CASE REPORT

Type II reaction without erythema nodosum leprosum masquerading as lymphoma

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Summary
Lepromatous leprosy is a multisystem disease that can involve many organ systems, with lymph nodes a common extra-cutaneous site to be affected. Rarely, multibacillary leprosy can be confused with other diseases like lymphomas and connective tissue diseases. Herein we report a patient of lepromatous leprosy with Type II lepra reaction involving lymph nodes who presented with generalised lymphadenopathy, acquired ichthyosis and constitutional symptoms but no cutaneous lesions to suggest erythema nodosum leprosum, and who was initially misdiagnosed as a case of Hodgkin’s lymphoma

Introduction

Mycobacterium leprae, the causative organism of leprosy primarily affects Schwann cells, endothelial cells and monocyte-macrophage system.1 Approximately one-third of patients may present with Type II lepra reaction (erythema nodosum leprosum) at the time of diagnosis.2 Generalised lymphadenopathy as a presenting manifestation of lepromatous leprosy (LL) has been rarely described.3–5 We report a patient having generalised painful/tender lymphadenopathy and acquired ichthyosis who was initially diagnosed with TB Lymphadenitis, then Hodgkin’s lymphoma but later was found to have lepromatous leprosy and Type II Lepra reaction.
Report of a case

A 36-year-old male was referred with recurrent episodes of high-grade fever of 12 months duration, plus burning sensation and pain in his hands and feet for 10 months. During the febrile episodes, the patient experienced associated pain in the calf muscles, redness of eyes, testicular pain and arthralgia. Almost simultaneously he noticed the development of dry, scaly lesions over both lower limbs and upper limbs. After nearly 8 months he developed painful nodular swellings over the axillae and inguinal regions. Fine needle aspiration cytology (FNAC) from the axillary swelling revealed granulomatous pathology and hence, with a possible diagnosis of tuberculosis, he was started empirically on anti-tubercular therapy with no significant clinical improvement. A lymph node biopsy from axilla was reported elsewhere as Hodgkin’s lymphoma. Finally, the patient was referred to the hematology unit of our institute for the management of suspected lymphoma and a dermatology opinion was sought for the management of his skin lesions.

A general physical examination revealed fever (39°C), pallor, hepato-spleno-megaly along with cervical, axillary and inguinal lymphadenopathy. Lymph nodes were multiple, firm to hard in consistency, tender and mobile. Mucocutaneous examination showed diffuse infiltration of the face including ear lobules and skin coloured papules and nodules over the forehead and ear lobes (Figure 1).

Over the upper and lower limbs, areas of ichthyosis were present (Figure 2).

Sensory examination revealed ‘glove and stocking’ anesthesia. Bilateral supra-clavicular, ulnar, median, radial-cutaneous, lateral popliteal and posterior tibial nerves were thickened. Motor examination did not reveal any muscle weakness.

INVESTIGATIONS AND COURSE

A complete blood count showed anemia with no atypical cells on peripheral smear examination. A slit skin smear showed bacteriological index to be 5+ on the Ridley scale and morphological index was 0%. A second FNAC from axillary lymph nodes showed

Figure 1. Diffuse infiltration seen over the face, especially forehead and ear lobes.
organised inflammation. Histopathology of skin biopsy from lower back showed thinned out flattened epidermis; perianal, perivascular and perineural circumscribed epithelioid cell granulomas and Langhan type giant cells with collection of foamy histiocytes in the dermis (Figure 3).

Histopathological examination of axillary lymph node revealed extensive effacement of architecture with numerous epithelioid cell granulomas having central polymorphic collection and nuclear debris and scattered multinucleated giant cells (Figure 4).

No acid fast bacilli (AFB) could be demonstrated on skin or lymph node biopsy specimens. Based on the clinical and histopathological findings, a diagnosis of untreated
LL with Type II reaction involving lymph nodes was made. The patient was started on WHO-recommended multidrug therapy, multibacillary regimen (MDT-MB). For Type II lepra reaction, oral prednisolone (starting at 60 mg/day followed by tapering to 40 mg/day), clofazimine 300 mg/day along with antipyretics were prescribed. Within 2 weeks of starting treatment, he showed significant improvement in his general condition and subsidence of enlarged lymph nodes.

Discussion

Although the clinical diagnosis of leprosy is rarely difficult in the presence of hypopigmented hypo-aesthetic patches, it may pose a diagnostic challenge in certain situations. There are several case reports in literature where leprosy has been confused with other diseases, common mimics being sarcoidosis, cutaneous lymphomas, Jessner’s lymphocytic infiltrate, multicentric reticulohistiocytosis, and connective tissue diseases.

Balachandran et al. have reported two cases of non-Hodgkin’s lymphoma and histiocytic lymphoma simulating lepromatous leprosy. In addition, there are many reports of association of leprosy with lymphomas. Leprosy can also mimic or coexist with connective tissue diseases like systemic lupus erythematosus (SLE) and rheumatoid arthritis. Auto-aggressive hanseniasis refers to a syndrome with immunopathologic and clinical features simulating auto-aggressive diseases. The exact aetiopathogenesis of this condition is not
known but may be caused by B cell stimulation along with a dysfunction of the T-suppressor lymphocytes.14

Lymph node involvement in leprosy can be due to Type II reaction or as a part of visceral involvement in lepromatous leprosy. Satti et al. recognised Hansen’s disease to be an important cause of lymphadenopathy in endemic areas.15 Gupta et al. studied the aspiration cytology of lymph nodes in leprosy and found 51-7% to have Type I and 25% to have Type II pattern with both patterns showing predominantly foamy histiocytes and corresponding to lepromatous spectrum of disease. A Type III pattern was seen in 16-6% patients, showing atypical histiocytes due to improved cell mediated immunity and corresponding to borderline spectrum, and another 6-7% patients showed a Type IV pattern with predominant epithelioid histiocytic cell population, analogous to tuberculoid spectrum of Hansen’s disease.16 Rodriguez reported a case of Type II reaction presenting with generalised lymphadenopathy.5 Bagla et al. reported a 70 year old male presenting with generalised lymphadenopathy and a clinical suspicion of Hodgkin’s disease. Histopathologic examination of cervical and axillary lymph nodes revealed aggregates of lepra cells loaded with lepra bacilli.3 Singh et al. reported two cases, one with localised and other with generalised lymphadenopathy who were initially diagnosed as tuberculosis and lymphoma respectively, and later only on lymph node biopsy a diagnosis of leprosy made.4

In the present case, the patient presented with constitutional symptoms, generalised lymphadenopathy, splenomegaly and acquired ichthyosis, all of which could be clinically consistent with lymphoma. However, tender lymphadenopathy is not usually seen in tuberculosis and lymphoma as they are usually painless. Hence, these features should lead the physician to suspect an alternate aetiology. Presence of cutaneous infiltration over face and thickened peripheral nerves along with sensory anesthesia should have raised the suspicion of leprosy, although the typical lesions of erythema nodosum leprosum seen in Type II reaction were missing. Finally, a simple test like slit skin smear should suffice to rule out leprosy aetiology in this clinical situation, along with a search for the other cardinal signs of thickened peripheral nerves or hypo-anaesthetic skin patches and for immunosuppression. The present case serves as an important reminder that the level of suspicion in diagnosing leprosy should remain high when encountering symptoms suggestive of connective tissue diseases or lymphadenopathy especially in regions where leprosy is currently or was earlier endemic.

References

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