

# Primary Pulmonary Lymphoma with Metastases – A Rare Presentation

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**Abstract** Primary pulmonary non-Hodgkin's lymphoma is a very rare neoplasm. It is represented most commonly by marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT) type. We here in report a case of 64 year old male presented with a lung mass that was diagnosed to be NHL, with metastases to liver and kidney. It represents only 3-4% of extranodal NHL, less than 1% of NHL, and only 0.5- 1% of primary pulmonary malignancies. Current treatment options are surgery, chemotherapy, and radiotherapy.

**Keywords:** lymphoma, primary, lung

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## 1. Introduction

The most common extranodal site of presentation for nonHodgkin's lymphoma (NHL) is the gastrointestinal tract, including stomach. Primary NHL of the lung is very rare, accounting for only 0.4% of all malignant lymphomas [1].

Primary pulmonary NHL is most commonly represented by marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue type (MALT lymphoma) [1]. Their development depends on MALT of the bronchus that is thought to be acquired as a result of chronic antigenic stimulation such as smoking, autoimmune disease or infection [1]. They usually pursue indolent courses, remaining localized to the lung for long periods before dissemination [1].

A good deal of progress has been made in understanding the pathophysiology of primary pulmonary lymphoma. The role of Epstein-Barr virus, for example, is now well documented in some cases of high-grade B-cell lymphoma and lymphomatoid granulomatosis. Similarly, it is possible that an infectious agent plays a role in the emergence of pulmonary mucosa-associated lymphoid tissue lymphoma (analogous to *Helicobacter pylori* in the stomach). The diagnosis of these clonal lymphoid proliferations has also benefited from advances in immunohistochemistry and molecular biology [2].

The current definition of PPL covers: 1) low-grade B-cell PPL (PPL-B), the most frequent form; 2) high-grade PPL-B; and 3) lymphomatoid granulomatosis (LG), a rare disorder [2].

## 2. Case Report

A 69-year-old man was referred with the diagnosis of a bronchogenic squamous cell carcinoma obtained by bronchoscopic biopsy at another hospital. The patient was a chronic smoker, presenting with off and on dyspnoea, hemoptysis, pain abdomen, loss of weight, decreased appetite. On physical examination, breath sounds were diminished on the left lower zones of the lung. No lymphadenopathy seen. In addition, he had an apparent hepatomegaly.

On investigation, hemogram was normal. Chest X-ray showed a pneumonic consolidation



**Figure 1.** Chest Xray showing consolidation in left lower zone on the left lower zone (Figure 1). Bronchoscopy showed no endobronchial lesion. Cytologic examination

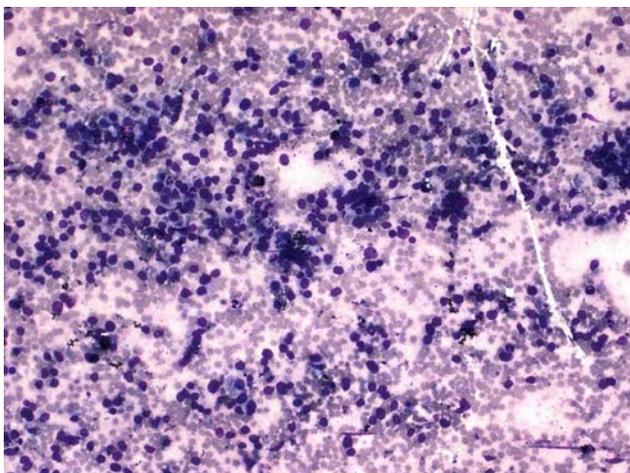
of the bronchoalveolar lavage and sputum was not definitive. Ultrasound (USG) abdomen showed multiple hepatic and bilateral renal space occupying lesions. On CT, evidence of multiple peripherally enhancing nodular lesions seen scattered in both lobes of liver, largest

measure 3.5cm, suggestive of metastasis (Figure 2). It also revealed multiple non enhancing hypodense nodular lesions in kidney, and large lobulated mass in left lower lobe of lung.

