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EXTRANODAL B CELL LYMPHOBLASTIC LYMPHOMA PRESENTING AS A LARGE CHEST WALL MASS

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Abstract- Precursor B-cell lymphoblastic lymphoma (PLBL) is an uncommon high-grade neoplasm of immature B cell type. It is most commonly prevalent in children and adolescents with extranodal involvement such as skin and bone. We report a case of 48 year old man with PLBL presenting as a large chest wall mass without any systemic symptoms. The patient was treated with neoadjuvant radiotherapy followed by chemotherapy with cyclophosphamide, hydroxyzine, oncovin, prednisolone (CHOP). The patient is still under treatment and follow up.

Keywords- Precursor B- cell lymphoblastic lymphoma, Extranodal, Chest Wall, Adult

Introduction

Chest wall tumors arise from a wide variety of benign and malignant pathologies. The majority of chest wall lesions are the result of metastasis or invasion from adjacent malignancies. Primary chest wall lymphoma is an uncommon tumor which account for less than 2% of all chest wall soft tissue tumors [1]. Lymphoblastic lymphoma (LBL) is a highly aggressive subtype of non-Hodgkin's lymphoma (NHL), and accounts for about 2% of all NHLs. The majority of acute lymphoblastic leukemias (ALLs) are of immature B-cell phenotype, whereas approximately 90% of LBL are of immature T-cell phenotype. The natural history of B cell lymphoblastic lymphoma B-LBL has not been well defined and molecular studies about its pathogenicity are still lacking [2]. The most common age of presentation is in persons younger than 18 years. The most common presentation of B-LBL is extranodal involvement sparing the bone marrow. The most commonly affected sites are skin (33%), lymph node (22%), bone (19%), and mediastinum (5%) [2].

Material and Methods

A 48-year-old male presented with a mass on the right side of the chest wall involving the right axilla since last six months. The mass was approximately 20x15cm in size, tender, non-itching, with ulcerated surface. There were no palpable lymph nodes or any other skin lesions in any region including the neck and groin. There was no history of fever, vomiting, pain abdomen, headache, bleeding, weight loss, seizures, respiratory or urinary complications. There was also not any history of exposure to drugs. Rest of his personal and family history was non contributory. On examination there was no jaundice, lymphadenopathy, hepatosplenomegaly or any bone tenderness. The systemic examination was unremarkable. Routine investigations revealed white blood cell count- 6.78×10^9/L, with normal differential count: platelet count -243×10^9/L, and haemoglobin -12.6 g/dL . Erythrocyte sedimentation rate was 24mm, aspartate transaminase 8 U/L, alanine transaminase 15 U/L, uric acid 6.7 mg/dL, lactate dehydrogenase 286 U/L, and alkaline phosphatase 57 U/L. Chest radiography revealed a homogenous white opacity of size 17x15cm with well defined borders on right chest wall. Ultrasonography of chestwall showed large hypoechoic lesion of size 17x13.5cm showing flow on color doppler with? underlying bone destruction. Computed topography scan of chest was advised but patient refused since he could not afford it.

Result

Fine needle aspiration cytology of the lesion showed singly scattered malignant small round cells with round to oval pleomorphic nucleus having negligible to scant cytoplasm and inconspicuous nucleoli. Cytological features were suggestive of malignant round cell tumor with possibility of round cell sarcoma or high grade lymphoma [Fig-1]. Biopsy of chest wall mass revealed small malignant cells with stippled chromatin, convoluted nuclear membranes and scant cytoplasm [Fig-2]. A large number of mitotic figures were also observed. On immunohistochemistry, the tumor cells were shown to express Tdt (patchy positive), CD99 (positive), CD20 (strongly positive), CD10 (focally positive), Vimentin (strongly positive) and were negative for LCA and CK ,suggesting a Pre B phenotype [Fig-3] and [Fig-4]. These finding were consistent with non-Hodgkin lymphoma (NHL) of the diffuse, high grade lymphoblastic type. The patient was advised to undergo radiochemotherapy.

