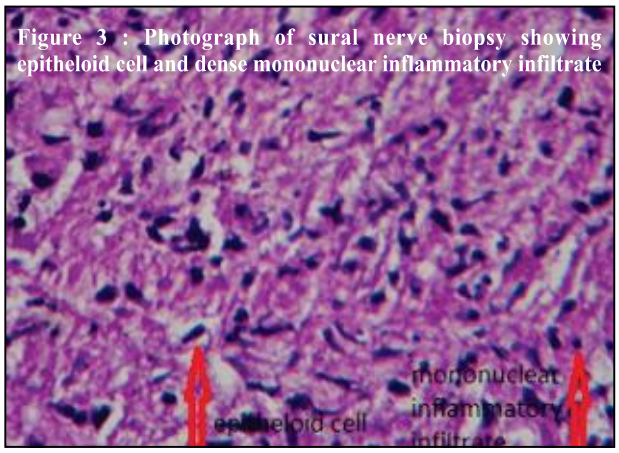

Figure 3 : Photograph of sural nerve biopsy showing epitheloid cell and dense mononuclear inflammatory infiltrate

Synovial biopsy of the left extensor tenosynovial sheath showed extensive mononuclear inflammatory infiltrate but no lepra bacilli.Pus drained from nasal abscess did not reveal lepra bacilli. Fite faraco stain of nerve biopsy showed lepra bacilli (figure 4).

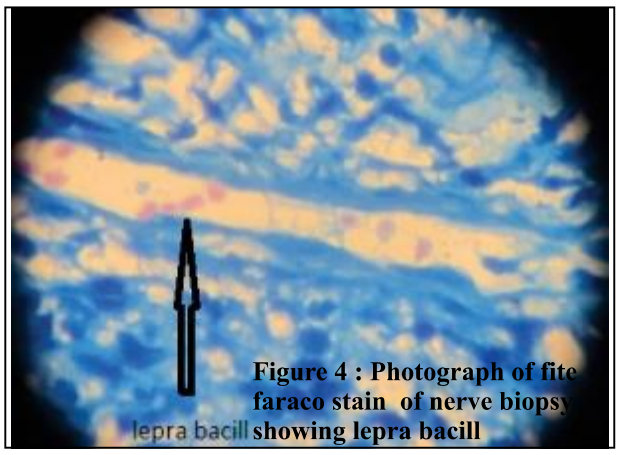

Figure 4 : Photograph of fite faraco stain of nerve biopsy showing lepra bacill

A diagnosis of pure neuritic (PN) leprosy with symmetric arthritis and tenosynovitis with recently detected type 2 DM with nasal cellulitis was made. Multi-drug therapy (dapsone, rifampicin, clofazimine) was started along with broad spectrum antibiotics to take care of his nasal infection. Because of his nasal cellulitis steroids were started a week later. Prednisolone (1 mg/kg/day) was started after his nasal infection healed. He had mild improvement in his symptoms at 8 weeks however his diabetic status deteriorated.In view of this his prednisolone was tapered and he was started on tab thalidomide 100 mg/day. He had significant relief in arthritis and tenosynovitis. He discontinued thalidomide after 3 months when he had complete remission of arthritis and tenosynovitis . At last follow-up, 6 months later, he was in complete remission. He was advised to continue drugs for the prescribed period and advised regular follow up.

Discussion:

Leprosy is a chronic granulomatous infectious disease caused by Mycobacterium leprae with predominant involvement of skin, nerves and eyes . The introduction of multi-drug therapy in 1988 has reduced the burden of disease considerably except in a few countries [1]. As of June 2006, the prevalence in India is 0.84 per 10 000, but nine states/union territories of the country still have not achieved the elimination target of the World Health Assembly [2] India is considered to be the epicentre of the problem, where 260 000 of the 408 000 people diagnosed across the world in 2004 reside [3]

The classical presentation of leprosy is in the form of hypesthetic/anaesthetic, anhidrotic macules, patches, plaques or papulo-nodular lesions. Neural involvement can manifest as paresthesias or as ensori-motor mononeuropathy, mononeuritis multiplex or polyneuropathy. Articular involvement in leprosy has been recognized since 600 B.C. in Chinese literature.[4] The two most common and well-recognized forms of articular

involvement, neuropathic joints and post-traumatic septic arthritis occur as a consequence of neuropathy [5].

In recent times, primary articular involvement in leprosy, due to infiltration by M. leprae or as part of lepra reaction, has been recognized. The reported prevalence varies from 1% to 70% [6] Acute and chronic symmetric polyarthritis involving hand joints, mimicking rheumatoid arthritis (RA), has been described with or without lepra reaction . Acute onset painful oedema of hands with marked restriction of movements and nodules along the extensor tendons was described in 1980 [7]. Pure enthesitis of the heel, sacroilitis, cryoglobulinaemic vasculitis, dermatomyositis, tenosynovitis and vasculitic rash are included in the spectrum of rheumatological manifestations of leprosy. However, almost all of these major reports are in patients with characteristic features of leprosy. In our patient male sex, absence of extraarticular manifestation of Rheumatoid arthritis, negative

serological investigations, thickened peripheral nerves, positive family history of Hansen's disease are pointers towards diagnosis of Hansen's arthritis.

Conclusion:

In developing country like India, where Hansen' s disease is still an existing entity a combination of tenosynovitis and thickened nerves in association with symmetric polyarthritis should raise a suspicion of Hansen's disease even in the absence of cutaneous features.

References

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