



Case Report

T-cell lymphoblastic lymphoma with generalized lymphadenopathy and bone marrow involvement

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Keywords:

Lymphoma;
Bone marrow;
Lymph node;
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ABSTRACT

A 16-year-old girl presented to the out patient department in Grande International Hospital with complaints of fever since 3 weeks, tonsillitis and multiple lymphadenopathy. Fine needle aspiration cytology from one of the cervical lymph nodes showed atypical lymphoid cells with prominent nucleoli in some cells; hence, a diagnosis of Non Hodgkin lymphoma was made. Bone marrow aspiration and biopsy also showed presence of similar atypical lymphoid cells. One of the lymph nodes was also excised for purpose of immunohistochemistry, and the biopsy showed effacement of lymph node architecture with diffuse proliferation of intermediate sized atypical lymphoid cells. These tumor cells showed positivity for CD3, TdT, and Ki-67. A diagnosis of T-cell lymphoblastic lymphoma was made.

Lymphoblastic lymphomas are rare, accounting for 2% of all lymphomas, of which majority are of T-cell type. T-CLL usually occurs in young males. In addition to lymphadenopathy, other common sites of involvement include mediastinum, CNS and bone marrow. It runs an aggressive course and overall survival of patients is poor.

INTRODUCTION

Lymphomas are less common in Asian patients than most Caucasians. The incidence in South Asians has been shown to be only about 3/100,000 cases, whereas it has been shown to be up to 16/100,000 cases in some European and American populations. Usually, patients present with multiple and large lymphadenopathy along with findings like mediastinal mass and other B symptoms that help us diagnose these cases. After morphological diagnosis of lymphoma has been made, immunohistochemistry should

be done to further subtype the lesion and guide therapy. Most non-Hodgkin lymphomas are of B-cell type while T-cell lymphomas account for only 20% of cases.¹

Following is a case of a rare type of T-cell lymphoma, which was diagnosed by bone marrow aspiration with biopsy and fine needle aspiration cytology and biopsy with immunohistochemistry of lymph nodes.

CASE REPORT

A 16-year-old girl presented to out patient department of Grande International Hospital with fever since 3 weeks. On examination, she had multiple lymphadenopathies along with tonsillitis. She also complained of fever.

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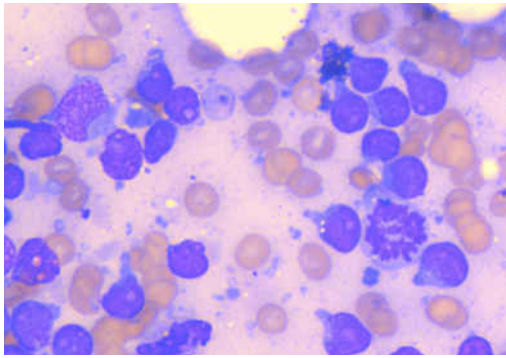


Figure 1: Fine needle aspiration cytology showing atypical lymphoid cells of intermediate size along with a mitotic figure. (Giemsa Stain, X400)

Fine needle aspiration cytology was done from one of the enlarged cervical lymph nodes, which was soft to firm in consistency and about 3x2 cm in size. Cellular material was aspirated and microscopy showed diffusely scattered atypical cells with round to oval to irregular nuclei and prominent nucleoli in some cells. Cells were intermediate to large in size and had moderate amount of cytoplasm. A diagnosis of Non Hodgkin lymphoma was made (fig.1).

Subsequently, the oncologist requested a bone marrow, so aspiration along with biopsy was performed to rule out bone marrow involvement. The peripheral blood counts done at that time showed pancytopenia; however, atypical cells were not seen. Bone marrow aspiration, on the other hand showed scattered atypical lymphoid cells similar to those seen in the lymph node. These atypical cells accounted for 15% of the nucleated marrow cells. (fig.2A) Bone marrow biopsy also showed similar types of atypical cells admixed with marrow cells of all three lineages. There was a diffuse pattern of marrow involvement, without any formation of nodules or paratrabeular aggregates. (fig.2B)

One of the lymph nodes was excised and histopathologic examination showed diffuse loss of lymph node architecture with atypical lymphoid cells of intermediate size, round to

oval to irregular nuclei with prominent small nucleoli in some cells. Cytoplasm was moderate in amount. Occasional mitoses and tingible body macrophages were also seen. (fig.3A)

Immunohistochemistry was also performed (on the lymph node biopsy) which was positive for CD3 and TdT and Ki-67 showed high proliferation rate. Other markers such as CD 10, CD 20, and Bcl-6 were all negative.(fig.3B-E)

Based on all these findings, a diagnosis of TCLL was made.