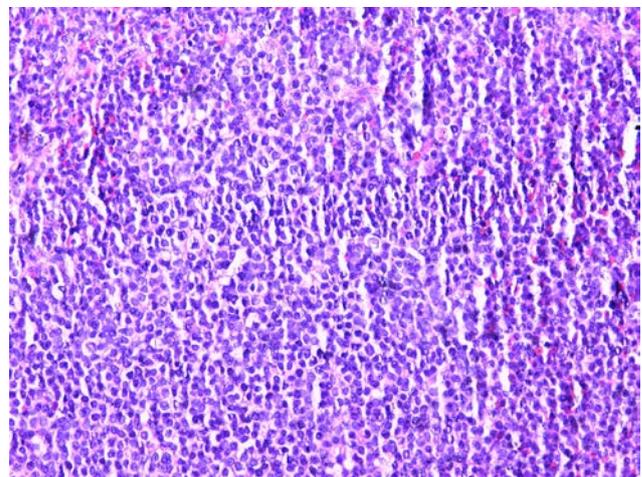


**Figure 2.** CT scan showing liver metastases

Fine needle aspiration(FNA) from lung mass showed cellular smears, suggestive of malignant small round cell tumor (Figure 3). Liver FNA performed showed scanty cellularity but showed lymphoid cells of similar morphology as seen in FNA lung. Histological examination showed a primary pulmonary lymphoma without any involvement of hilar or mediastinal lymph nodes (Figure 4). Immunohistochemical staining with CD20, CD23, CD 43, CD79a showed positivity on the lymphoid cells, and revealed a primary lowgrade marginal zone B cell lymphoma of lung. The patient was referred to medical oncology for further chemotherapy. There was no second malignancy detected despite exhaustive work-up.



**Figure 3.** FNA smear showing small round cell tumor (Leishman%2c 200x)



**Figure 4.** Diffuse sheets of neoplastic small round lymphocytes (200X, H&E)

### 3. Discussion

PPL is very rare, it represents only 3-4% of extranodal NHL, less than 1% of NHL, and only 0.5-1% of primary pulmonary malignancies. The incidence of primary pulmonary lymphoma peaks in the 6<sup>th</sup> and 7<sup>th</sup> decades of life, and the ratio of male to female patients is close to 1:1. Clinical description is nonspecific. When present, symptoms are various, such as cough, mild dyspnea, chest pain, and occasionally hemoptysis. The radiographic findings are nonspecific also and include solitary nodule, multiple ill-defined nodules, consolidated mass with air

bronchograms, pleura effusions, atelectasis, and cavities. PPL originate frequently from the B cell-lineage which is represented most commonly by marginal zone B-cell lymphoma of MALT of the bronchus that is thought to be acquired as a result of chronic antigenic stimulation such as smoking, autoimmune disease, or infection [3].

Reactive lymphoid proliferations such as pseudolymphoma, lymphoid interstitial pneumonitis, lymphomatoid granulomatosis, and follicular bronchiolitis are morphologically difficult to distinguish from primary malignant lymphoid tumors [1].

A variety of histologic subtypes of NHL may manifest as primary pulmonary lymphoma. The most common histologic subtypes of primary pulmonary lymphoma are low-grade lymphoproliferative processes that are well-differentiated B-cell tumors that appear to arise from bronchus associated lymphoid tissue (BALT). BALT forms part of the wider system of low-grade malignant lymphomas of MALT type such as those found in the gastric area. This form may be referred as a subtype of marginal-zone Bcell lymphoma [2].

The second most frequent histologic type of non-Hodgkin lymphoma to involve the lung is diffuse large Bcell lymphoma [1,2].

The prognosis of patients with primary pulmonary NHL is known to be relatively favorable [4]. The treatment options include watch and wait approach, surgery in localized tumors, chemotherapy if the lesions are diffuse or involve both lungs and radiotherapy. But in the absence

of comparative series, the efficacy of these treatments cannot be analyzed [3]. Metastases in liver and kidney from Pulmonary lymphoma has not been mentioned in literature to best of our knowledge, However, in a study, recurrences and metastases in the lung, stomach, lymph nodes and salivary glands were seen in about 46% of the cases of low-grade B-cell lymphoma of the bronchus-associated lymphoid tissue.

## 4. Conclusion

In conclusion, primary non-Hodgkin's lymphoma is a rare entity of the lung. An open thoracotomy is very likely to provide a diagnosis as well as a therapeutic resection.

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