A RARE SITE OF PRESENTATION OF B CELL LYMPHOBLASTIC LYMPHOMA

ABSTRACT:

Lymphoblastic lymphoma is a neoplasm of immature B cells that accounts for approximately 2% of all the lymphomas. LBL is highly aggressive, but frequently curable with current therapy. The prognosis in all age groups has dramatically 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.

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 devoid of lymphocytes. 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 disseminated disease may represent secondary extra-nodal disease spread. Currently, it is accepted to operationally define as extra-nodal those lymphomas with no or only “minor” nodal involvement associated with a clinically dominant extra-nodal component. As for the definition, 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.

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.

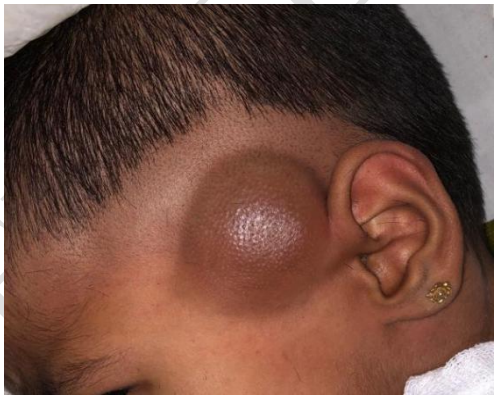

Fig. 1. B CELL LYMPHOBLASTIC LYMPHOMA

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.

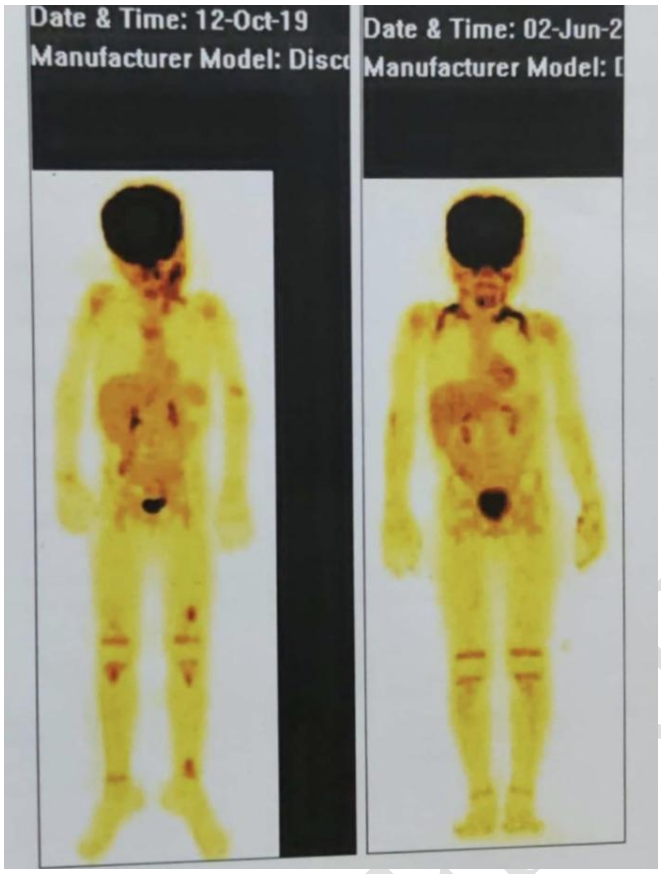

Fig. 2. FNAC of the left temporal swelling

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

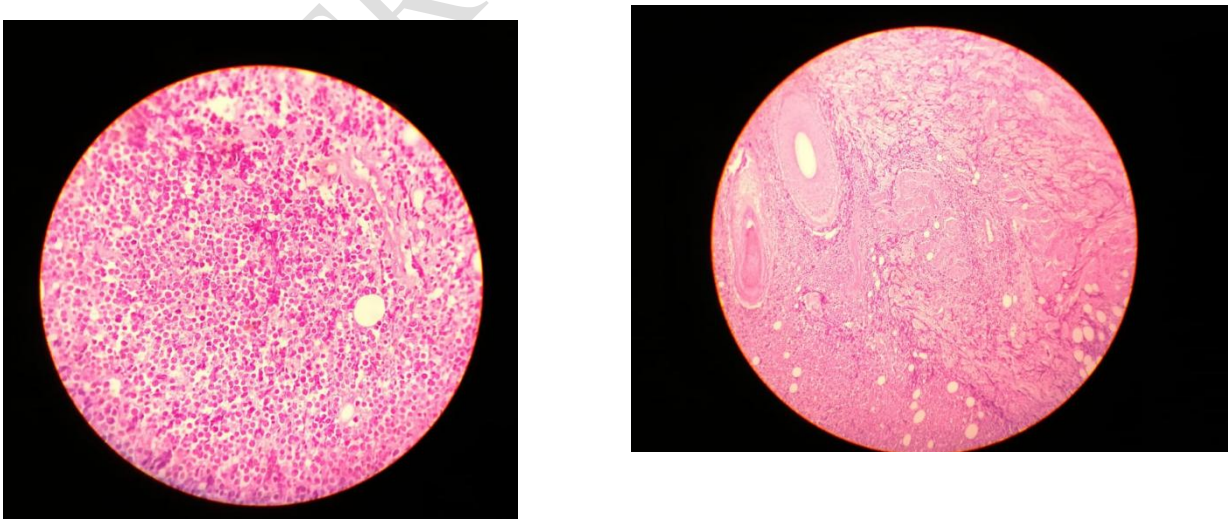

Fig. 3. Histopathology

Fig. 4. Model assay

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 preference 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]. Liver, spleen, and testes are some of the other sites that are rarely involved. 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 variably express CD1a, CD2, CD3, CD4, CD5, CD7 and CD8. Child underwent chemotherapy for about six months and is in the phase of remission now.

CONCLUSION:

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

REFERENCES:

1. Lymphoblastic lymphoma, Sergio Cortelazzo et al. Crit Rev Oncol Hematol. 2011 Sep.

2. A Rare Presentation of Precursor B-cell Lymphoblastic Lymphoma in a Child Zarmina Javed and Faiza Hanif

3. Precursor B-cell Lymphoblastic Lymphoma A Predominantly Extranodal Tumor With Low Propensity for Leukemic Involvement, Lin, Pei M.D.; Jones, Dan M.D., Ph.D.; Dorfman, David M. M.D., Ph.D.; Medeiros, L. Jeffrey M.D.

4. B-cell lymphoblastic lymphoma: a study of nine cases lacking blood and bone marrow involvement and review of the literature Anirban Maitra, Robert W McKenna, Arthur G Weinberg, Nancy R Schneider, Steven H Kroft

5. Outcome of children with primary resistant or relapsed non-Hodgkin lymphoma and mature B cell leukemia after intensive first line treatment: a population-based analysis of the Austrian Cooperative study group; Paediatric blood cancer 2005;44:pp.70-76, Attarbaschi A, Dworzak M., Steiner M., Urban C., Fink F.M., Reiter A., et al

6. Case series of precursor B-cell lymphoblastic lymphoma, Kim JY, Om SY, Shin S-J, Kim JE, Yoon DH, Suh C. Blood res. 2014;49:270-274

7. Clinical presentation, evolution, and prognosis of precursor B-cell lymphoblastic lymphoma in trials LMT96, EORTC 58881. Ducassou S, Ferlay C, Bergeron C, et al. Br J Haematol. 2011;152:441-451