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A CASE REPORT OF LYMPHOEPITHELIOID LYMPHOMA-LENNERTS LYMPHOMA

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Abstract :

Lymphoepithelioid lymphoma (Lennerts lymphoma) is a variant of peripheral T cell

lymphoma , not otherwise specified with peculiar histological picture characterised by massive proliferation of epithelioid cell clusters intermingled with many, mostly small-sized lymphoid cells.

Immunohistochemistry shows reactivity with T cell markers and proliferation-associated antigen Ki67.

Keyword : Lennerts lymphoma, peripheral T-cell lymphoma, epithelioid histiocytes, CD3, Ki67.

INTRODUCTION:

Lymphoepithelioid lymphoma (Lennerts lymphoma) is a variant of peripheral T-cell lymphoma,not otherwise specified, with unique histologic morphology characterised by two predominant cell populations - epithelioid cells and T-lymphocytes.[1,2]Ly mphoepithelioid lymphoma was first described by Lennert [3] in 1952 and later by Lennert and Mestdagh [4] as a special variant of Hodgkin's disease. Later in WHO it was included in the peripheral T cell lymphoma- not otherwise specified (ICD-0 code 9702/3) [1], which accounts for 30% of all the peripheral T cell lymphomas.[1] Older individuals are most commonly affected in their 6th or 7th decade.[1,2] Males are more commonly affected with male to female ratio of 2:1. [1,2] Patients usually present with peripheral lymphadenopathy and hepatosplenomegaly.

CASE REPORT:

60 years old male presented with bilateral cervical lymphadenopathy, persistent cough and fever. Physical examination revealed mild hepatosplenomegaly. Complete hemogram showed Hb:12.8 g/dl, WBC:7400cells/cumm, platelets:2,03,000cells/cumm. Peripheral smear and bone marrow aspiration study were normal. All relevant investigations were done to rule out tuberculous lymphadenitis. With clinical impression of lymphoma, biopsy from the lymph node was sent for histopathological study.

Microscopic examination revealed complete effacement of lymph node architecture by diffuse proliferation of small lymphoid cells and epithelioid histiocytes (Fig 1). Histiocytes forming numerous microgranulomas are noted (Fig 2). Reed-Sternberg cells are not seen.

Histopathological study was reported as Non Hodgkin's lymphoma morphologically Lennerts lymphoma.

Immunohistochemistal study was carried outwhich showed strong uniform reactity

with pan T-cell marker CD3 (Fig 3) and CD45 (Fig 4). Proliferation antigen ki67 showed high proliferation rate with strong nuclear positivity (Fig 5). Immunostaining were negative for CD20, CD79a, CD15 and CD30 (Fig 6). Hence, the results were consistent with lymphoepithelioid lymphoma.

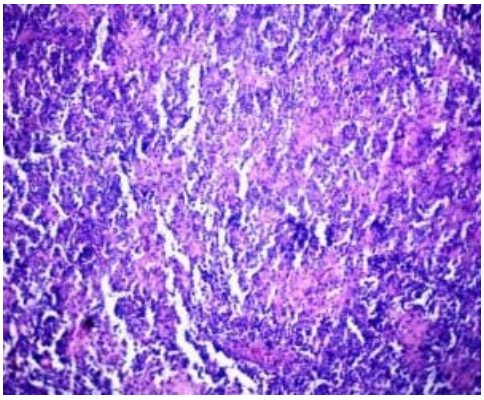


Fig 1. Diffuse effacement of lymphnode architecture by epithelioid histiocytes and small lymphoid cells- H&E (10 x)

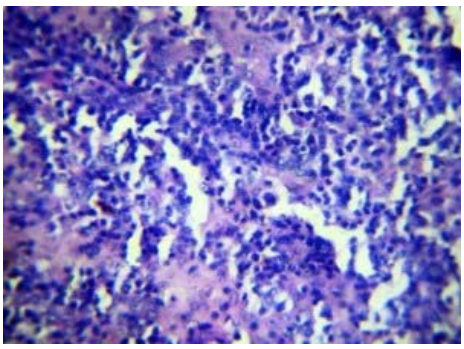


Fig2. Epithelioid clusters forming microgranulomas (40 x)

