

Lymphoid Atypia in Dusty Lungs: A Case Report of Lennert's Lymphoma with Pneumoconiosis

Dr Aryakrishna SL, MD, DM¹, Dr Sugeeth M T, MD, DNB, DM, DNB^{1*}, Dr Pallavi Nair, MD, DM¹

¹Assistant Professor, Department of Medical Oncology, Department of Medical Oncology, Regional Cancer Centre Trivandrum

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*Corresponding author: Dr Sugeeth M T

Assistant Professor, Department of Medical Oncology, Department of Medical Oncology, Regional Cancer Centre Trivandrum

Abstract

Case Report

Lennert's lymphoma is a type of rare T cell lymphoma. It poses a great diagnostic dilemma. When compared to other T cell lymphomas it has a better prognosis. Here we are presenting a case of forty-two-year-old gentleman who is an arc welder presenting with loss of weight, loss of appetite, lymphadenopathy and on evaluation was found to have pneumoconiosis and biopsy from the lymph node was suggestive of Lennert's lymphoma stage, IIIB. He was treated with multiagent chemotherapy with CHOP and is currently on follow up

Keywords: Lennert's lymphoma, pneumoconiosis, CHOP Chemotherapy.

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INTRODUCTION

Lennert Lymphoma (LL), or the lymphoepithelioid variant of peripheral T-cell lymphoma, is a rare entity first described by Karl Lennert in 1952 [1]. It was initially considered a variant of Hodgkin's Lymphoma (HL), but categorized as a non-Hodgkin lymphoma in 1975 when Lennert et al referred it as "lymphoepithelioid cellular lymphoma" [2]. Oftentimes, this specific variant tends to remain localized to lymph nodes with only rare extra-nodal extension. It is thought to have a better prognosis in comparison with other peripheral T-cell lymphoma variants [2]. Here we report a case of Lennert's lymphoma with associated pneumoconiosis.

CASE REPORT

We report a case of a forty-year-old gentleman, an arc welder with Diabetes mellitus, who presented with loss of appetite and loss of weight of one year duration. On evaluation, he was found to have bilateral lung nodules and a few mediastinal nodes for which he was started on empirical anti-tuberculosis (ATT) treatment. He developed axillary nodes while on ATT and was referred to our institution. On examination, he had multiple cervical and axillary lymph nodes.

His blood hemogram, Serum biochemistry, and Electrolytes were within normal limits. His LDH level was 291 units /litre. His viral markers were negative. Biopsy from the left axillary node showed infiltration of atypical small to medium-sized lymphoid cells with irregular nuclear membranes, prominent nucleoli, and scanty cytoplasm with effaced architecture. Mixed with are seen many clusters of epithelioid histiocytes. On immunohistochemistry (IHC), these atypical lymphoid cells were positive for CD3, CD4 and negative for CD8, CD20, and CD33. CD7 was down-regulated, and the picture was suggestive of Lennert's lymphoma. Bone marrow evaluation showed marrow infiltration by lymphomatous cells. Hest Xray showed multiple nodules scattered throughout lung fields suggestive of pneumoconiosis (Fig 1). Positron Emission Tomography (PET-CT) scan showed multiple supra and infradiaphragmatic lymphadenopathy. He also had lung nodules suggestive of occupational lung disease. He was diagnosed with Lennert's Lymphoma, Stage IV, with an International Prognostic Index (IPI) score of 3. Cardiology evaluation was within normal limits. Pulmonology opinion was taken and was advised to change occupation as there was a chance of worsening of interstitial lung disease, and clearance for chemotherapy was obtained.



Fig. no 1: X ray PA view of chest showing multiple nodules scattered through out lung fields suggestive of pneumoconiosis

He was treated with multiagent combination chemotherapy with cyclophosphamide, Adriamycin, Vincristine, and Prednisolone (CHOP) for six cycles. PET CT scan at the end of six cycles was in complete metabolic remission with stable findings in the lung suggestive of pneumoconiosis. The patient has been on follow-up since December 2022 for the past 30 months.

