



A RARE CASE OF PRIMARY PLEURAL LOW GRADE MARGINAL ZONE NON-HODGKIN'S LYMPHOMA: A CASE REPORT

Pulmonary Medicine

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ABSTRACT

A primary pleural based marginal zone low grade NHL is very rare. Primary pleural Lymphomas account for less than 0.5% off all Non-Hodgkin Lymphomas and DLBCL is the most common subtype of primary pleural lymphomas. Here, we report a rare case of marginal zone lymphoma of low grade with Mib index of 15-20% presenting as a pleural based mass in a middle age individual. A 52 years old male presented with right sided pleural effusion with pleural thickening of soft tissue density. Thoracoscopy showed pleural based mass lesion and pleural biopsy revealed low grade marginal zone NHL which is very rare with very few cases documented till date. Diagnosis was based on Bcl-2 and CD20 marker positivity on IHC. Treatment of such Low-grade Marginal zone lymphoma is with Bendamustine and Rituximab.

KEYWORDS

Primary pleural Lymphoma, Marginal zone Lymphoma, Thoracoscopy guided forceps and cryo-biopsy.

Case Report

A 52 years old male patient presented with complaints of cough, fever and breathlessness for almost 1-2 months. He was evaluated with Chest X-Ray which showed Right Pleural Effusion and pleural fluid analysis suggestive of exudate with high ADA [54.76-ADA, Protein-8.7, Sugar-103] and cytology was negative for malignant cells. With India being endemic for Tuberculosis and based on pleural fluid analysis, patient was started on ATT. Despite ATT, patient had recurrent pleural effusions and hence CT Chest was done which showed Right Moderate to Massive Pleural Effusion with thickened and clearly demarcated pleura with soft tissue density (Figure-1: A & B). Autoimmune work-up was negative.

Despite pleural fluid analysis suggestive of exudate with high ADA most probably Koch's, in view of CT Chest findings, Right side Medical Thoracoscopy was done which showed pleural based mass lesion (Figure-2: A). Forceps and Cryo-Biopsy was done under Thoracoscopic guidance (Figure-2: B & C).

Histopathological examination was suggestive of Marginal Zone-Low Grade Non-Hodgkin's Lymphoma (Figure-3: A, B & C). Hematoxylin and Eosin (H & E) stain showed monotonous atypical Lymphoid cells infiltrating Pleura. Further Immuno Histo-chemical (IHC) tests revealed diffuse CD-20 and Bcl-2 Positivity suggestive of B-Cell Lymphoma with Mib1 Index of 15-20% while CD-3, CD-5, CD-23, Cyclin-D1, TdT, Sox-11, CD-10 were all negative.

Further PET-CT Whole Body was done which revealed FDG avid Right Pleural Mass with Pleural Thickening (Figure-1: C).

DISCUSSION

Primary pleural lymphoma is very rare with incidence of about 0.3 to 1% of extra nodal lymphomas. It is also very rare in immunocompetent patients without any previous history of Tuberculosis, HIV or empyema(1). It has diverse clinical presentations without specific symptomatology.

Our patient presented with dyspnoea, low grade fever, right sided chest pain. We did Thoracoscopy guided pleural mass biopsy which showed typical histopathological and Immuno-Histochemistry (IHC) findings of low grade pleural marginal zone B-cell lymphoma. Primary pleural NHL is extremely rare. Diffuse Large B Cell Lymphomas are most common Primary Pleural NHL followed by Follicular type(2,3).

Extra Nodal Marginal-zone Lymphoma or EMZL or mucosa-associated lymphoid tissue lymphoma is a relatively uncommon clonal B-cell neoplasm which comprises only 5–10% of all non-Hodgkin lymphoma. They arise in various epithelial tissues, mostly gastrointestinal, salivary glands and cutaneous tissues. Lung

involvement more commonly presents as lung nodules (4,5). Primary Pleural involvement is very rare but has been reported in a few case reports (6,7). Extra Nodal Marginal Zone Lymphomas arise from Mucosa-associated Lymphoid Tissue (MALT) and are most commonly associated with chronic inflammation secondary to infections or autoimmune conditions(8).

Primary pleural lymphomas should be differentiated from other pleural malignancies and plaques. Other differentials include Adenocarcinoma and pleural mesothelioma.

Adenocarcinoma commonly presents with primary lung mass with irregularly nodular pleura with variable sized nodules whereas pleural mesothelioma presents with heterogeneously thickened pleura. CT Thorax is the primary investigation which is used to evaluate pleural effusion and pleural nodularity. In this case, CT Thorax showed right sided moderate pleural effusion with soft tissue density thickening of pleura in costal region.

Here, Medical Thoracoscopy guided Forceps Biopsy as well as cryo-biopsy was used. Pleural Cryo-biopsy gives advantage of Deeper sections as well as preserved Cellular architecture(9).

PET-CT is more accurate in overall staging of lymphoma compared to simple CT scan. Immuno-Histochemistry findings were malignant cells positive for CD20 and Bcl-2 with Mib-1 index around 15-20% suggestive of low-grade marginal zone B-cell lymphoma.