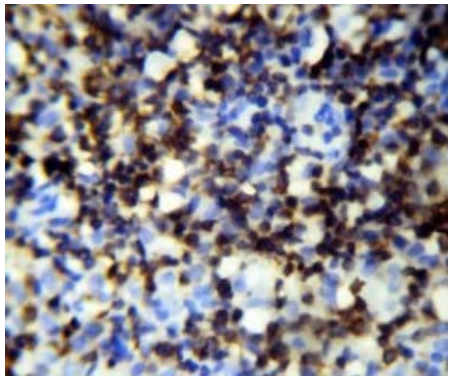
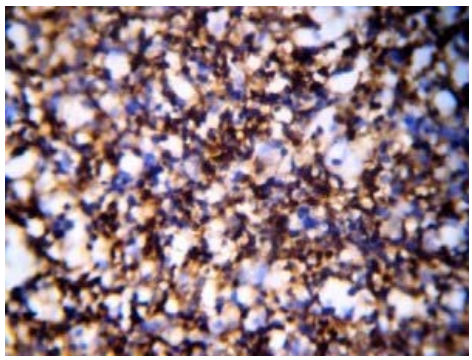
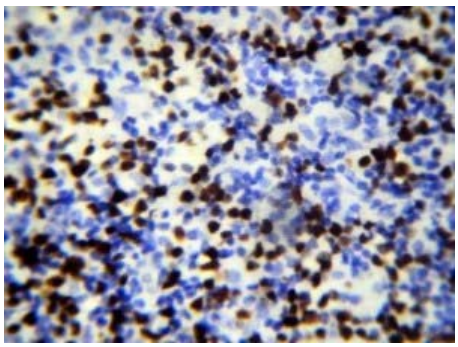
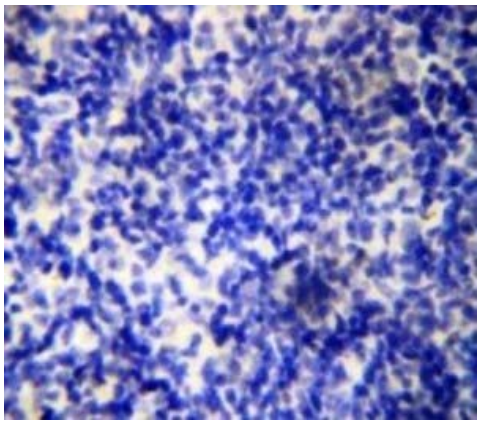
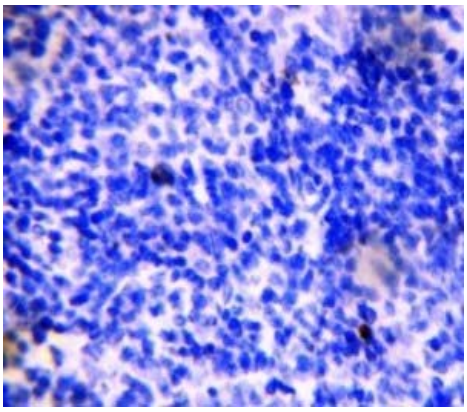


Fig3. Immunoreactivity with CD3 showing perinuclear positivity (40 x) Fig4. Immunoreactivity with CD45 RO (40 x) Fig5. High proliferation rate shown by immunostaining with Ki67 (40 x) Immunostaining with B-cell markers CD20 and CD79a shows negativity (40 x)









DISCUSSION:

Lymphoepithelioid lymphoma (Lennerts lymphoma) is a particular variant of peripheral T-cell lymphoma, not otherwise specified with unique histologic morphology characterised by two predominant cell populations -epithelioid cells and T-lymphocytes, mainly T helper /inducer phenotype.[5,6] In the Non-Hodgkin Lymphoma Classification Project,[7] peripheral T-cell lymphoma, not otherwise specified represented 6.0% of all non-Hodgkin lymphomas and approximately half of all T/NK-cell neoplasms. The incidence of Lennerts lymphoma increases with age, and are more common in men than in women.They usually present with peripheral lymphadenopathy. Generalised disease involves

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Histologically, lymph node in Lennerts lymphoma shows diffuse or rarely interfollicular infiltrates with effacement of lymph node architecture.[1,2] Confluent clusters of epithelioid histiocytes are seen admixed with predominantly small lymphoid cells with scant cytoplasm, round or irregular nuclei and dense chromatin showing mild to moderate atypia.[1,2] Some medium sized and large atypical lymphoid cells are also seen intermingled, usually in proximity to epithelioid cells.[1,3] Reed-Sternberg like cells, if present are very sparse. High endothelial venules are not prominent.[1,2] There can be admixed inflammatory cells composed of eosinophils and few plasma cells.

