Case Report

Lennert's lymphoma

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ABSTRACT

Lymphoepithelioid cell lymphoma (Lennert's Lymphoma) is a rare morphological variant of peripheral T-cell lymphoma characterized by the presence of numerous clusters of epithelioid histiocytes without formation of discrete granulomas and the intervening atypical lymphocytes. Lennert's lymphoma is often misinterpreted as granulomatous lymphadenitis or Hodgkin's disease. This report describes fine needle aspiration cytology and histological findings in a case of Lennert's lymphoma.

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In 1952, Lennert[1] described an unusual form of malignant lymphoma characterized by presence of small evenly distributed clusters of epithelioid cells in Hodgkin's disease such as abnormal tissue but virtually lacking typical Reed-Sternberg giant cell. Later this lymphoma was termed lymphoepithelioid cell lymphoma (Lennert's Lymphoma) and grouped in the new World Health Organization classification as a lymphoepithelioid cell variant in the peripheral T-cell lymphoma (PTCL), unspecified entity.[1] Histological and cytological features of Lennert's lymphoma mimics a wide range of lesions with high content of epithelioid cells. We report a case of this rare disease.

Case Report. A 50-year-old Palestinian lady presented in January 2004 with 3 months history of itching, intermittent fever and weight loss. Clinically, the patient had a right supraclavicular lymphadenopathy. Initial investigations showed eosinophilia and erythrocyte sedimentation rate of 25 mm/hour. Biochemistry tests were normal apart from lactate dehydrogenase of 1130 IU/L. Fine needle aspiration of right supraclavicular lymph node showed cellular smears composed of lymphoid cells, histiocytes, plasma cells and eosinophils. The lymphoid cells were small to medium size, some with irregular nuclei and some others with large nuclei showing nucleoli. Few were binucleated. The histiocytes were seen as single cells or in clusters. Some of these clusters showed epithelioid cell morphology (**Figure 1**). No acid-fast bacillus seen in ZN stain. Lymph node biopsy was suggested to rule out lymphoma.

On further examination, there were multiple lymph nodes in the occipital, right upper cervical, right supraclavicular and bilateral axillary regions. Spleen was palpable 3 cm below the costal margin. The computerized tomography (CT) scan of the chest and abdomen showed multiple lymphadenopathy of varying sizes in mediastinal, para aortic, iliac and inguinal regions. Left iliac group of lymph nodes showed evidence of necrosis.

Excision biopsy of the right supraclavicular lymph node was carried out. Two lymph nodes measuring 2 x 1.5 x 0.8 cm and 1.5 x 1 x 0.8 cm were received in the Histopathology Section. Microscopy showed lymph nodes with loss of normal architecture and replacement by small aggregates of epithelioid cells and mixed population of lymphoid cells, eosinophils, plasma cells and

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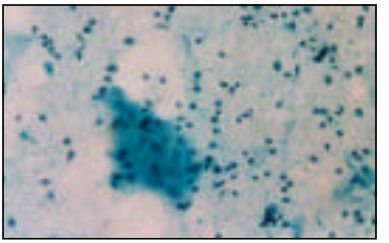

Figure 1 - Fine needle aspiration cytology smear showing cluster of epithelioid cells, small to large lymphoid cells, eosinophils and plasma cells (papanicolaou stain x 400).

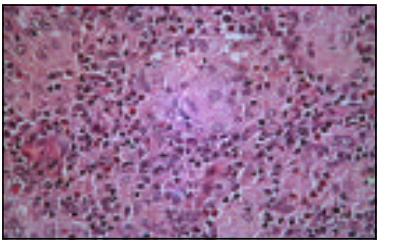

Figure 2 - Effacement of normal lymph node architecture by polymorphic infiltrate consisting of epithelioid histiocytes, small lymphocytes, scattered large lymphoid cells, and eosinophils (hematoxylin and eosin x 400).

histiocytes. The lymphoid cells were of varying sizes (small, medium and large). Some showed irregular nuclei and some were in mitosis (**Figure 2**). There was no necrosis. Infiltration in the capsule and perinodal fat was seen. No organisms were demonstrated in ZN, PAS or Giemsa stains.

On immunohistochemistry, the lymphoid cells were diffusely positive for T-Cell marker (CD45RO). The histocytes were CD68 positive. There were scattered CD5 positive cells. In February 2004, the patient developed right hemiplegia and aphasia due to left middle cerebral artery (MCA) infarction, which was confirmed by brain CT scan. Patient received five cycles of cyclophosphamide, doxorubicin, vincristine, prednisolone (CHOP). Except for receiving granulocyte colony stimulating factor (GCSF) for neutropenia she tolerated the chemotherapy. She had marked clinical and radiological improvement.

Discussion. Lennert's lymphoma is an uncommon neoplasm accounting for approximately 3.1% of the lymphomas.[2] Lennert's lymphoma is a peripheral T-cell lymphoma of CD4 type. The median age of presentation is 60 (range 21-87) years with a male to female ratio of 1.3:1. The patient usually presents with lymphadenopathy and there is a tendency of early generalization. The majority present in advanced stage with constitutional symptoms.[3] Fine needle aspiration cytology (FNAC) of lymph nodes is a first line investigation in patients with lymphadenopathy. Immunophenotyping by flow cytometry or by immunohistochemistry has greatly improved the accuracy of cytological diagnosis of lymphomas. Fine needle aspiration cytology is an accepted means of diagnosing and typing common forms of lymphomas, particularly small lymphocytic lymphoma and large B cell lymphoma. The diagnosis of peripheral T-cell lymphoma in FNAC smears is often difficult due to the heterogeneous population of cells. The cytopathological features of Lennert's lymphoma are not well documented in the literature except for few case reports.[4-6] Lennert's lymphoma should be distinguished from lesions rich in epithelioid cells such as angioimmunoblastic T-cell lymphoma, Hodgkin's disease, lymphoplasmacytic lymphoma with a high content of epithelioid cells and from inflammatory epithelioid cell reactions. Angioimmunoblastic T-cell lymphoma type of peripheral T-cell lymphoma differs from classic Lennert's lymphoma by the presence of arborizing high endothelial venules and dendritic cell-lymphocyte complexes.[7] The polymorphic nature of the infiltrate and occasional presence of Reed-Sternber (RS) such as cells often elicit a mistaken diagnosis of Hodgkin's disease. However, Hodgkin's disease with high epithelioid cell content mainly in the mixed type shows many Hodgkin's and RS cells. Fibrosis, necrosis and eosinophilia are frequently seen in Hodgkin's disease but rare in Lennert's lymphoma.[8] Lymphoplasmacytic lymphoma can be easily distinguished from Lennert's lymphoma by the presence of plasmacytoid and plasma cells with precursor cells and by the demonstration of monotypic immunoglobulin in the cytoplasm of plasma cell series. Lennert's lymphoma is often misinterpreted on cytology as granulomatous lymphadenitis.[5,9] Inflammatory epithelioid cell reaction commonly seen in tuberculosis, toxoplasmosis and infectious mononucleosis can be differentiated by the demonstration of responsible organism/serology and by their classical histological findings.

The clinical course of Lennert's lymphoma is quite unpredictable with few spontaneous but mostly unstable remissions. The clinical outcome is generally poor. Patsouris et al[3] observed transformation of Lennert's lymphoma into high

grade malignant lymphoma in 8% of their cases. A median survival is approximately 16 months. Although histopathological and histochemical studies are required for a definite diagnosis, the awareness of this rare morphological variant helps in early diagnosis by FNAC.

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