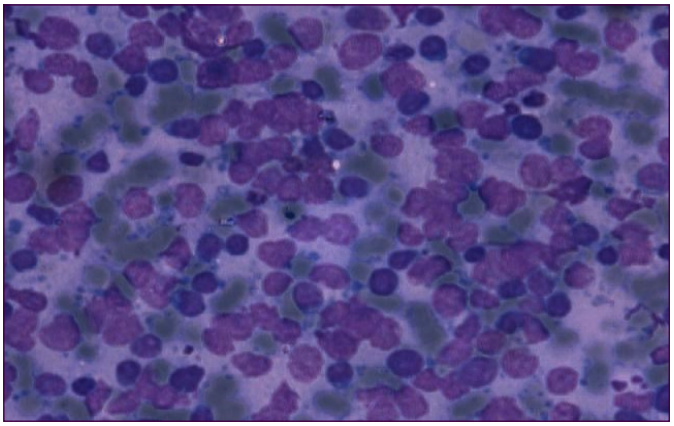

Fig. 1- Cytology smear showing small round cell tumor (Leishman Stain; 400x).

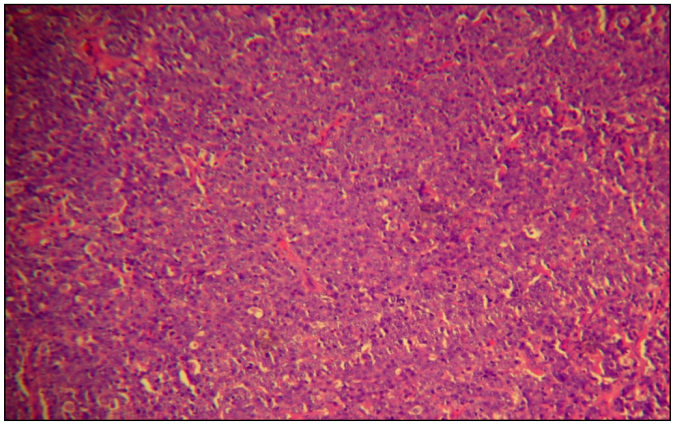

Fig. 2- Histopathological Section showing NHL of diffuse,high grade . (Hematoxylin and eosin stain; 400x).

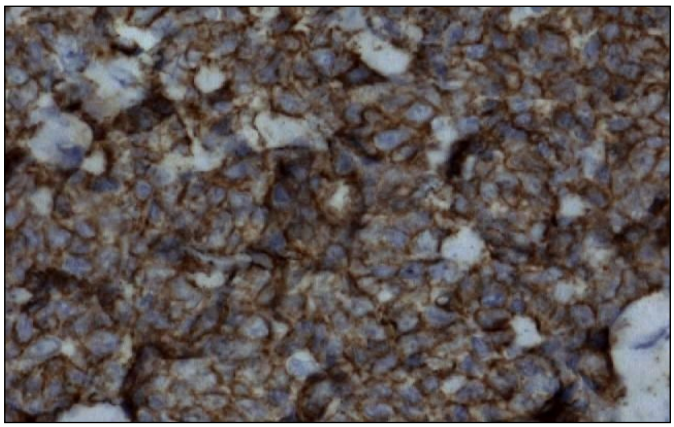

Fig. 3- Immunohistochemistry showing membranous positivity for CD20 (400x).

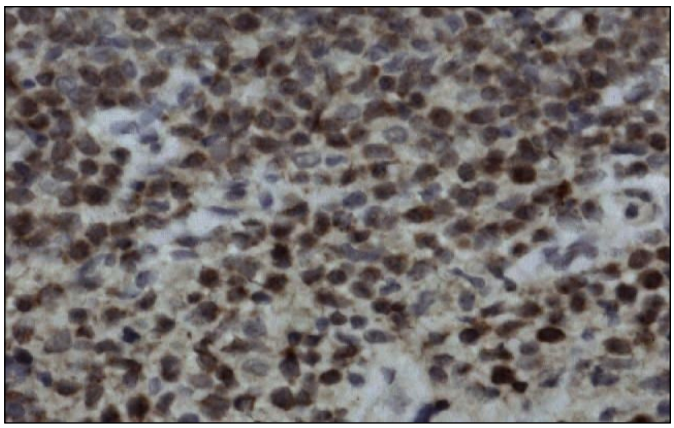

Fig. 4- Immunohistochemistry showing nuclear positivity of Tdt (400x)

