

Case Report

A rare case of primary pulmonary Hodgkin's lymphoma

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ABSTRACT

A 46 years old male presented with fever, chest pain, dry cough, weight loss, and breathlessness over the preceding 3 months. CT scans of thorax revealed diffuse sclerosis with multiple ill-defined small erosion and pleural thickening of the right lung with bulky lymph node in a subcarinal region measuring approximately 13.5×11×27.5 mm and 20×5 mm soft tissue parenchymal lesion in a right apical zone. Examination of pleural fluid cytology was not diagnostic. Bronchoscopic fluid cytology was also not contributory. Hilar and pre aortic lymph node biopsies showed only reactive change. Video-assisted thoracoscopic surgery (VATS) with biopsy from all suspicious areas of lung and pleura was taken, which on evaluation confirmed the diagnosis of primary pulmonary Hodgkin's lymphoma. We present here a rare case of primary pulmonary Hodgkin's lymphoma with a review of the literature.

Keywords: VATS, PPHL, Hodgkin's lymphoma

INTRODUCTION

Primary pulmonary lymphoma is defined as clonal lymphoid proliferation affecting one or both lungs (parenchyma and/or bronchi) in a patient with no previous extrapulmonary involvement at the time of diagnosis or during the subsequent 3 months. Primary lymphoma of the lung is a rare disorder and represents only 0.3% of all primary pulmonary malignancies, less than 1% of all the cases of non-Hodgkin lymphoma, and 3% to 4% of all the extra nodal manifestations of non-Hodgkin lymphoma.¹ Lymphomas in the lung are usually diagnosed by video-assisted thoracic surgery, open-wedge biopsy, or lobectomy. The lung is one of the more common sites where extra nodal lymphomas arise, following the gastrointestinal tract, skin, and nervous system.¹ Primary pulmonary lymphoma is diagnosed when the lung, bronchus, or both are involved without evidence of mediastinal adenopathy or a mass on the chest imaging and without any diagnosis of extra thoracic lymphoma previously or during the subsequent 3 months. Most cases of primary pulmonary lymphoma of the lung

are of B-cell lineage, and the disease is frequently located in the bronchus-associated lymphoid tissue. These neoplasms are most commonly seen in adults with a median age of about 60 years and rarely develop in children. Most studies show a slight male predominance. Patients often present with pulmonary symptoms such as dyspnea, hemoptysis, chest pain, or constitutional symptoms. However, a majority of low-grade primary pulmonary lymphoma patients are asymptomatic. Radiologic findings of primary pulmonary lymphoma are non-specific, with single or multiple, unilateral or bilateral lesions forming nodules, masses, or infiltrates resembling consolidated lung.¹ Few patients (less than 10%) develop pleural effusions.

CASE REPORT

A 45 years old male patient presented with complaints of breathlessness, fever, and vague right-sided chest pain for the past 3 months and was admitted to SMIMER Medical College and Research Institute in February 2020. Clinically there was no evidence of peripheral

lymphadenopathy or splenomegaly. Respiratory system examination revealed decreased air entry over the right chest. The patient had a history of smoking. He was a social drinker and worked in a cloth factory. There was no history of dental manipulation, hospitalizations, or surgical procedures. The patient has no family history of carcinoma.

On presentation, the patient's body temperature was 36°C (96.8°F). His blood pressure was 120/76 mmHg. The heart rate of 110 beats/min. The physical examination was normal.

His hemogram showed a hemoglobin level of 112 g/l; a haematocrit of 31.1%; a white blood cell count of $29.6 \times 10^9/l$ (82% neutrophils and 9.9% lymphocytes) with no other abnormalities; and an erythrocyte sedimentation rate of 127 mm/h. Biochemistry did not show abnormalities except for a low serum iron level 21.48 $\mu\text{mol/l}$, a ferritin level of 707.80 p/mol/l, an albumin level of 25 g/l, and a lactate dehydrogenase level of 201 U/ml. Microbiological testing of bronchial alveolar lavage fluid and bronchial washings did not reveal any pathogens. Serology for hepatitis and HIV were negative.



Figure 1: Chest X-ray showing right pleural effusion.

Chest X-ray revealed right-sided pleural effusion with the underlying collapse of the lung (Figure 1) for which pleural tapping was done and sent for pleural fluid cytology and no evidence of dysplasia/malignancy seen in fluid. CT scan report of the chest revealed diffuse sclerosis with multiple ill-defined small erosion and pleural thickening of the right lung with bulky lymph node in the subcarinal region measuring approximately 13.5×11×27.5 mm. And 20×5 mm soft tissue parenchymal lesion in the right apical zone (Figure 2).

In the bronchoscopy report, the right lung was grossly normal with few nodules in the left upper and lower lobe, and the patient was referred to the surgery department for lung biopsy and the patient underwent video-assisted thoracoscopic surgery (VATS) with lung biopsy.

Biopsy were taken from multiple whitish nodules on lung surface, parietal pleura of upper lateral chest wall (Figure 3 and 4) and sent for histopathological and IHC examination.

