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A Rare Site of Presentation of B Cell Lymphoblastic Lymphoma

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Lymphoblastic lymphoma is seen accounting to about 2% of all the lymphomas and it is a neoplasm of precursor B cells. Lymphoblastic lymphoma is highly aggressive, but is now frequently curable with recent therapy. Lymphoblastic lymphoma is associated with radiation exposure and also exposure to pesticide. The lymphoblasts infiltrate nodal or extranodal structures and is commonly associated with large mediastinal masses with a high chances for spreading to bone marrow and the central nervous system. The prognosis in all age groups has drastically improved with the use of intensive ALL-type chemotherapy regimes, with a disease-free survival of 73-90% in children and 45-72% in adults.

Keywords: B. cell; lymph nodes; bone.

1. INTRODUCTION

Approximately one-third of non Hodgkin lymphomas (NHL) arise from sites other than lymph nodes, spleen or the bone marrow. They may also arise from sites normally without lymphocytes [1,2]. The designation of stage III and IV lymphomas as primary extra nodal NHLs is indeed questionable in extra-nodal involvement in the presence of mainly nodal or

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disseminated disease may represent secondary extra-nodal disease spread [3-5]. Currently, it is accepted to define as extra-nodal the lymphomas with nil or only “minor” nodal involvement associated with a clinically dominant extra-nodal component [6,7]. There is no consensus about the staging of primary extra-nodal lymphomas: the Ann Arbor staging system is at present widely used for describing the extent of the disease. However, specific sites of extra-nodal lymphoma involvement may require additional work-up procedures.

2. CASE REPORT

5/F, presented to our OPD with complaints of swelling in the left temporal region for 2 months duration and swelling in the left side of the neck for 2 weeks duration, progressive increase in the size of the swelling was noticed with no discharge or skin changes over swelling. Child also had complaints of high grade, continuous fever for 4 days. No known co-morbids. Normal birth and developmental history are noted. On examination, the Child was moderately built and nourished. Vitals are stable and on local examination:4x3cms non-tender, firm, hyper pigmented, mobile swelling + in the left temporal region which is non pulsatile 5x5cms firm, swelling + in the left cervical region are noted. No discharge seen, non pulsatile, No other nodes were palpable in the neck.

Routine investigations like CBC, urine routine, liver and kidney function tests were done which showed leucocytosis with other parameters within normal limits.

Child was evaluated and FNAC of the left temporal swelling was done which showed smear with lymphocytes and smudge cells on a background showing blood. No keratin seen.Child was then planned for left temporal swelling excision and biopsy with split skin grafting and excision biopsy of left cervical region swelling.Lesion over left temporal region was seen well above the pericranium but adherent to the skin and subcutaneous tissue, cross section appeared like fish flesh. SSG was done for the raw area.Specimen sent for HPE and immunehisto-chemistry.Biopsy report concluded to be B cell lymphoblastic lymphoma positive for markers Tdt, CD79a, CD45, CD43 and negative for CD3, CD20. Child was then started on T. Prednisolone 60mg, Inj. Vincristine, Inj. Daunorubicin, Inj.L Asparaginase, Inj. Cytarabine.

Treatment was taken for 6 months and then FDG-PET was done which showed complete metabolic response to the therapy.

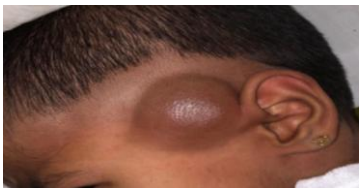
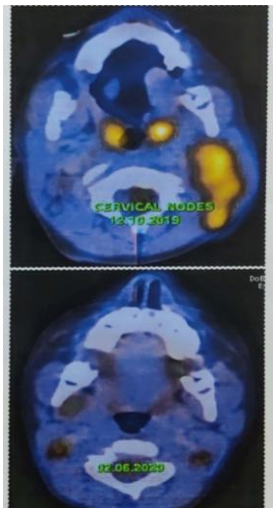


