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Extra Nodal Marginal Zone Lymphoma of Lung: A Clinical Report

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Abstract

Extra Nodal Marginal Zone lymphoma constitutes 0.4 % of lymphomas and 3.6 % of non-Hodgkin's lymphoma. Extra Nodal Marginal Zone lymphoma may occur in many sites including the GI tract, Orbit, Salivary glands, thyroid, respiratory tract, skin, genitourinary tract, and breast. Lung involvement is usually rare and usually seen in the 6th decade of life. Pulmonary parenchyma does not contain organized lymphoid tissue physiologically, but some conditions (such as follicular bronchiolitis, pulmonary inflammatory processes & acute infections) can lead to lymphocytic hyperplasia. The management of lung extranodal marginal zone lymphomas include surgery, chemotherapy, immunotherapy & Radiation therapy alone or in combination but optimal treatment is not well defined.

Keywords: Extra Nodal Marginal Zone lymphoma, Non-Hodgkin's lymphoma, Lung, surgery, chemotherapy, immunotherapy & Radiation therapy.

Introduction

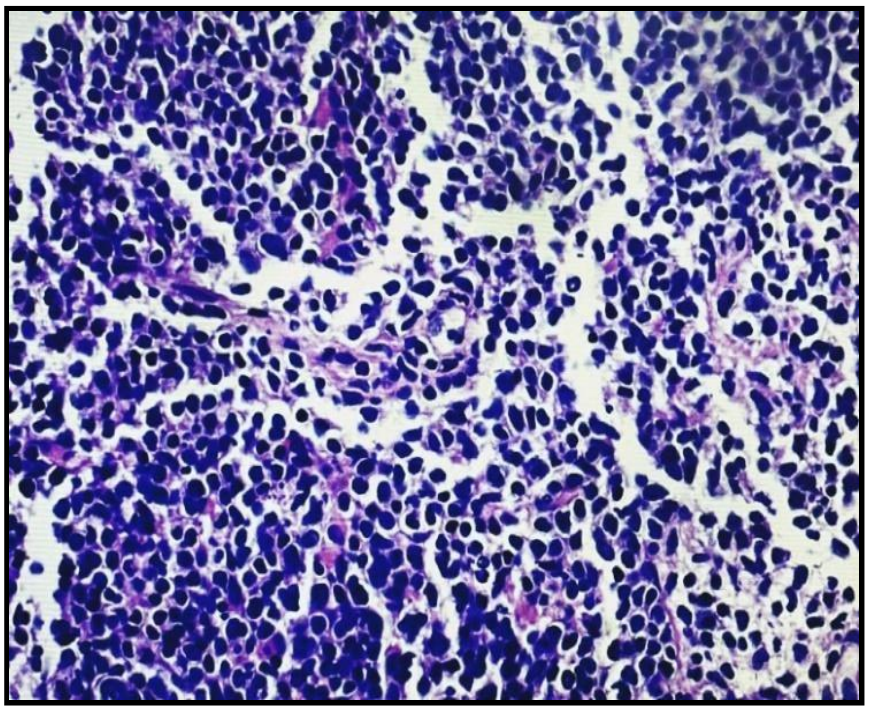
Mucosa-associated lymphoid tissue-derived (MALT) lymphoma is the most frequent subset of primary pulmonary lymphoma. Gastrointestinal tract involvement is the most frequent primary location, with lung location representing 15 % of cases (1). MALT lymphoma is a low-grade B-cell extra nodal lymphoma characterized by a proliferation of clonal marginal zone lymphocytes (MZLs) that invade epithelial structures and form characteristic lymphoepithelial lesions. Much evidence demonstrates that MZLs are associated with chronic antigenic stimulation, either by autoantigen or pathogen, leading to the accumulation of lymphoid tissue in involved organs **[1]** Helicobacter pylori is the best-characterized causative pathogen and is responsible for gastric MALT lymphoma. Other chronic infections seem associated with MALT-derived lymphoma at other sites, even if their roles in pathogenesis are not as firmly established as that of H. pylori. However, an association between a specific pathogen and lung MALT lymphoma has never been shown **[1]**. Pulmonary marginal zone lymphoma carries a favourable outcome with a 5-year overall survival (OS) rate of 90 % **[2]**.10 Presence of mediastinal lymphadenopathy, extrapulmonary MZL, and use of chemotherapy regimens including anthracyclines or cyclophosphamide have been associated with shorter progression-free survival (PFS) and time to progression. Overall, clinical and radiological variables associated with worse outcomes remain poorly understood **[2]**. Patients with pulmonary MALT lymphoma were managed by a multidisciplinary team comprising several specialists, including a pulmonologist, thoracic surgeon, radiologist, and oncologist **[3]**.