In general, extra-nodal pleural B-cell lymphomas are rare(10,11). Here, Extra-Nodal Primary Pleural Marginal Zone NHL which is low grade is extremely rare. Treatment of such Low-grade Marginal zone Lymphomas is Bendamustine and Rituximab. If patient was having moderate to high grade lymphoma R-CHOP therapy would have been considered.

In general, pleural lymphomas which are low grade have good prognosis with overall survival of 90% (5-year survival) but extrapulmonary spread causes poor prognosis(3).

CONCLUSION

This is a very rare case of primary low grade marginal zone NHL or extra nodal marginal zone lymphoma in a middle-aged immunocompetent individual. Primary Pleural Lymphomas as such are very rare and DLBCL variety is most common followed by Follicular type. Here we emphasize the need for CT imaging and Thoracoscopic pleural biopsy even in patients with high ADA in Tuberculosis endemic countries like India. Thoracoscopy guided Cryo-Biopsy of Pleural mass gives additional advantage of well preserved Cellular architecture.

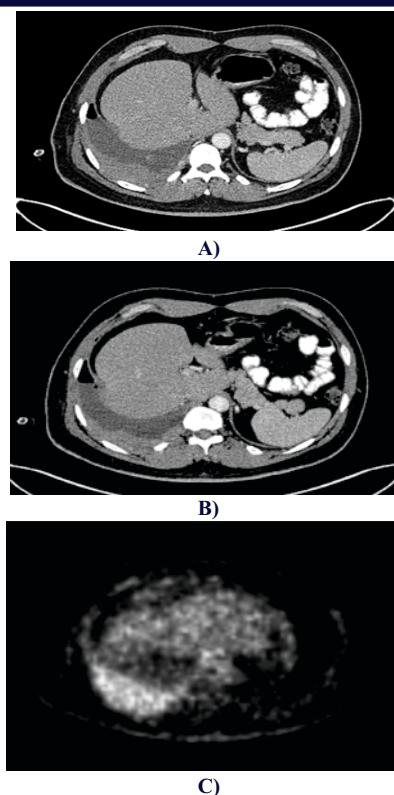


Figure 1: A & B) CT Chest with Contrast showing Right sided Pleural Effusion with Heterogenous Enhancement in Pleura suggesting Pleural mass. C) PET Image showing Enhanced Pleural mass in Right Pleural cavity.

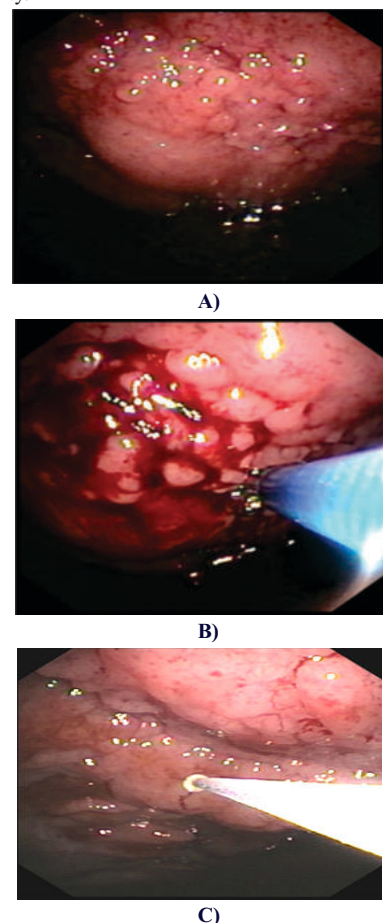


Figure 2: Medical Thoracoscopy view of A) Pleural Mass, B) Forceps Biopsy, C) Cryo-Biopsy

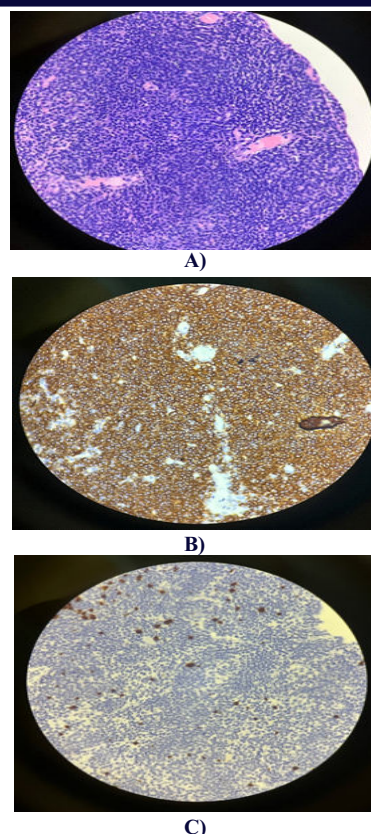


Figure 3: Histopathological and Immuno-Histochemical Images. A) Microscopic Image showing monotonous Low-Grade Atypical Lymphoid cells infiltrating Pleura (H & E Stain). B) Diffuse CD-20 Positivity on IHC. C) Mib1 Index of 15-20% on IHC.

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