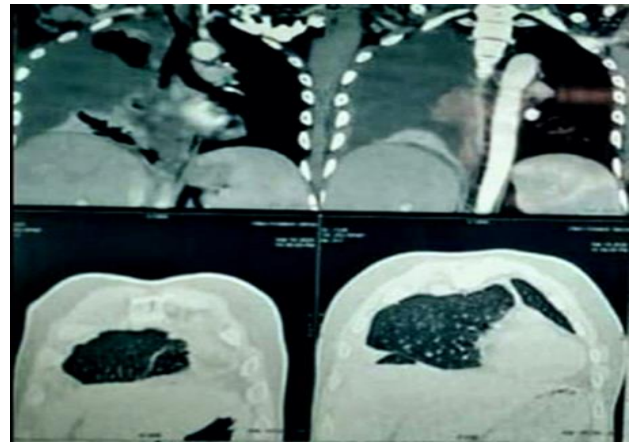


Figure 2: CT scan showing diffuse sclerosis, pleural thickening of right lung, and fibro calcific nodules in both lungs.

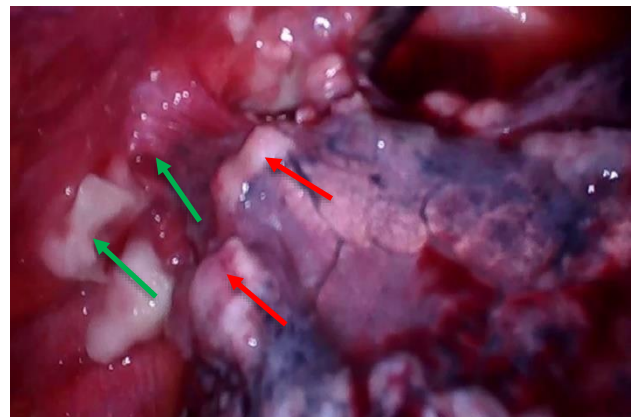


Figure 3: Multiple nodules on lung surface (green arrow) and parietal pleura (red arrow) intra operative findings on VATS.

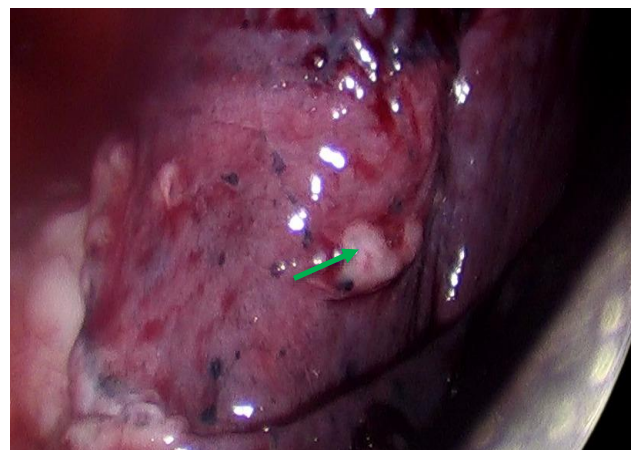


Figure 4: Multiple nodules on lung surface (green arrow).

Initially histopathology report reported as a mesothelioma. However, second pathology report with additional immunohistochemistry noted positive for CD30, PAX5, CD15 and background lymphocytes are predominantly CD3 positive suggestive of classical Hodgkin's lymphoma (Figure 5) and, showing a polymorphous lymphoid infiltrate with surrounding fibrosis and sharp interface with adjacent lung parenchyma (Figure 6).

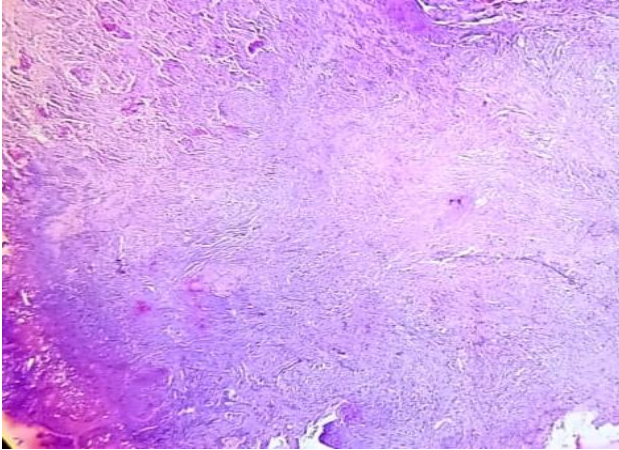


Figure 6: A rare occurrence in the lung, showing a polymorphous lymphoid infiltrate with surrounding fibrosis and sharp interface with adjacent lung parenchyma.