Case Report

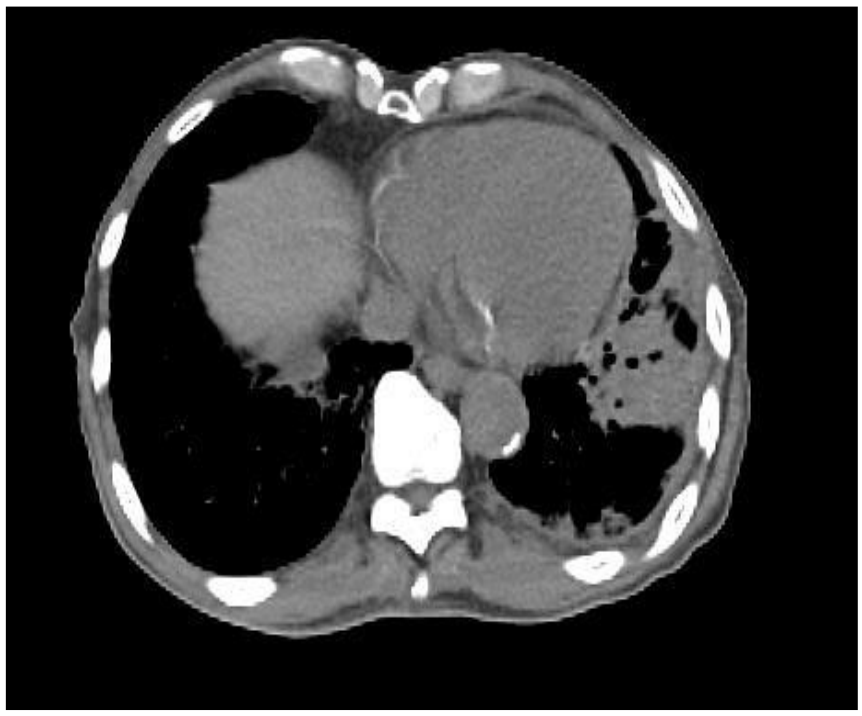
A 67-year gentleman presented in November 2021 with complaints of headache, cough with expectoration & fever for 1 year. The patient also gave a history of dyspnoea on exertion. A patient doesn't give a history of haemoptysis, blood-tinged sputum, weight loss, or loss of appetite. He was diagnosed and treated with Tuberculosis for 10 months. No history of any other known medical co-morbidities. The patient has no significant family or personal history.

Diagnostic Assessment

An MDCT Thorax done in October 2021 showed an Ill-defined suspicious soft tissue density lesion approximately measuring 5.5 X 4.5 cm seen in left lower lobe consolidation with specks of calcification. PET-CT scan done in November 2021 Showed a Low-grade FDG uptake noted in the ill-defined consolidation-like area involving the lower lobe of the left lung, measuring 5.4 X 7.9 X 3.7 cm. He was evaluated and diagnosed to have Low-Grade Non-Hodgkin's lymphoma (NHL) on Left Lung Biopsy. Immunohistochemistry (IHC) was done, and it was positive for CD20, BCL2, CD43 & negative for CD5, CD10, and CD23. CD3 highlights the background T cells. Ki67: 10 % which is confirmed as Extranodal marginal zone lymphoma.



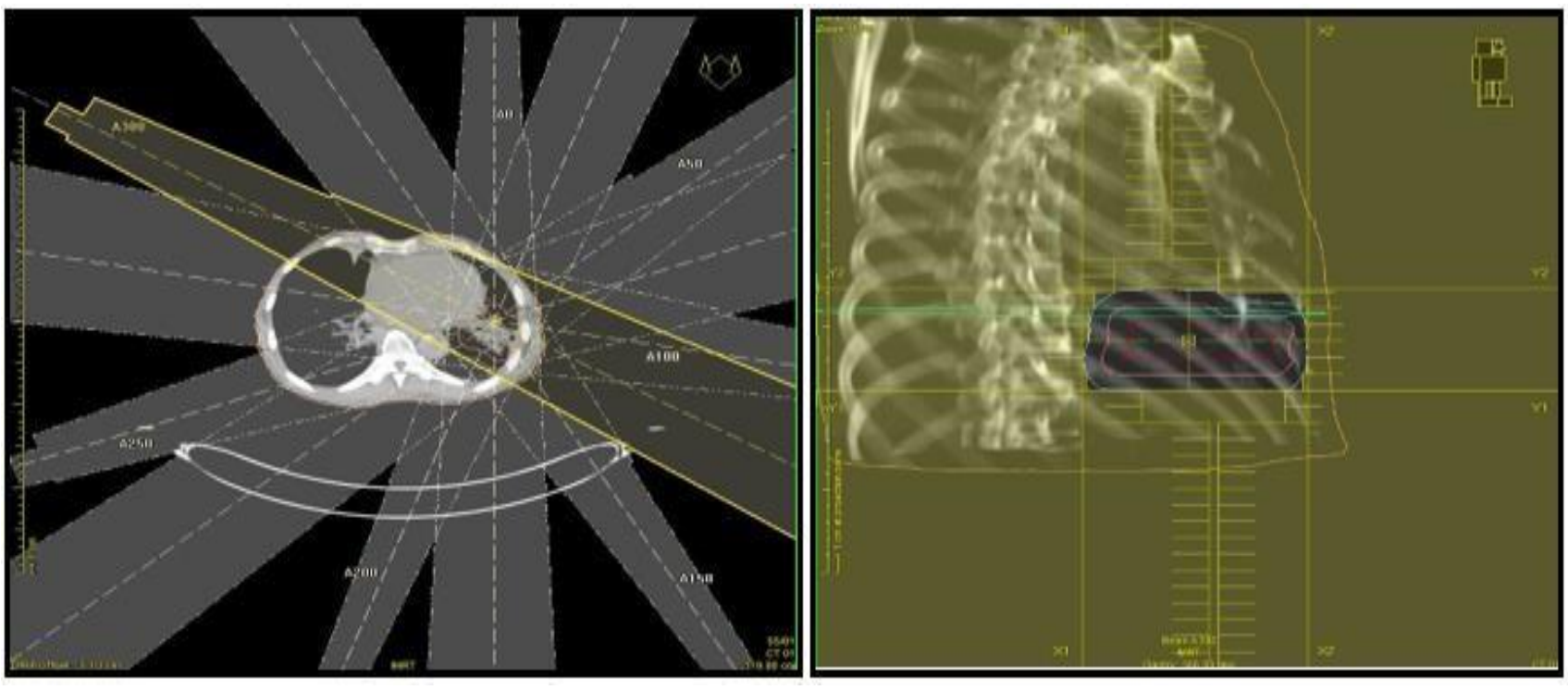
(A) Histopathological picture depicting Non-Hodgkin's lymphoma –Extra Nodal Marginal Zone Lymphoma



