

Anaplastic Large Cell Lymphoma presenting as a mediastinal mass – A case report

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Abstract

Lymphoma involving the mediastinum are very rare. We report a case of lymphoma presenting as a mediastinal mass. A 30 years old male presented with history of cold since 3 months with loss of appetite and history of weight loss. Fine needle aspiration cytology from the site revealed anaplastic large cell lymphoma. It was confirmed by immunohistochemistry. The tumor cell was positive for ALK – 1, but negative for CD20. To our knowledge this is a rare case of T cell lymphoma involving mediastinum. Laboratory studies only showed a markedly high lactate dehydrogenase (LDH) level and CT scan revealed heterogeneously enhancing lobulated soft tissue in anterior mediastinal and right hemithorax as described.

Keywords: Anaplastic large cell lymphoma, ALK Positive, Mediastinum.

Introduction

Anaplastic large cell lymphoma is a rare type of lymphoma which usually presents at young age and has a special predilection for cutaneous and extra nodal organs

[1]. Anaplastic large cell lymphoma has a characteristics histology and is differentiated from other lymphoma by CD30 membrane expression. Later it was found that most of this tumor have a balanced translocation t(2;5). This resulted in differentiation of this tumor into two subtypes ALK (Anaplastic lymphoma kinase) positive and ALK negative. ALK positive subtype occurs in younger patients and has a favorable diagnosis [2]. In our case it was Anaplastic large cell lymphoma of mediastinum which was ALK positive.

Case report

A 30 years old male patient presented with complaints of cough, cold since 3 months with loss of appetite and history of weight loss. CT scan revealed heterogeneously enhancing lobulated soft tissue density mass lesion in anterior mediastinum and right hemithorax. It is seen extending from arch of aorta superiorly to aortic valve level inferiorly. It is measuring about 7.8 x 11 x 9.6 cm. Few enlarged supra clavicular and mediastinal lymph nodes are seen, largest measuring 3.9 x 2.7 cm. FNAC was done. FNA smears were

hemorrhagic. Microscopy revealed many singly scattered large cells having moderately pleomorphic nuclei, coarse chromatin, indistinct nucleoli and scant to moderate eosinophilic cytoplasm. Binucleated and multinucleated cells were present. Based on these cytological findings three differentials were kept

- 1) High grade lymphoma.
- 2) Thymoma.
- 3) Metastatic germ cell tumor.

Biopsy and Immunohistochemistry was advised. Biopsy showed aggregates of large sized lymphoid cells having coarse granular chromatin, indistinct nucleoli, and scant cytoplasm. Immunohistochemistry showed positivity for ALK, CD30, CD3 positive and negative for CD20. So final diagnosis of Anaplastic large cell lymphoma (ALK positive) was offered.

Discussion

Anaplastic large cell lymphoma (ALCL) is a lymphoid neoplasm of T or null cell origin and one of the most common forms of peripheral T cell lymphoma. Two subtypes of ALCL are recognized by the World Health Organization classification system: ALK – positive ALCL and ALK – negative ALCL. ALK positive ALCL is associated with translocation involving ALK, the Anaplastic Lymphoma Kinase gene, located on chromosome 2p23. ALCL is a lymphoma most commonly seen in children and young adults, and it has a male predominance. ALCL, ALK positive shows clonal rearrangements of the T- cell receptor genes.

The ALK gene encodes a tyrosine kinase receptor, which is normally silent in lymphoid cells. In the presence of an abnormal fusion partner, most commonly nucleophosmin, a housekeeping gene [t (2,5) (p23; q35)], the formation of the heterodimer results in catalytic activation of the ALK domain and in the oncogenic properties of the ALK protein. ALCL, ALK positive

