

Alk- Negative, Anaplastic Large Cell Lymphoma- A Case Report



Medical Science

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Dr.Sujata.S.Giriyani

Department of pathology, Karnataka institute of medical sciences, Hubli, Karnataka.

Dr. Kiran.S.M.

Department of pathology, Karnataka institute of medical sciences, Hubli, Karnataka.

ABSTRACT

Anaplastic large cell lymphoma (ALCL), Anaplastic lymphoma kinase (ALK)-negative (ALCL-ALK-) is a provisional entity in the WHO 2008 Classification that represents 2–3% of NHL and 12% of T-cell NHL.

Patients with ALCL-ALK- are usually adults with a median age of 54–61 years, and a male-to-female ratio of 0.9.

ALCL-ALK- presents with lymph node involvement in 50% of cases; extranodal spread (20%) is less common.

Staging work-up for ALCL-ALK- is similar to that routinely used for nodal NHL. Overall prognosis is poor, with a 5-year of 30–49%, which is significantly worse when compared to reported in patients with ALCL-ALK+ (5-year: 70–86%)5.

INTRODUCTION : Anaplastic large cell lymphoma(ALCL) is rare type of Non-Hodgkin's lymphoma characterized by peripheral, mediastinal or abdominal lymph node involvement1.

This case study is to focus on clinical presentation, cytology , histopathology and immunohistochemistry of ALCL.

CASE REPORT:

A 63 yrs old male patient presented with abrupt onset of left axillary swelling of 15 days duration. On examination Left central group of axillary lymph node enlarged which was solitary firm (m) 4x3 cms. Small left cervical level V node enlarged, (m) 2x1 cms.

No h/o fever, cough, loss of appetite or weight loss.

Hemogram findings , Biochemistry findings, Bone marrow studies were within normal limits.

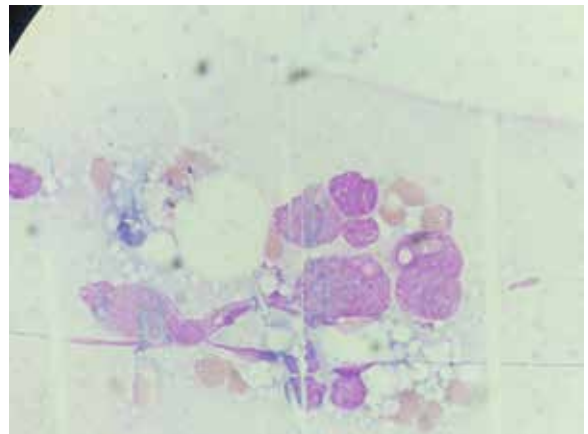
On PET CT : Bulky metabolically active left lower neck, left axillary, left deep pectoral lymphadenopathy with small metabolically active right subcarinal portocaval and right external iliac lymphnodes. Metabolically active intrasplenic focal lesion.

FNAC of left axillary lymphnode was done.

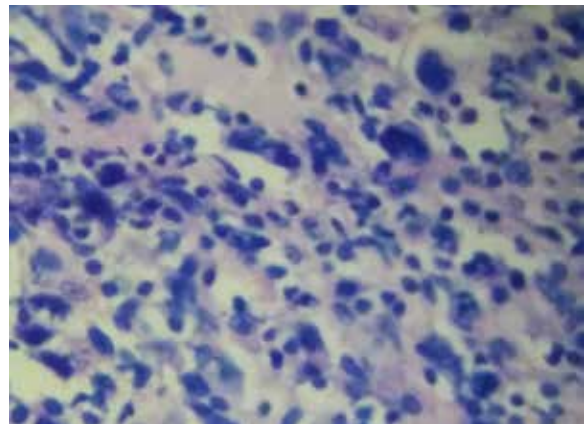
Fine needle aspiration: Smears were moderately cellular showing polymorphic population of lymphoid cells along with dispersed large atypical cells having large irregular nucleus, coarse chromatin, 1 to 2 prominent nucleoli and moderate amount of cytoplasm. Some showed vacuolations. Background showed lipid material with few fragments of adipose tissue.

Differential Diagnosis of

(1)Anaplastic large cell Lymphoma./ (2) Poorly differentiated carcinoma was done and advised biopsy.



Dispersed large atypical cells having large irregular nucleus, coarse chromatin, 1 to 2 prominent nucleoli



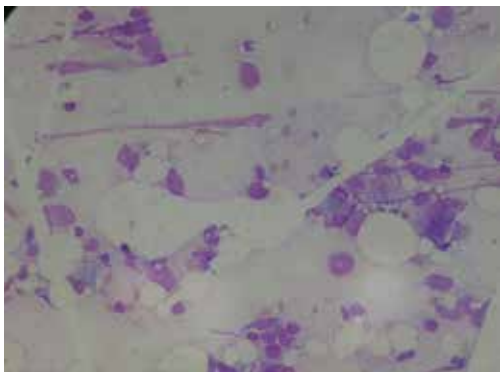
Histopathology: suggestive of Anaplastic Large cell lymphoma.

IMMUNOHISTOCHEMISTRY: The neoplastic large cells were positive for CD 30, EMA(focal), CD45 and were **negative for ALK**, CD68, CD15, PLAP, CD117, EBER, CD56, CD138, CD10 and CD22.

Background T lymphocytes expressed CD3, CD8>CD4, CD5 and CD43.

Ki-67 proliferation index is 80%.

Impression: Consistent with ALK negative Anaplastic Large Cell Lymphoma.



DISCUSSION:

ALCL is a type of non-Hodgkin lymphoma that features in the World Health Organisation (WHO) classification of lymphomas. Defining features consist of a proliferation of predominantly large lymphoid cells with strong expression of the cytokine receptor CD30 and a characteristic growth pattern².

ALCL can be classified in three distinct forms:

1. Systemic ALCL, ALK positive, which affects children or young people.
2. Systemic ALCL, ALK negative, which affects older people.
3. Primary cutaneous ALCL².

ALCL accounts for approximately 3% of adult non-Hodgkin lymphomas & 10% to 20% of childhood lymphomas³. The ALK positive subtype usually affects children & young adults⁽¹⁾. The ALK negative subtype is more commonly found in older patients over the age of 40⁽¹⁾. With treatment ALK-positive patients (5-year survival of 70-80%) have better prognosis than ALK-negative patients (5-year survival of 33-49%)².

ALCL-ALK negative is generally responsive to doxorubicin-containing chemotherapy, but relapses are frequent. CHOP is the most commonly used regimen to treat systemic ALCL with complete remission rates of 56%, and a 10-year DFS of 28%. Encouraging results have been reported with more intensive chemotherapy regimens.

CONCLUSION:

Anaplastic large cell lymphoma is a rare type of NHL, which is characterised by peripheral, mediastinal or abdominal lymph-node involvement and is confirmed by FNAC, histopathology or and immunohistochemistry.

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