DISCUSSION

Lennert's lymphoma (LL) is classified under "Peripheral T-cell lymphoma - Not otherwise specified" by 4th edition of WHO classification system (1). The median age of presentation was 60 years in retrospective analysis by Patsouris *et al.*, [2]. Our patient was of age 50 years. Splenomegaly and lymphadenopathy are the usual findings on presentation, and our case also presented with lymphadenopathy. Various risk factors are involved in the development of lymphoma, one of which is environmental exposure to various toxins [3]. Occupational exposure may have contributed to the development of lymphoma in our patient, even though conclusive evidence is absent.

Histologically, LL shows diffuse infiltrates consisting predominantly of small cells with marked nuclear irregularities, along with clusters of epithelioid

histiocytes that are scattered within these infiltrates. Clear cells or high endothelial venules are less frequent than in peripheral T-cell lymphomas of angioimmunoblastic or T-zone type. Few Reed-Sternberg (RS)-like cells, eosinophils, and plasma cells may also be seen [5]. However, in our case RS RS-like cells were not identified. Atypical lymphoid cells were positive for CD3, CD4, and negative for CD8, CD20, CD33, and CD7 was down-regulated.

The response to chemotherapy is often poor, with a mean overall survival being about 42 months. Standard treatment approaches for PTCL NOS include combination chemotherapy with the CHOP regimen, Fludarabine and cyclophosphamide, and ifosfamide, carboplatin, and etoposide (ICE). If CD52 is positive, alemtuzumab may be used with standard chemotherapy [7]. For those who relapse, reduced intensity conditioning and allogeneic stem cell transplantation after salvage therapy, integrating alemtuzumab is considered. However, the prognosis remains poor, with the majority of the series reporting a 5-year overall survival of approximately 30-35% using standard chemotherapy. Our patient was also treated with CHOP chemotherapy and had a complete metabolic response post-chemotherapy and has been followed up since 2022.

Table 1: Reported cases of Lennert's lymphoma

| | Age | Sex | Presenting Symptom | Stage | Treatment | Follow Up | Outcome |
|-----------------------------|-----|-----|---|-------|-----------|-----------|---------|
| Parimal <i>et al.</i> , [8] | 67 | M | Lymphadenopathy | IIIS | CEPP | -- | -- |
| | 45 | F | Lymphadenopathy | IIIA | CHOP | -- | -- |
| | 27 | F | Lymphadenopathy | IIIBS | CHOP | -- | --- |
| | 68 | M | Lymphadenopathy | NA | NA | -- | -- |
| | 32 | F | Lymphadenopathy | NA | NA | -- | -- |
| Yin <i>et al.</i> , [9] | 65 | M | painless mass in the subcutaneous soft tissue of the left forehead. | | CHOP | 20 months | NED |
| Ding <i>et al.</i> , [10] | 58 | f | Left inguinal lymphadenopathy | IV | CHOEP | 60 months | NED |

| | Age | Sex | Presenting Symptom | Stage | Treatment | Follow Up | Outcome |
|-----------------------------------|-----|-----|---|-------|-----------------|-----------|----------|
| Kapoor <i>et al.</i> , [11] | 50 | M | Generalized lymphadenopathy B symptoms | IIIB | CHOP | --- | --- |
| Roundtree <i>et al.</i> , [12] | 85 | F | Lymphadenopathy Skin lesions | IV | ---- | ---- | --- |
| MacGillivray <i>et al.</i> , [13] | 73 | M | Tonsillar mass | | Supportive care | 2months | Deceased |
| Mishra <i>et al.</i> , [14] | 23 | M | Thyroid mass | III | On evaluation | 2 months | -- |

NED: No evidence of disease, NA: Not applicable, CHOP: Cyclophosphamide, Adriamycin, Vincristine, Prednisolone, CHOEP: Cyclophosphamide, Adriamycin, Vincristine, Etoposide, Prednisolone

CONCLUSION

Lennert's lymphoma is a rare T cell lymphoma. It may pose a diagnostic dilemma as it can mimic Hodgkin lymphoma at times. Due to its rarity, lack of clinical trials optimal treatment of this group of lymphoma is still unclear. However, when compared to other T cell lymphomas it has a better outcome.

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