accounts for approximately 3% of all adult non – Hodgkin lymphomas and 10% to 20% of the childhood lymphomas. ALCL, ALK positive is most frequent in the first three decades of life and has a male predominance [3]. Malignant tumors of mediastinum are rare. Their management is complex and depends mainly on their histologic type, local aggressiveness and possibility or not of chest wall reconstruction [4]. Clinical signs are not specific; chest pain and signs of inflammation are always found. Chest x rays, CT and magnetic resonance imaging give precise information about extension, detect pulmonary metastasis and aid in assessment of mediastinal lymph nodes [5,6]. The diagnosis is usually obtained by surgical biopsy as some authors have reported that needle biopsies may be insufficient because of limited efficacy. However, even surgical biopsies may be uncertain as tissue cannot be distinguished [7,8]. The main predictors of survival in patients with primary systemic ALCL are the ALK status of the tumor and the International Prognostic Index (IPI). Patients with newly diagnosed anaplastic large cell lymphoma (ALCL) are usually treated with an anthracycline- based chemotherapy regimen with the goal of achieving a complete remission. For ALK positive patients, ALK inhibitors Crizotinib or Ceritinib is an option [9].

Complications

Mediastinal T cell lymphoma (ALK positive) complications include Bone marrow suppression, organ dysfunction or adverse effects related to high dose chemotherapy.

We are presenting this case not only for its rarity but to put emphasis on FNAC as initial mode of diagnosis for mediastinal lesion.

Conclusion

Mediastinal lymphomas mimic inflammatory conditions but each lymphoid malignancy is unique and possesses a

characteristic immunophenotype. It may therefore be emphasized that correct immunophenotyping in the context of a panel is indispensable in the diagnosis of lymphoid neoplasm.

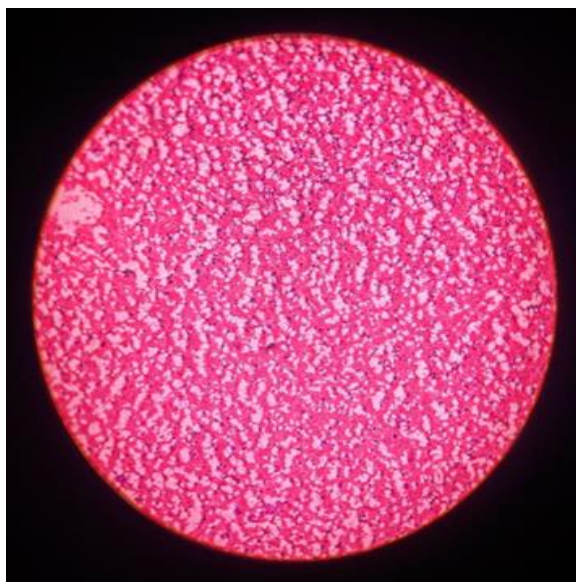


Figure 1: On 10 x examination

This figure shows many singly scattered large neoplastic cells with lymphocytes, RBCs, and atypical figures in the background.

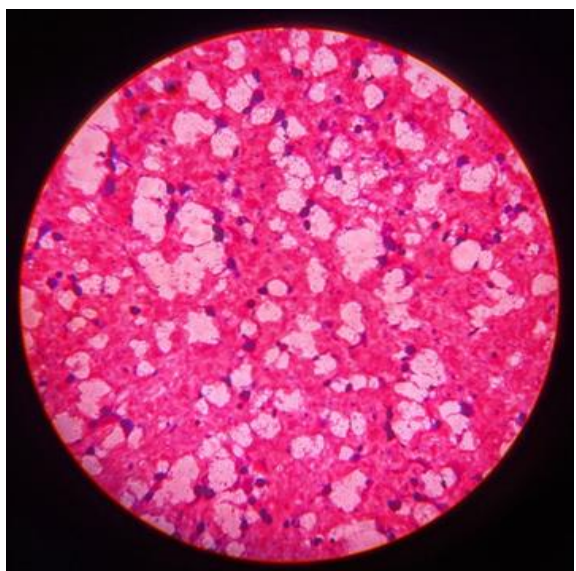


Figure 2: On 40 x examination

This figure shows round to oval cells which have round, moderately pleomorphic nuclei.