Immunohistochemistry shows strong and diffuse positivity of small lymphoid cells with pan T-cell markers and T-cell associated antigens CD3, CD2 and CD45 RO. Most cases express CD4 (helper/inducer T -cell), with a smaller subset of neoplasms positive for CD8 (suppressor/cytotoxic T-cell). B-cell markers CD20, CD79a are usually negative.[9] CD15 can be expressed exceptionally in large lymphoid cells and CD30 shows variable expression. Proliferation is usually high and Ki67 rates exceeding 75% are associated with worse prognosis.[1]In keeping with the expression pattern of normal T cells, 95% of which are positivfor the T-cell receptor (TCR) aß, most cases of peripheral T-cell lymphoma show monoclonal TCR 8 gene rearrangements.
Lymphoepithelioid lymphoma must be distinguished from other conditions associated with prominent epithelioid infiltrate[10] which includes

1. Classical Hodgkin's lymphoma

2. Reactive granulomatous lymphadenitisEpithelioid cell granulomas, multinucleated giant cells in a background of polymorphouspopulation of lymphocytes without cytological atypia differentiates granulomatouslymphadenitis from lymphoepithelioid lymphoma.

3. Angioimmunoblastic T-cell lymphoma with a high content of epithelioid cells.

Clear cells or high endothelial venules are less frequent in lymphoepithelioid lymphoma than in Peripheral T- cell lymphomas of angioimmunoblastic type.Lennerts lymphoma is highly aggressive lymphomas with poor response to therapy.

Frequent relapses are common and have low overall 5 year survival rate ranging from 25% to 45%.[5] Involvement of bone marrow is a negative prognostic factor [1] and survival rate for stage 4 disease is < 10%.[2] These lymphomas can transform into a large cell lymphoma of T- cell lineage with loss of epithelioid histiocyte reaction.[2]

CONCLUSION:

In the presence of epithelioid granulomaswith the background of monomorphic lymphoid population, Lennerts lymphoma should be considered. The diagnosis of Lennerts lymphoma could be confirmed by immunophenotyping and TCR genetic rearrangement studies.

BIBLIOGRAPHY:

1.Ralfkiaer E, Muller-Hermelink HK, Jaffe ES. Peripheral T-cell lymphoma, unspecified. In: Jaffe ES, Harris NL, Stein H, et al. eds. World Health Organization Classification of Tumours. Pathology and genetics of tumours of haematopoietic and lymphoid tissues. Lyon, France: IARC Press, 2001; 227 –229.

2 Christopher D.M.Fletcher. Diagnostic Histopathology of Tumors. 3rd edition, Elsevier, 2007; 1214-1218.

3 Lennert K: Zur Histologischen Diagnose der Lymphogranulomatose. Frankfurt, FRG, Habil-Schrift, 1952.

4 Lennert K, Mestdeagh i: Hodgkin's disease with constantly High content ofepithelioid cells. Virchows Arch [B] 344:1, 1968.

5 H.L.Ioachim, L.J.Medeiros. Lymph Node Pathology. 4th edition, Lippincott Williams & Wilkins, 2009; 482-489.

6. Y. Daneshbod. Cytologic findings of peripheral T-cell lymphoma (PTCL) with high epitheloid cell content (Lennert's lymphoma) in imprint smear. A case report.*cytoJournal2006, 3:1-4.*

7 The Non-Hodgkin's Lymphoma Classification Project. A clinical evaluation of the

8 International Lymphoma Study Group classification of non-Hodgkin's lymphoma. Blood 1997;89: 3909–3918.

9 A.C. Feller, G.H. Griesser et al ; Lymphoepithelioid Lymphoma (Lennert's Lymphoma) Is a Monoclonal Proliferation of Helper/Inducer T Cells. *Blood, Vol 68, No 3 (September), 1986: pp 663-667.*

10 Picker LJ, Weiss LM, Medeiros LJ, et al. Immunophenotypic criteria for the diagnosis of non-Hodgkin's lymphoma. Am J Pathol 1987;128:181–201.

11 E.S. Jaffe et al. Hematopathology, Elsevier, 2011, 541-553.