

CASE REPORT

Lymph node abscess and cardiac involvement in a patient with nodular lepromatous leprosy (LL) with erythema nodosum leprosum (ENL): A rare occurrence

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Summary

With the world's focus on reducing the leprosy patient load to the extent of elimination, finding and reporting the rarer presentations of leprosy becomes important for prompt treatment. Also, these untreated patients may serve as a potential source of infection in community. We report a 35-year old man diagnosed to have lepromatous leprosy and erythema nodosum leprosum with inguinal lymph node abscess and suspected cardiac involvement that proved fatal. We stress the importance of detailed workup to look for associated systemic involvement for timely intervention and favourable outcome.

Case Report

A 35-year old married, man, resident of a village in Uttar Pradesh, North India, labourer by profession, presented to the emergency department with fever, bone and joint pain, muscle aches, left inguinal abscess and numerous cutaneous nodules all over the body. The nodules were present for the last 4 years with gradual progression and were asymptomatic to begin with. The crops of reddish painful nodules started appearing 8 weeks before presentation and were associated with fever, pain in the groin and adjoining scrotal area.

Subsequently, the swelling (abscess) in the left groin burst open, 2 weeks before presentation, with oozing of pus and blood. He also reported to have markedly diminished sensation and weakness, with inability to perform fine manual movements, but without any

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ulceration of the digits. He was under various treatments in his village, details of which were unavailable. The possibility of injudicious use of antibiotics could not be ruled out.

Examination revealed a temperature of 102° F, normal blood pressure, pulse rate of 56 per minute and respiratory rate of 34 per minute. He complained of high grade evening fevers in previous weeks, but it was now continuous. There was presence of oedema in the feet and lower legs that was woody hard in consistency. Cutaneous examination of the face revealed lesions infiltrating the eyelid margins, ears and forehead, leading to accentuation of forehead and other facial folds. The patient had ciliary and superciliary madarosis with loss of the lateral third of the eyebrows. Additionally there were innumerable, fleshy, skin coloured to erythematous papulo-nodular lesions all over the body including face, chest, back, abdomen, gluteal region, upper and lower extremities (Figure 1).

The body folds, scalp, inguinal region, palms and soles were spared. In addition, there were bright red, erythematous, evanescent tender nodules distributed predominantly on the extremities, face and lower back along with post inflammatory hyperpigmentation. The hard palate and nasal septum were intact. There was a large ulcerating, necrotic lesion in left inguinal region overlying left inguinal lymph node (Figure 2).



Figure 1. A - Face of the patient showing coalescing papulo-plaque lesions on the forehead, eyebrows, ears, nose and cheeks. There was marked induration and loss of eyebrows. B - The back and ears show the extensive lesions. Scalp has patchy hair loss and the back shows lesions with dry, ichthyotic-looking skin.

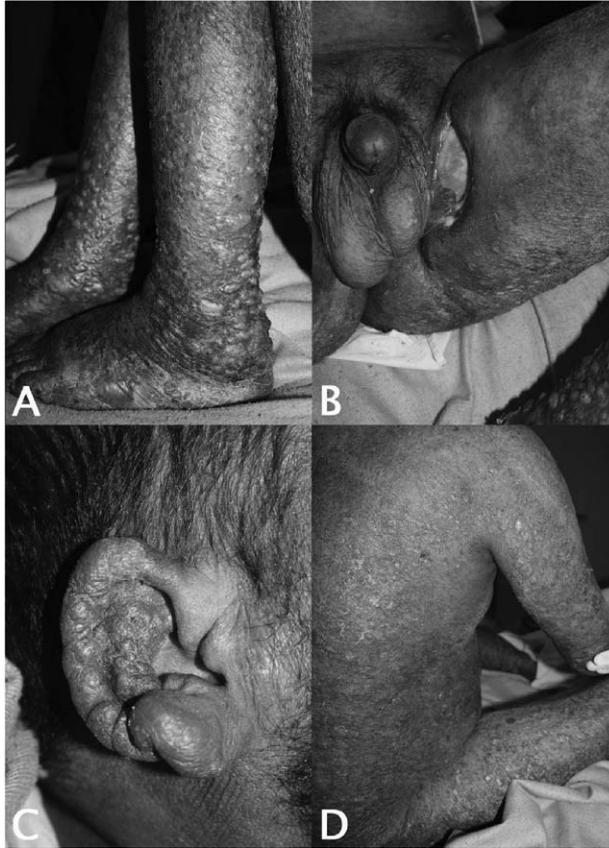


Figure 2. A - Lower extremities showing taut shiny oedematous skin with multiple erythematous and skin coloured papulo-nodular lesions. B - Inguinal abscess on right side with lesions on thighs and very few but discrete lesions on scrotum. Note the sparing of inguinal folds. C - Close up view of ear showing cauliflower ear due to lepromatous leprosy and ENL lesions. D - Whole body showing dry ichthyotic skin with papulo-plaque lesions. Note the sparing of warmer axillary regions.

