

## Bilateral Symmetric Polyarthrititis with Deformities – Is it Always Connective Tissue Disease ?

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### Abstract

*Leprosy is a chronic granulomatous infectious disease which is caused by Mycobacterium leprae and has varied manifestations including arthritis. Cutaneous and neurological manifestations are classical presentations of leprosy. Musculoskeletal involvement is the third most common manifestation but is less frequently reported. Joint involvement can present as acute symmetrical polyarthrititis or chronic polyarthrititis resembling rheumatoid arthritis. Leprosy is seen in high prevalence in some developing countries however, it is rarely seen in non-endemic regions. Arthritis can be present in all types of leprosy. Chronic arthritis is known to exist even in paucibacillary forms, resolved or treated disease and in patients without reaction, suggesting a perpetuated inflammatory process. In these cases leprosy can mimic some autoimmune diseases such as rheumatoid arthritis.*

**Key words:** *Leprosy, arthritis, charcots arthropathy, arthritis mutilans, arthritis robustus, connective tissue disease.*

### Case report

A 53-year-old male patient, resident of Delhi, car driver by occupation and without any previous co-morbidity, presented with chief complaints of polyarthrititis with severe deformities in all four limbs involving small joints of both hands and feet since two years. These deformities were however painless and there was no history of early morning stiffness. He also complained of non-healing deep ulcers on the dorsal surface of his right lower limb around the medial malleolus, on the heel and over the ball of great toe since last 6 months. There was no history of fever. There was no evidence of psoriasis or other skin disease and the past and family history of psoriasis was negative. There was no history of diabetes or tuberculosis. He was treated

previously as arthritis robustus but to no effect.

He sought consultation in surgery and dermatology clinic for non-healing ulcers and was treated for unknown reasons as mucormycosis although all routine and special investigations, including nerve and ulcer edge biopsy, were inconclusive. He didn't improve, and was referred to our rheumatology clinic to evaluate the cause of joint deformity. On physical examination, vitals were within normal limits. Musculoskeletal examination revealed thinning of the thenar and hypothenar eminences. There was flexion deformity with contractures at proximal and distal inter phalangeal joints involving 3rd, 4th and 5th digits of right hand, 5th digit of left hand (Fig. 1 & 2) and 2nd, 3rd, 4th and 5th digits of bilateral lower limbs (Fig. 3). In lower limbs, deformity



**Fig. 1 and 2:** Showing joint deformity and ulcer on both hands.

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was also associated with mild synovitis and resorption of digits and there was generalised skin erythema with multiple discharging sinuses over the ulcer area. There was no involvement of axial skeleton. His cardiovascular, respiratory and abdominal examination were within normal limits. His neurological examination was grossly normal and there was no thickening of peripheral nerves on palpation.



**Fig. 3:** Showing flexion deformity and contracture in left foot.

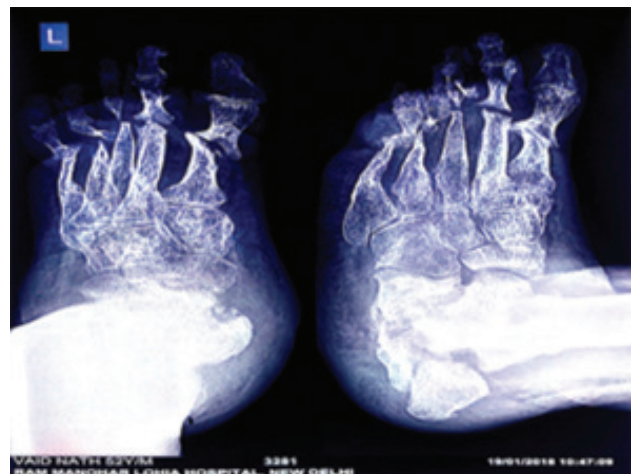
Laboratory investigations revealed haemoglobin of 14.7 g/dl, WBC count of 7,000 mm<sup>3</sup>/cumm<sup>3</sup> with normal platelet counts. Liver and kidney function tests were normal. Inflammatory markers were raised (CRP of 10.5 mg/l and ESR of 38 mm/hr). Urine examination was essentially normal. His rheumatoid factor, anti CCP, ANA, anti dsDNA, extracted nuclear antigen profile were negative.