Discussion

Pre B-LBL is an uncommon high-grade neoplasm of immature B-cells. It is a neoplasm of cells frozen at the level of immature lymphoid, either T lineage thymocytes or immature B- lineage bone marrow lymphoid cells. In the WHO classification, it is separated into B- cell and T -cell type lymphomas and is called precursor lymphoblastic lymphoma /lymphoblastic leukemia. It can involve blood, bone marrow and occasionally nodal/extranodal involvement (B-LBL/LBL). By convention, the term 'lymphoma' is used when the process is confined to a mass lesion with no evidence of peripheral blood involvement and no less than 25% blasts in the bone marrow [3]. Majority (85%) of the acute lymphoblastic leukaemia tumors belong to the B-cell lineage, whereas most of the LBL express T cell markers [3]. Lymphoblastic lymphoma predominates in young adults and adolescents with a median age at the time of diagnosis of 20 years (in adults: median age, 27 years for men; 50 years for women) and a slight male predominance (male-to-female ratio, 2:1) with a tropism for the head and neck region. Interestingly in our patient, the lesion was located in chest wall. These tumors show a rapid progression often with involvement of the peripheral blood and bone marrow, central nervous system and gonads. Like all other non-Hodgkin lymphomas, lymphoblastic lymphoma is also associated with exposure to radiation or pesticides and congenital or acquired immunosuppression. Lymphoblastic lymphoma is highly aggressive, presenting as stage IV disease in more than 70% of the patients. Immunity is hampered by gross lymphadenopathy which allows development of opportunistic infections. Lymph nodes may be enlarged to the extent of compressing the adjacent structures. In 30-50% of patients, the lymphoblasts infiltrate bone marrow causing ineffective hematopoiesis. Histologically, they show diffuse effacement of normal nodal architecture with capsular destruction and extensive infiltration of adjacent soft tissues in single file cell pattern. Mitotic rate is also high and a starry sky pattern may be present focally.

Pre B-LBL may present a diagnostic challenge due to absence of mature B cell markers like CD20. Thus, it must be distinguished from other high grade lymphoid tumors, myeloid sarcoma, thymoma and other small blue round cell tumors including neurublastoma, Ewing's sarcoma/peripheral neuroectodermal tumor (PNET), Merkel cell carcinoma, rhabdomyosarcoma, Wilms tumor, neuroendocrine carcinoma and metastatic small cell carcinoma [2]. Since no single diagnostic criteria has been established for Pre B LBL, comprehensive immunohistochemistry (IHC) staining is crucial for final diagnosis [2]. Tdt is an extremely useful marker as it is positive in virtually all cases of lymphoblastic lymphomas, but negative in other types of NHL. B-LBL should also be distinguished from Burkitt type lymphoma and blastic variant of mantle cell lymphoma [4]. Myeloid sarcoma can be distinguished by the presence of eosinophils, scant cytoplasm and MPO positivity since occasionally these may be Tdt positive causing diagnostic challenge. Thymomas can be distinguished by the presence of large epithelial cells with vesicular chromatin and keratin positivity. Small blue round cell tumors especially Ewings sarcoma is characterized by sheets of uniform small round cells that are slightly larger than lymphocytes with scant cytoplasm which may appear clear because of glycogen. They are CD99 positive, Tdt negative. The remaining other small round cell tumors can be differentiated by characteristic immunohistochemical pattern.

Patients with B-LBL respond well to intensive multiagent chemotherapy with CHOP or CHOP like regimens (ALL type regimens or intensive NHL type regimens) which produces a disease free survival of 60-80% in children and 55-95% in adults [1]. B lymphoblastic lymphomas have a better prognosis with a high remission rate, especially in the early stage disease. With the current treatment regimens, the overall survival rate of 5 years in children with lymphoblastic lymphoma is 80-90% and the overall survival rate in adults is 45-55%. Disease free survival rates of 5 years range from 70% to 90% in children and from 45% to 55% in adults. Adverse prognostic factors include high stage, involvement of the central nervous system and a high serum lactate dehydrogenase level.

Conclusion

The diagnostic rate using fine needle aspiration for the diagnosis of chest wall lymphoma is low and therefore not only histology but comprehensive IHC staining is also crucial for the final diagnosis. Although the primary treatment of choice for such lymphomas with or without chest wall involvement is chemotherapy, surgery followed by chemotherapy can provide satisfactory outcome in some patients in whom chest wall is the only site of involvement.

Conflicts of Interest: None declared.

References

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