(B) CT image showing Extra Marginal Zone Lymphoma involving Left Lower lobe of Lung

Therapeutic intervention:

After giving proper counselling about disease & treatment, patient was started on Chemotherapy with R-CVP (Inj. Rituximab + Inj. Cyclophosphamide + Inj. Vincristine + Tab. Prednisone) had given Q3wk for 6 cycles. After completing 6 cycles of chemotherapy, he was planned for radiation therapy. The planning CT scan was fused with the Pre-Chemo PET-CT scan & Radiation therapy was delivered on a 6MV linear accelerator, with a dose of 36Gy in 18# to the involved site using IMRT technique with regular monitoring of the patient.



(C) Image Showing IMRT plan for Extra nodal Marginal Lymphoma of Left Lung

Discussion

Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT), also known as MALT lymphoma, comprises 6 % to 8 % of all non-Hodgkin lymphomas (NHL) **[2]**. MALT lymphomas most commonly affect the stomach; however, it has been reported in virtually all tissues. Extranodal marginal zone lymphoma may occur in many sites including the gastrointestinal tract, and less commonly, the salivary glands, thyroid, respiratory tract, skin, ocular adnexa, genitourinary tract, and breast **[3]**. MALT lymphoma originating in the lung, also known as pulmonary marginal zone lymphoma (PMZL), is a rare disease representing 9 % –14 % of MALT lymphomas **[2]**. However, it is the most common NHL affecting the lung. The chronic antigenic stimulus is considered the underlying cause of PMZL. PMZL is associated with smoking in 35 % – 45 % of patients. Respiratory symptoms are usually present at the time of PMZL diagnosis and diverse patterns of lung abnormalities are observed in imaging studies. The diagnosis of extranodal marginal zone lymphoma, of any site, is problematic due to the extensive overlapping features with reactive lymphoproliferative processes **[3]**. Despite these limitations in interpreting the pathologic features alone, correlation with the clinical, laboratory, and CT findings frequently allows the diagnosis of extranodal marginal zone lymphoma to be made with confidence **[3]**. Pulmonary MALT lymphoma staging featured the following evaluations: comprehensive medical history, physical examination, and radiologic imaging, including computed tomography (CT) scan of the chest and abdomen and positron emission tomography/CT **[4]**. Patients with pulmonary MALT lymphoma were managed by a multidisciplinary team comprising several specialists, including a pulmonologist, thoracic surgeon, radiologist, and oncologist. Pulmonary marginal zone lymphoma carries a favourable outcome with a 5-year overall survival (OS) rate of 90 %. Treatment includes a multidisciplinary team includes surgery, chemotherapy, and Radiation therapy depending on the stage and condition of the patient. In this case, the reported patient was treated with Rituximab-based chemotherapy followed by radiation therapy to the involved site which has shown very good response on following up the patient for 1 month, 3 months, and 6 months post-chemo-Radiation therapy.

Conclusion

The presentation of Extranodal marginal zone lymphoma of the lung shares clinical features of chronic infection and metastatic cancer. The disease responds favourably to the R-CVP regimen. Estimated survival Rates at 5 and 10 years are reported as 90 % and 72 % respectively but recurrence is as high as 50 % and ongoing survival surveillance is thus crucial. However, future studies with larger numbers of patients are needed to fully evaluate the role of different modality approaches in the treatment of Extranodal marginal zone B-cell lymphoma.

Patient Perspective: Satisfactory

Informed consent: Informed consent was taken prior to Imaging and Prior management.

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