Fig. 1. B cell lymphoblastic lymphoma



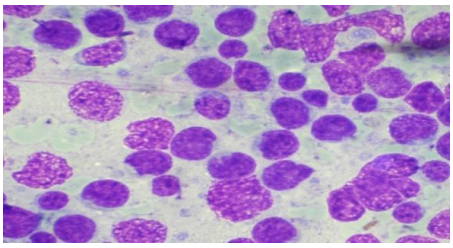


Fig. 2. FNAC of the left temporal swelling

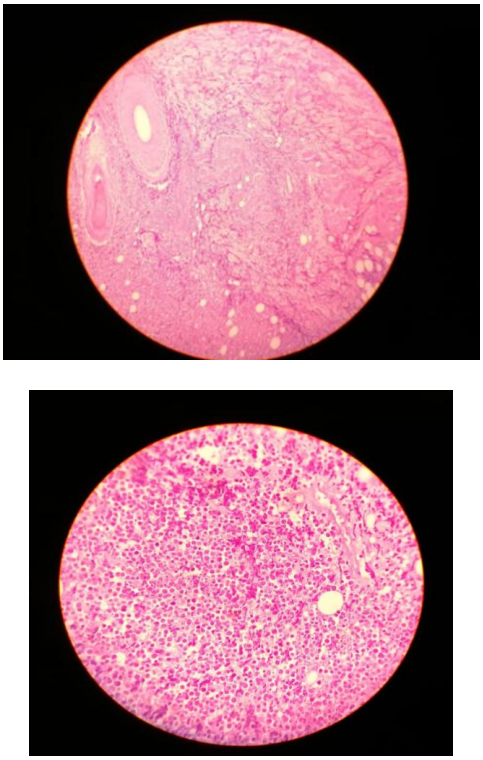
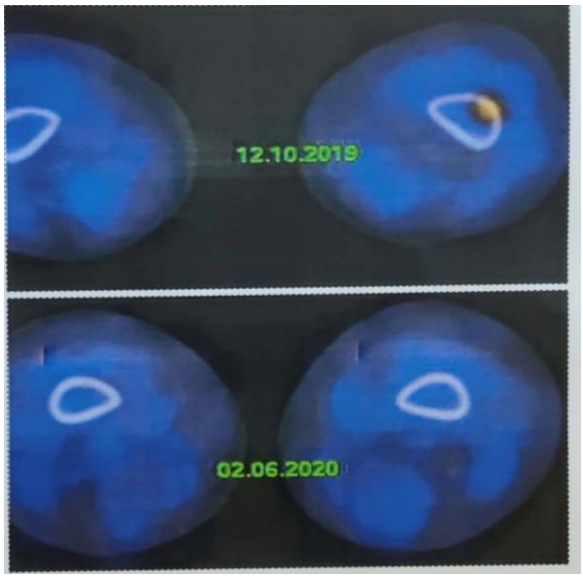


Fig. 3. Histopathology



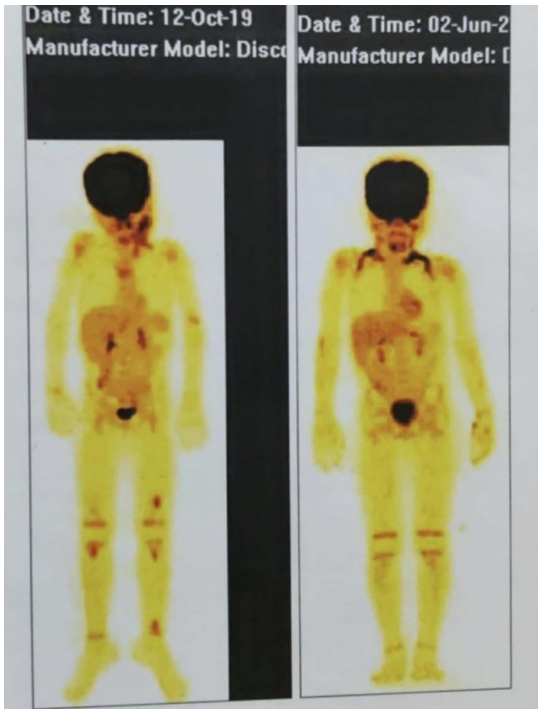


Fig. 4. Model assay

3. DISCUSSION

B cell lymphoblastic lymphoma is a rare type of fast growing non-Hodgkin lymphoma.B cell type is is commonly extranodal with skin being involved most commonly (33%) followed by lymph nodes (22%), bone (19%) and mediastinum (5%). Lymphoblastic lymphoma has a high predilection for the bone marrow, with a reported frequency at diagnosis of 21% as well as a reported frequency of 5–10% for the central nervous system. Central nervous system involvement is more commonly associated with relapse, especially in cases without adequate central nervous system prophylaxis [3]. Some of the rarely involved sites include liver, spleen, and testes. This is a rare presentation of lymphoblastic lymphoma where the child presented with a swelling in the left temporal region and underwent excision biopsy with split skin grafting. They are positive for CD10, CD 24, PAX5, and TdT in most cases, while the expression of CD20 and the lineage independent stem cell antigen CD34 is variable and CD45 may be absent. Surface immunoglobulin is usually absent. In T-LBL, neoplastic cells are usually TdT positive and variablyexpress CD1a, CD2, CD3, CD4, CD5, CD7 and CD8. Child underwent chemotherapy for about six months and is in the phase of remission now.

4. CONCLUSION

Thus, we conclude that about 90% of the B cell lymphoblastic lymphomas are curable with regular treatment and close follow up.

CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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