Ulcer swab for nocardiosis, histoplasmosis and mucoromycosis was negative. Venous and arterial doppler of upper and lower limb were normal. Radiograph of right foot showed marked osteopenia with resorption of metatarsals with multiple fractures and destruction of calcaneum. X-ray of left hand showed flexion deformity at DIP and PIP (Fig. 4 and 5). So a provisional diagnosis of polyarthritis with non-healing ulcer (probably vasculitis associated arthritis *versus* arthritis mutilans *versus* arthritis robustus) was made. The patient was kept on antibiotics and antifungals but did not get relief. DEXA scan was suggestive of osteopenia, however nerve conduction velocities revealed sensory neuropathy in all four limbs.

So a repeat biopsy was taken from left ulnar and left peroneal nerve and from the edge of ulcer at deformity which showed well defined granuloma, lymphocytic infiltration of perineurium and endoneurium with *Mycobacterium leprae* bacterial index of (2+). He was immediately started on rifampicin 600 mg monthly and



**Fig. 4:** Showing mild synovitis and resorption of digits along with flexion deformity.



**Fig. 5:** Showing deformity and resorption of toes.

dapsone 100 mg daily along with wound hygiene. His ulcers began to heal within a month and completely healed in 3 - 4 months. Meanwhile his contractures also got improved and there was no new joint deformity. He has been kept on follow-up and is doing well since then. He has been referred to physiotherapy out-patient department for further support.

## Discussion

Leprosy is a disease, more common in endemic areas and developing countries such as India, although now it is more found in only certain pockets of country. Because of increased travel in today's world, it is possible that we may come across patients of leprosy in non-endemic areas. It is important to be updated about different manifestations of

leprosy in order to avoid unnecessary diagnostic tests and start proper treatment, as early as possible.

Exact pathogenesis of the joint involvement in leprosy is still not fully elucidated. Lepra reactions<sup>1,2</sup> (Types I and II lepra reaction), and direct infiltration<sup>2</sup> of the synovium by bacilli are thought to be the underlying mechanisms of joint involvement.

Chauhan *et al*<sup>3</sup> classified the arthritis in leprosy into the following groups: (1) Charcot's arthropathy secondary to peripheral sensory neuropathy; (2) swollen hands and feet syndrome; (3) acute polyarthritis of lepra reaction; and (4) chronic arthritis from direct infiltration of the synovium by lepra bacilli.

Shiva Prasad *et al*<sup>4</sup> in his retrospective case series study of 44 patients of leprosy with musculoskeletal manifestations revealed arthritis (n = 22), swollen hands and feet syndrome (n = 11), tenosynovitis (n = 9), painful swollen feet (n = 9), arthralgia (n = 7) as the commonest manifestations and vasculitis was seen in only one patient. Arthritis and tenosynovitis were part of spontaneous onset lepra reaction in 28 cases.

Atkin *et al*<sup>5</sup> studied 77 patients with leprosy and found that ten patients had generalised enthesitis and twenty patients had leprosy with manifestations of arthritis.

Although metropolitan cities like Delhi may have very few patients with leprosy, it is important to include leprosy in the list of possible differential diagnoses of arthritis, in India

where leprosy is prevalent or in those patients who have travelled from endemic areas. Sometime, it is difficult to differentiate symmetrical polyarthritis due to leprosy from rheumatoid arthritis as in our patient. However, seronegative male patients without pain, early morning stiffness, rheumatoid nodule and poor response to treatment with DMARDs are the clinical features distinguishing from rheumatoid arthritis.

## Conclusion

Presentation of leprosy as a chronic polyarthritis and multiple deformities is unusual. This case shows that leprosy can present without skin manifestations and thickened nerves but with severe disabling, deforming chronic arthritis or Charcot's arthropathy.

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