DISCUSSION

Lymphoblastic lymphoma is a rather rare type of lymphomas, accounting for about 2% of the cases. Of these, 85-90% cases are of T-cell origin and the remainders are of B-cell origin. It occurs more commonly in childhood, adolescence and young adults. Male to female ratio is 2:1.²

Patients with TCLL are younger and have a more extensive disease. There is a higher rate of mediastinal, CNS, bone marrow and stage IV disease. Other less common sites include tonsils, liver, spleen, omentum, skin, and testis.³ Initially, there may be mediastinal and lymph node involvement, but eventually, bone marrow involvement occurs, leading to leukemia like picture.^{3,4} B-cell lymphoblastic lymphoma patients have more of skin, lymph node and bone involvement.⁵

Typical morphological features of these tumor cells are tumor cells of small to medium in size and having scant cytoplasm, convoluted or round nuclear contours, and immature nuclear chromatin with inconspicuous nuclei. Larger blasts, like those seen in this case, may also be present and their nuclei show fine nuclear chromatin with prominent nucleoli.⁶

Immunophenotyping of these TCLL tumor cells show expression of TdT, CD 34, CD 99 and variable CD 2, CD 3, CD 4, CD 5, CD 7, and CD 8.⁶

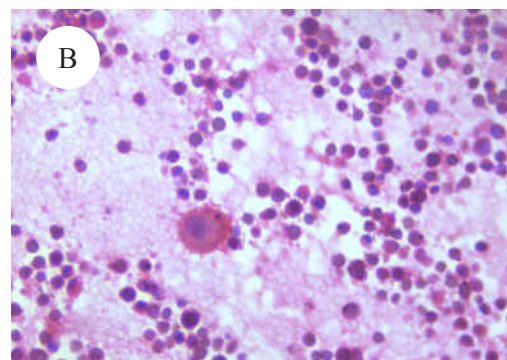
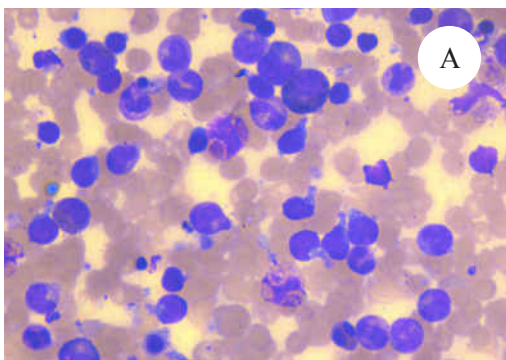


Figure 2A. Bone marrow aspiration showing scattered atypical cells admixed with hematopoietic cells. (Giemsa stain, X100).

Figure 2B. Bone marrow biopsy showing scattered, intermixed atypical lymphoid cells. Note the megakaryocyte and other hematopoietic cells. (Giemsa stain, X100).