Bilateral ulnar, radial and common peroneal nerves were uniformly thickened and tender. Bilateral glove and stocking anaesthesia was present up to just below the knee. His haematological investigations showed anaemia with a haemoglobin of 8.6 gm%, neutrophilic leucocytosis ($19,700/\text{mm}^3$) and erythrocyte sedimentation rate of 34 mm in 1st hr. His liver function tests were deranged with serum bilirubin of 3.0 mg/dL and SOT/SGPT 238/246 IU/L respectively. ELISA for HIV was negative. The chest X-ray showed cardiomegaly. The ECG findings revealed supraventricular heart rhythm, with left bundle branch block. The ultrasound revealed mild hepatosplenomegaly with right inguinal lymph node enlargement with an abscess in the left inguinal region.

Based on the clinical features and aforesaid investigations, the differential diagnoses of nodular/histoid leprosy, reticulohistiocytosis, subcutaneous sarcoidosis, cutaneous localised leishmaniasis, lymphocytoma cutis, mycosis fungoides and post kala azar dermal Leishmaniasis were considered. The presence of nerve thickening and glove and stocking anaesthesia was strongly in favour of Hansen's disease.

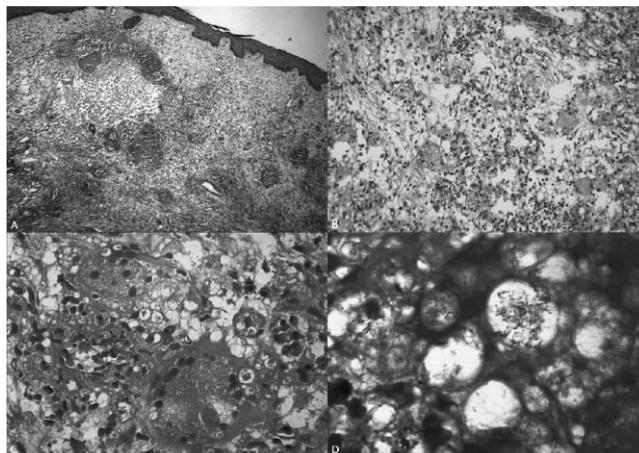


Figure 3. A - Low power view showing foamy macrophage granulomas with peripheral neutrophils and vasculitis. B - Medium power view showing foamy macrophages, giant cells, vasculitis with polymorphonuclear infiltrate. C - High power view showing very clear giant cells, foamy macrophages and PMNs. D - Modified Wade-Fite stain showing leprosy bacilli in clumps with few peripheral bacilli showing BI +6

A slit skin smear was then performed, revealing AFB in globi on ZN staining with a Bacteriological Index (BI) 6 + and morphological index of 10%. (Figure 3)

A biopsy from the tender ENL nodule revealed epidermal atrophy, with foamy macrophages forming large, loose granulomas surrounded by dense neutrophilic infiltrate. The macrophages infiltrated deep down into the dermis. Medium to small sized vessels showed fibrinoid necrosis with plump endothelial cells and neutrophils. At places the foamy macrophages are seen forming giant cells. Modified Wade-Fite stain showed the BI as 6+ . (Figure 3)

Based on clinical, biopsy and slit skin smear evidence, he was diagnosed to be a case of lepromatous leprosy with erythema nodosum leprosum, with ruptured left inguinal lymph node abscess, but with no visible deformity. The WHO regime of MDT-MB for Hansen's disease was proposed, but unfortunately he succumbed to cardiac failure with left bundle branch block before therapy could be started.

Discussion

Lymph node involvement in leprosy is well documented both in reactional and as well as non-reactional states. It was first reported on cytological diagnosis by Cavett *et al.*¹ Subsequently, Kar *et al.*² from India reported on the frequency of inguinal, cervical, axillary, epitrochlear and pre-auricular lymph node involvement (with figures of 76%, 70%, 70%, 65% and 10% respectively) on biopsy. *M. leprae* has been demonstrated in lymph node aspirates from leprosy patients. While in tuberculoid leprosy, regional lymph nodes are involved, there is extensive and even visceral lymphatic involvement towards the lepromatous pole.² In another study involving 26 patients with tuberculoid leprosy, 12% showed the presence of AFB in lymph nodes.³ In 18 patients with borderline tuberculoid (BT) leprosy, 61% showed granuloma and 11% showed the presence of AFB in lymph nodes.³ The involved nodes are usually without matting or suppuration. Generalised lymphadenopathy has also been reported with Type 2 reaction.⁴

Our case highlights the importance of lymphatic involvement, even to the extent of inguinal abscess formation, which is relatively rare; as noted above, the patient was HIV negative.

Cardiac involvement is a known complication of leprosy.⁵ Cardiac dysautonomia may present as abnormal responses in heart rate and blood pressure on standing, or the absence of responses to perioperative manipulations such as intubation, extubation or anticholinergic drugs, or an abnormal response to the Valsalva manoeuvre.^{6,7} It is also common to find tachycardia, ST and T wave changes, bundle branch block, extrasystoles and prolongation of QT interval.⁸ The phenomenon of sudden death has been reported in leprosy, particularly amongst lepromatous patients. Lepromatous leprosy (LL) is associated with autoantibody formation⁹ and may result in autonomic nerve dysfunction along with degenerative changes of striated muscle fibres.^{10,11} Although no autopsy was done in this patient, leprosy-related cardiac involvement can be strongly suspected. Such patients must be carefully assessed preoperatively for cardiovascular autonomic dysfunction, if any surgical interventions are planned. We also need to be aware of the phenomenon of sudden death in these patients.

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