CASE REPORT

Primary Pulmonary Lymphoma Presenting as Lung Mass

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ABSTRACT

The commonest cause of lung mass in an elderly patient is bronchogenic carcinoma. We are reporting an unusual case of lung mass that was diagnosed following exploratory thoracotomy and pneumonectomy. Sputum examination, bronchoscopy and percutaneous fine needle aspiration cytology were inconclusive. On histopathology, a diagnosis of non-Hodgkin’s lymphoma (NHL) was made. There was no involvement of any other site on detailed work up. The patient was advised chemotherapy.

Key words: Lymphoma, Bronchoscopy, Lung mass.

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INTRODUCTION

For investigation of a lung mass seen on radiography, a sputum examination, followed by fibreoptic bronchoscopy and/or percutaneous diagnostic procedure is performed to achieve a microbiological or histopathological diagnosis. Occasionally, when bronchoscopy is inconclusive, the mass seems resectable and there is no evidence of distant metastasis, exploratory thoracotomy is performed both for diagnosis and treatment. Primary pulmonary lymphoma (PPL) is a rare disorder, often clinically latent, and an accurate pre-operative diagnosis is difficult because of non-specific clinical presentation and radiographic findings. Primary non-Hodgkin’s lymphoma (NHL) of the lung affects elderly patients and often the diagnosis is made on thoracotomy or thoracoscopy¹. Since the condition is very rare, there are no specific treatment guidelines and it is done as for NHL in other organs of the body.

CASE REPORT

A 75-year-old housewife, a nonsmoker, presented with five months history of progressively increasing shortness of breath, chest pain, vague ill health and easy fatigability. On direct questioning she complained of undocumented low-grade fever and dry cough. There was no history of haemoptysis. She received antibiotics without any response. On examination, she was afebrile and there was no clubbing, oedema feet or peripheral lymphadenopathy. There was a dull note on percussion and reduced air entry in the left lower chest.

On investigation, hemogram, renal and liver functions were within normal limits. Chest radiograph revealed a homogeneous shadow in the left lower zone suggesting a mass with pleural effusion (Figure 1). Contrast enhanced CT scan of the chest revealed a mass lesion in the left lower lobe with minimal pleural effusion (Figure 2). Ultrasound of the abdomen was
normal. Sputum for acid fast bacilli (AFB) and malignant cells was negative on three occasions each. On bronchoscopy there was a polypoid lesion in the left lower lobe from which a biopsy was taken which was inconclusive. Bronchoscopic aspirate was also negative for AFB and malignant cells. A percutaneous fine needle aspiration of the lung mass was also inconclusive and patient was taken up for exploratory thoracotomy.

At surgery, there was a hard mass of 6 x 6 cm size in the left lower lobe which was extending up to secondary carina and lower part of the left main bronchus. There were multiple enlarged lymph nodes around the left main bronchus. There was minimal pleural effusion without any pleural deposits. The mass was free from the chest wall, diaphragm and mediastinal structures. In view of these operative findings, a left pneumonectomy and removal of all the enlarged lymph nodes was performed. The post-operative period was uneventful.

Histopathological examination revealed sheets of small lymphocytes with occasional plasmacytoid features infiltrating and obscuring the pulmonary alveoli (Figure 3). At places the mucus glands of the bronchi were completely infiltrated by these cells. Hilar lymph nodes also revealed involvement by the tumour. The tumour cells were immunohistochemically positive for the B-lymphocyte marker, CD20+. A diagnosis of primary pulmonary non-Hodgkin’s lymphoma, diffuse, B-cell immunophenotype was established. Work-up for involvement of other sites including bone marrow examination was negative. Patient was treated with chemotherapy (CHOP regimen). She developed septicaemia and had upper gastrointestinal haemorrhage and succumbed to these complications.

Figure 1. Chest radiograph showing a mass lesion in the left lower zone. A co-existing pleural effusion can not be ruled out.

Figure 2. Contrast enhanced CT scan of the chest (Bone Window) showing mass in the left lower lobe with minimal pleural effusion.

Figure 3. Section from lung showing a diffuse infiltrate of lymphocytes obscuring alveolar spaces and alveolar septae. The infiltrates have relatively well circumscribed margins in this field and normal uninvolved alveoli are identified with carbon pigment (H & E x 10).
DISCUSSION

Although diagnosis of primary pulmonary lymphoma (PPL) have been achieved with bronchoalvelor lavage, bronchial biopsy and percutaneous biopsies in some case reports, the majority of cases require thoracoscopic or open lung biopsy1-3. The diagnosis of PPL in the present case was based on the criteria described by L'Hostel4 which is a modification of original Salzstein criteria5. Two pre-requisite features are: (1) involvement of lobar, lobar or mainstem bronchus either unilaterally or bilaterally with or without hilar or mediastinal involvement and (2) no evidence of extrathoracic lymphoma at the time of diagnosis or for three months following diagnosis4. Primary lymphomas arising from the bronchus (Mucosa) associated lymphoid tissue (MALT) are rare accounting for 3.6% of all extranodal lymphomas and less than one per cent of all malignant lymphomas5. Previously they were classified as pseudo-lymphomas since they were more often found to be solitary lesions, had indolent course and a favourable prognosis6.

However, modern immunohistochemical and molecular genetic methods have revealed that these tumours contain clonal population of lymphoid cells, majority being monoclonal B cell proliferation, thereby representing true lymphomas7. Lung is a rare site for extranodal lymphoma8. The lymphomatous involvement of the lung can occur by either extension of nodal disease into the lung9 or by involvement of MALT10, which includes closely related bronchus associated and gut associated lymphoid tissue. It is more commonly seen in between 5th and 7th decade with a mean age of 50 years and a male to female ratio of 1.07-1.011. Patients may have non-specific symptoms such as cough with or without expectoration, shortness of breath on exertion and chest pain. Fever, weight loss and night sweats may also be present. In most series, leucocyte counts and differential counts were within normal limits and no leukemic phase of lymphoma was demonstrated in any series12. Biochemical tests are also reported to be within normal limits12. Involvement of lung usually is diagnosed only at routine chest radiography. Pleural effusion is uncommon in patients with NHL (10%)12, however, our patient had minimal pleural effusion. An association with autoimmune disorders such as systemic lupus erythematosus, Sjogren’s syndrome or Hashimoto’s thyroiditis has been described12.

The staging system most commonly adopted is a modified Ann Arbour staging classification to suit lymphoma of the lung13. According to this staging system our patient was in stage II 2E. The treatment of primary pulmonary NHL is surgical as suggested by Salzstein5. The chemotherapy of choice for aggressive lymphoma is CHOP therapy which can result in cure rate of 30-40% even in stage III and IV. Good symptom control and radiological response was achieved with chemotherapy in disseminated low grade lymphoma by Toh and Ang8. The prognosis is marginally better with MALT type of PPL as compared to non-MALT type of lymphoma. No prognostic factors have been identified1.

Primary pulmonary lymphoma, though rare, is a distinct clinicopathological entity. Most of these are low grade B cell lymphomas arising from mucosa associated lymphoid tissue (MALT). The symptoms are non-specific and the chest radiograph gives the first clue to lung involvement. Treatment depends on the stage of the disease. PPL should be a differential diagnosis in patients with atypical shadows where there is difficulty in achieving diagnosis by sputum examination, bronchoscopy and FNAC.

REFERENCES