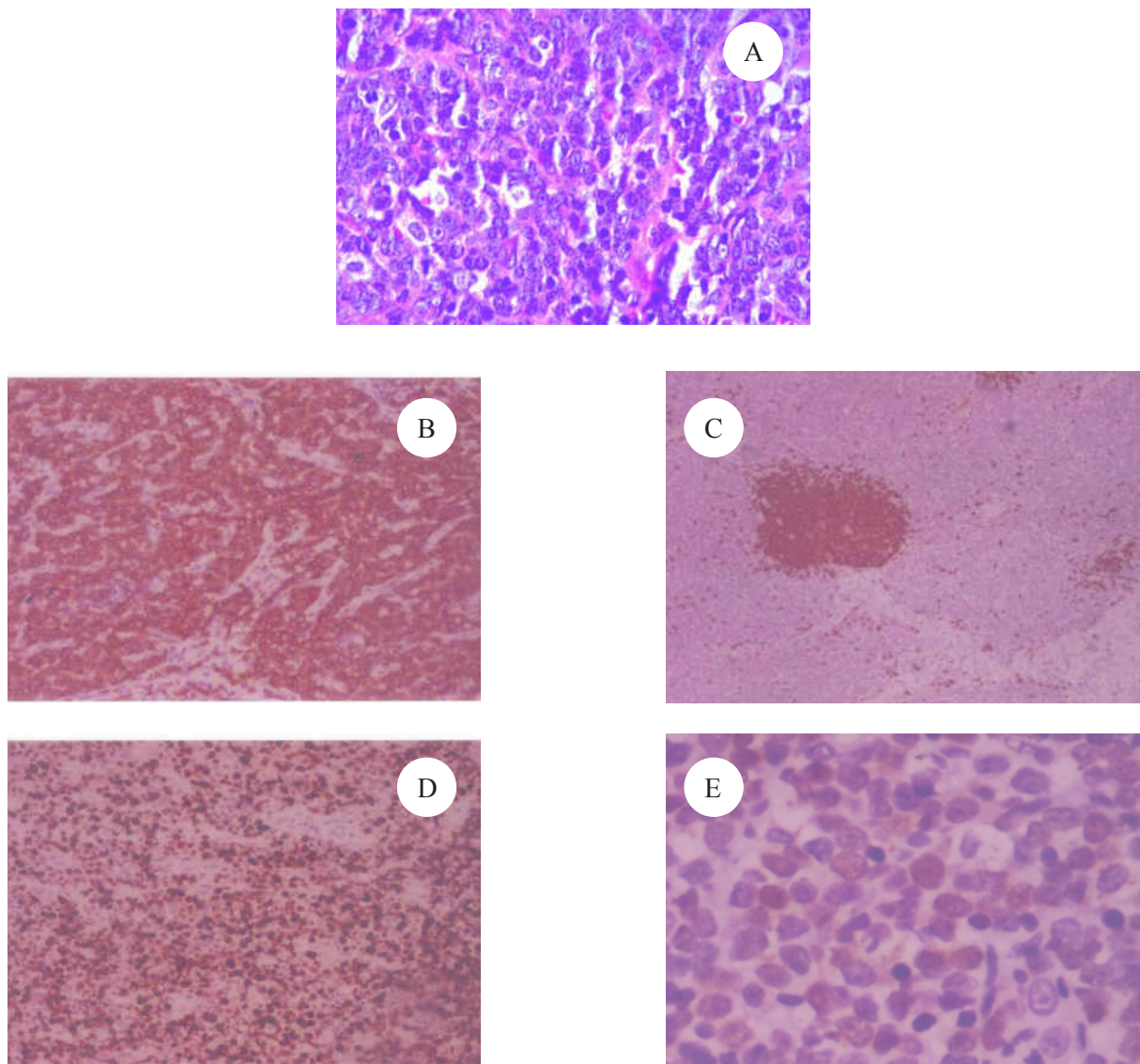


Figure 3. Lymph node biopsy showing diffuse proliferation of intermediate to large sized cells with occasional mitotic figures (A). CD3 shows diffuse positivity (B) whereas CD20 shows only focal positivity (C). Ki67 and TdT show positivity in most of these tumor cells (D and E, respectively).

Morphological differential diagnoses include Burkitt lymphoma, diffuse large B-cell lymphoma, small lymphocytic lymphoma, B1 thymoma, myeloid sarcoma, Ewing sarcoma/PNET, small cell carcinoma, and rhabdomyosarcoma depending on the site and presentation.⁶

TCLL is an aggressive disease with high risk of induction failure and relapse. Bone marrow examination is done for staging purposes in these patients. Overall survival of these patients is extremely poor.^{7,8}

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