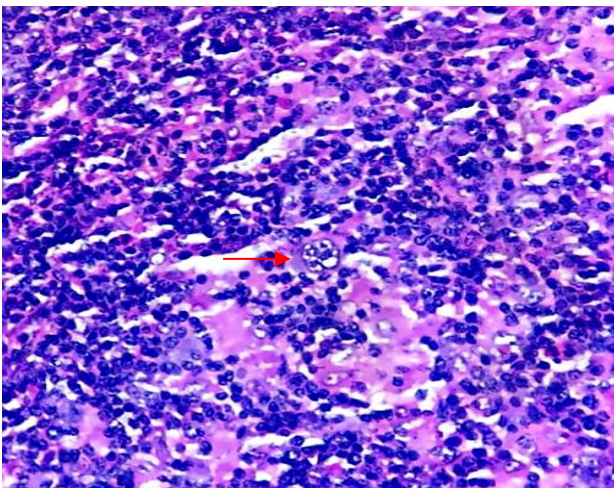


Figure 5: Higher power shows Reed-Sternberg and Hodgkin cells admixed with eosinophils, lymphocytes, plasma cells, and rare neutrophils.

The initial clinical presentation and history led to a provisional diagnosis of pulmonary tuberculosis initially leading to start of empirical treatment for mycobacterium tuberculosis infection with four drug regimen.¹

After final diagnosis, the patient was referred to the cancer department for further treatment, but patient died during the early course of treatment.

DISCUSSION

The criteria for the diagnosis of PPHL include; histological features of Hodgkin's lymphoma, restriction of the disease to the lung with or without minimal hilar lymph node involvement and adequate clinical and/or pathological exclusion of the disease at distant sites.^{1,2} Our case meets these criteria.

Primary pulmonary lymphomas originate from mucosa-associated lymphoid tissue arising from lymphoid follicles or peri bronchial lymph nodes and extending to the parenchyma, and are therefore described as separate entities from peripheral nodal lymphomas and from lymphomas affecting the lungs secondarily.¹ These tumors may be a type of Hodgkin's (PPHL) or non-Hodgkin's lymphoma (NHL).¹ Primary pulmonary lymphomas are rare, and of these, only 1.5-2.4% are attributable to PPHL.^{2,3}

Hodgkin lymphoma is most commonly seen in the lung as secondary involvement. Hodgkin lymphoma affects young adults with a mean age of 42 years and with slight female predisposition.

These patients present with single or multiple parenchymal masses, endobronchial lesions, or pneumonia like consolidation. It is difficult to diagnose HL in lung, especially, on scant endobronchial tissue or transthoracic needle biopsies or in frozen sections, because of the lack of identification of Hodgkin cells or Reed-Sternberg cells in a small sample. The diagnosis of HL in lung is based on the recognition of diagnostic Reed-Sternberg cells within the appropriate reactive cellular infiltrate in the background. Nodular sclerosis and mixed cellularity are the common histologic types seen. Differential diagnoses for classic HL include solitary fibrous tumor with extensive inflammation and inflammatory myofibroblastic tumor. Immunohistochemically, the tumor cells are positive for CD15, CD30, Pax5, and rarely CD20, and negative for T-cell markers.^{2,3} Factors that correlate with a poor prognosis include age greater than 60 years, B symptoms, multiplicity and bi-laterality of lung lesions, pleural effusion, and cavitation.^{1,3} It is suggested that chemotherapy is recommended over radiotherapy in this disease because of the risk of radiation pneumonitis.

Radiologically, PPHL typically involves the superior portions of the lungs, whereas secondary pulmonary involvement from Hodgkin's lymphoma shows a more random miliary distribution without zonal predilection - Many presents as a solitary mass, alveolar consolidation or multiple nodules. Cavitory pulmonary lesions have a wide differential diagnosis. The present case emphasizes that no radiological appearance is pathognomic for PPHL.

Bronchoscopic evaluation is warranted to exclude other pathologies, although endobronchial lesions are rare, and

bronchial cytology often fails to yield Reed-Sternberg cells. Diagnosis usually requires an open thoracotomy and lung biopsy.

Owing to the lack of survival data, prognostic factors affecting survival of PPHL are not well defined, but several factors have been suggested: 'B' symptoms, bilateral disease, multi-lobar involvement, penetration of the pleura with or without associated pleural effusion, cavitation, age greater than 60 years and clinical relapses. 14 of 23 patients with PPHL survived and four relapsed or died, which suggests that the prognosis for PPHL is not as poor as previously reported.

Management plans vary in the literature. Although surgical biopsy is required in most cases for diagnosis, the indications for surgical treatment are poorly defined. It has been generally accepted that, for diffuse and bilateral lesions, the combination of multiple-agent chemotherapy and radiotherapy is required. Recent authors have recommended chemotherapy over radiotherapy due to the risk of radiation pneumonitis.

CONCLUSION

A definitive diagnosis of primary pulmonary Hodgkin's lymphoma (PPHL) is very difficult without lung biopsy, which can be safely done by VATS with reduced morbidity. Histopathology with IHC is a must to confirm

its diagnosis offering a better chance to the patient to receive specific treatment thus improving prognosis and preventing the spread of the disease. This entity despite its rarity has to be kept in mind as a differential diagnosis while evaluating neoplastic pulmonary lesions.

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Ethical approval: Not required

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