CASE REPORT

Anaplastic Large Cell Lymphoma and Lepromatous Leprosy: A Rare Coexistence

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Summary Lepromatous leprosy (LL) has been reported in the literature with Non Hodgkin Lymphoma and rarely with Hodgkin Lymphoma. However, an extensive search of the literature shows no case report describing anaplastic large cell lymphoma (ALCL) in association with LL. We report a case of a young male with LL who was found to have ALCL. This is an interesting case of coexistence of an endemic infectious disease and a rare lymphoma involving the same lymph node, with a brief review of the literature.

Introduction

Leprosy (Hansen’s disease) remains an endemic infectious disease in certain parts of the world, including India, despite several attempts being made for its eradication. Lepromatous leprosy (LL) is at one end of the disease spectrum, where patients present with widespread disease involving skin, upper respiratory tract, eyes, testes, lymph nodes and superficial sensory and motor nerves. The association of leprosy with various visceral and lymphoreticular malignancies is known. We describe a patient with LL who failed to respond to multibacillary therapy (MBT) and developed rare lymphoma subsequently. This case illustrates that persistent lymphadenopathy in a leprosy patient, especially one not responding to therapy, should alert the clinician to considering other possibilities including rare coexistent lesions; the correct diagnosis of which is important for appropriate management and follow up.

CASE REPORT

A 27 year-old male presented in December 2010 to the dermatology out-patient department with raised tender lesions on the trunk and bilateral cervical swellings. Clinical examination
revealed numerous firm skin coloured to erythematous papules predominantly over the limbs bilaterally, and hypo-pigmented, hypoaesthetic and erythematous plaques over the trunk. He had clawing of medial two fingers of both hands, bilateral ulnar nerve thickening and glove and stocking loss of sensation, but the patient had a normal gait. He did not have leonine facies or any testicular atrophy. His past history revealed that he patient presented with similar skin lesions 8 months earlier (April 2010). A skin biopsy was taken, histopathological examination of which showed features of LL and strongly positive Fite stain (BI = 4). He was put on MBMDT regimen (Dapsone, Clofazamine and Rifampicin) but Dapsone was stopped after a few months when he developed pancytopenia and continued on the remaining two drugs. However, he did not respond well as there was no decrease in the skin lesions; rather he developed enlarged cervical and axillary lymph nodes (LN), mild hepatosplenomegaly and moderate pallor. His haemoglobin was 8.5 g/dl with mild leucopenia (2300/cumm) and thrombocytopenia (78000/cumm). Examination of the peripheral blood film showed no atypical cells. Fine needle aspiration cytology (FNAC) was done of the cervical LN which revealed atypical large lymphoid cells with indented, lobulated and doughnut shaped nuclei and frequent mitoses (Figure 1a).

These cytological features were suggestive of a high grade Non-Hodgkin Lymphoma possibly T cell type. A complete excision of the LN was then undertaken. Histological examination showed a complete effacement of architecture by atypical cells showing marked nuclear pleomorphism. These cells had a moderate amount of eosinophilic cytoplasm and a large nucleus with opened up nuclear chromatin and prominent nucleolus. A few of the cells were multinucleated and showed nuclear indentation (horse shoe shaped & kidney shaped nuclei) (Figure 1b).

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**Figure 1.** (a) FNAC smear showing presence of numerous large atypical cells with multinucleation, horseshoe shaped nuclei (May Grunwald Giemsa, 40×), (b) High power view showing presence of these multinucleated atypical cells amidst a polymorphous background (H&E stain, 40×). Inset- showing presence of large foamy histiocytes (lepra cells) (H & E stain, 10×).
Interspersed amongst these atypical cells, were clusters of foamy histiocytes (lepra cells) (Figure 1b, inset). The tumor cells expressed diffuse positivity for CD30 and T-cell marker (CD45RO), focal positivity for Epithelial Membrane Antigen (EMA) (Figure 2a and 2b); and were negative for CD15 and B cell marker (CD20).

A Wade-Fite stain showed a few fragmented bacilli. Thus a final diagnosis of concurrent anaplastic large cell lymphoma (ALCL) with LL was made.

Discussion

Leprosy is an endemic chronic infectious disease caused by *Mycobacterium leprae* (*M. leprae*); the incubation period of which varies from 6 months to several decades. The host immune responses in leprosy determine the spectrum of its clinical presentation. LL is characterised by cellular anergy to lepra bacilli leading to extensive cutaneous lesions, lymphadenitis and systemic involvement. Though infection with *M. leprae* has not been shown to increase the risk of neoplasia, leprosy has been associated with various visceral and lymphoreticular malignancies. Whether leprosy predisposes to, or is merely associated with malignancies, is still a controversial issue which requires a more detailed consideration. Amongst the nonepithelial malignancies, malignant lymphoma (including Hodgkin lymphoma, B and T cell Non-Hodgkin lymphoma) and mycoses fungoides are the common associations. The autopsy series of 252 patients with leprosy reported by Furuta et al. showed presence of malignancy in 85 cases (33·3%). Malignant lymphoma was found in six cases (2·4%) in this series, of which four patients had LL and remaining two were of

![Figure 2.](image-url)
borderline and tuberculoid type each. The authors documented that the increased risk of lymphoma in LL may possibly be related to immunosuppression. Ishida et al. have reported the details of these lymphoma cases. However, further studies need to be done to ascertain as to which types of leprosy showed higher incidence of malignant lesions as no definite mechanism has been described.\(^9\) The literature also documents development of LL in established cases of B and T cell lymphomas.\(^5,7\) According to Sujita et al. a generalised decrease in cellular immunity in lymphomas is postulated to be associated with activation of the dormant bacilli leading to LL.\(^7\) Though various lymphomas have been documented with leprosy, no case of LL with coexisting ALCL has ever been reported in literature to the best of our knowledge.

ALCL is a peripheral T-cell-derived malignancy, representing around 2–3% of all lymphoid neoplasms. It is an aggressive lymphoma which frequently presents in advanced clinical stage with systemic symptoms and extranodal involvement. In a case of leprosy, lymphadenitis can be seen as a part of disease itself or during reaction in patients. In this patient, the persisting skin lesions and development of massively enlarged bilateral cervical and axillary LNs prompted the dermatologist to undertake a more detailed examination, and further work up. Finally, cytological and histopathological examinations of the involved LN led to the diagnosis of ALCL with concurrent lepromatous lymphadenitis. Early FNAC and quick work up of the patient facilitated the management of the patient and prevented further progression of both the diseases. To summarise, concurrent leprosy and ALCL are rare. LL patients can frequently present with generalised lymphadenopathy, thus easily masking an underlying lymphoma. Therefore, clinicians should bear this fact in mind and consider other differential diagnoses as well in leprosy patients presenting with generalised lymphadenopathy.

References