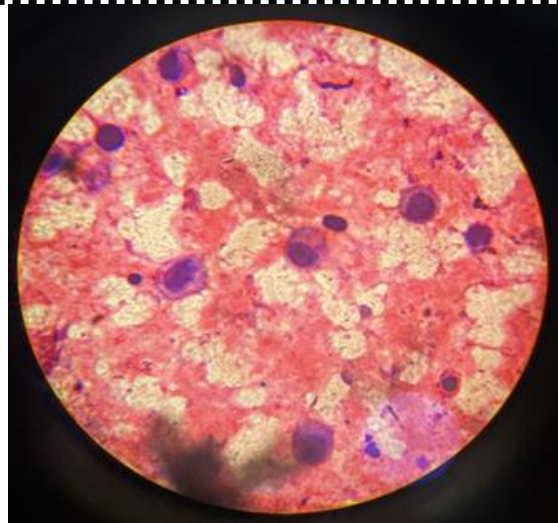


Figure 3: On 100 x examination

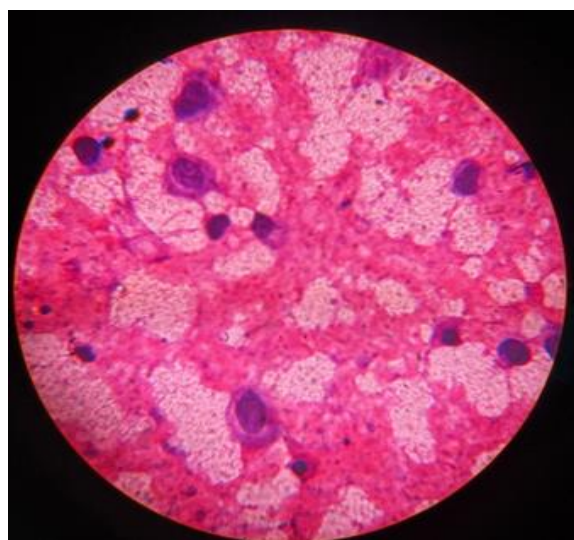


Figure 4: On 100 x examination

Figure 3 and 4 they show round to oval cells having moderately pleomorphic nuclei, having coarse granular chromatin, occasional prominent nucleoli and scant to moderate amount of eosinophilic cytoplasm.

References

1. Zareifar S, Bordbar MR, Karim M, Geramizadeh B, Rasekhi AR. T-cell lymphoblastic lymphoma of the sternum. *J Clin Oncol*. 2009;28(4):e51-3.
2. International Journal of Advances in Medicine| May- June 2018| Vol 5| Issue 3 page: 757-759.
3. Thoracic Pathology: A volume in the High Yield Pathology Series 1st Edition.

4. Sekiguchi N, Nishimoto J, Tanimoto K, Kusumoto S, Onishi Y, Watanabe T. Primary mediastinal large Bcell lymphoma: a single-institution clinical study in Japan. *Int J Hematol.* 2004;79:465-71
3. Faries PL, D'Ayala M, Santos GH. Primary immunoblastic T-cell lymphoma of the sternum. *J Thorac Cardiovasc Surg.* 1997; 114 (4):684-5.
5. Faries PL, D' Ayala M, Santos GH. Primary immuno blastic T cell lymphoma of the sternum. *J Thorac Cardio vase Surg.* 1997; 114 (4): 668-5.
6. Morton LM, Wang SS, Devesa SS. Lymphoma incidence patterns by WHO subtype in the United States, 1992-2001. *Blood.* 2006;107:265.
7. Al-Hamad ani M, Haberman TM, Cerhan JR. Non Hodgkin lymphoma subtype distribution, geode mographic patterns, and survival in the US: A longitudinal analysis of the National Cancer Data Base from 1998 to 2011. *Am J Hematol.* 2015;90:790.
8. Adams SV, Newcomb PA, Shustov AR. Racial patterns of peripheral t-cell lymphoma incidence and survival in the United States. *J Clin Oncol.* 2016;34:963.
9. Herling M, Rassidakis GZ, Jones D. Absence of Epstein- Barr virus in anaplastic large cell lymphoma: a study of 64 cases classified according to World Health Organization criteria. *Hum Pathol.* 2004;35:455.