

# Primary pulmonary Hodgkin lymphoma

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## Abstract

Primary pulmonary Hodgkin lymphoma (PPHL) is a rare disease. Herein, we report a case of PPHL with diagnostic concerns encountered during initial evaluation which is of paramount importance to keep the differential diagnosis in cases with high index of suspicion for this rare entity.

## Introduction

Primary pulmonary Hodgkin lymphoma (PPHL) is a rare disease. It occurs when the clonal lymphoid proliferation affect the lungs, and has no extra pulmonary spread at time of diagnosis or within the following 3 months. PPHL without peripheral lymphadenopathy or hepatosplenomegaly is exceptionally uncommon.<sup>1,3</sup> Pulmonary involvement is reported to occur in 15-40% of patients with Hodgkin's disease. However, the presentation of PPHL is very rare, with just over 70 cases recorded internationally between 1927 and 2006.<sup>1</sup>

A case of PPHL is herein reported with a brief description, as a diagnostic challenge. We assessed the clinical, radiological, diagnostic and pathological features along with treatment and prognosis.

## Case Report

A 15-year-old female of Saudi origin presented with persistent fever for more than one month duration. Over a period of one year, she had productive cough, weight loss and night sweats. She had no relative past medical history and was a non-smoker.

Respiratory examination revealed marked decrease air entry with dull percussion of the left upper lung zone. Physical examination did not reveal clubbing, peripheral lymphadenopathy or hepatosplenomegaly.

Laboratory investigation revealed anemia and severe leucopenia. Chest X-ray showed a persistent homogenous opacity occupying left upper lung zone for more than 5 weeks (Figure 1A). Chest computed tomography (CT) scan images showed consolidation and collapse of the left upper lung lobe with air bronchogram and pleural thickening (Figure 1B-D). CT guided biopsy was insignificant.

Bronchoscopy revealed normal macroscopic appearance of airways and absence of endobronchial lesions or foreign body.

The internal medicine and infectious disease team suggested tuberculosis (TB) pneumonitis as an initial diagnosis and started the patient on anti TB treatment in addition to antibiotics without improvement over a period of one month (if TB is suspected then acid-fast bacilli stain and mycobacterial culture should have been done).

Patient's conditions deteriorated and she became severely ill with persistent fever, significant weight loss and marked leucopenia. Based on clinical picture and failure to response to medical treatment, we thought that this is a case of lung gangrene that will soon develop cavitation into the consolidated lobe. We decided to perform exploration and lobectomy. The patient underwent left posterolateral thoracotomy which revealed a hepaticized and necrotic left upper lung lobe with extensive adhesions to chest wall, left upper lobectomy was carried out. There was neither mediastinal nor hilar lymphadenopathy. No pleural effusion and no pleural nodules or any other lesions.

## Histopathology

Histopathological examination showed Hodgkin lymphoma (HL), nodular sclerosis type. Images (Figure 2A) of lung tissue showed sheets of polymorph cells separated by fibrous septa; Figure 2B showed the sheets composed of small lymphocytes, histiocytes, eosinophils, neutrophils, plasma cells, large multiloculated giant cells and some Reed Sternberg cells with occasional lacunar cells. Figure 2C,D shows the neoplastic Reed Sternberg cells interacting with CD15 and CD30 in an immunohistochemical analysis.

## Outcome and follow-up

After the left upper lobectomy the patient markedly improved, fever subsided and her general condition and laboratory investigation improved. She received postoperative chemo and radiotherapy. She remains well without evidence of recurrence 6 years later.

She only had a postoperative eventration of the left diaphragm, due to intraoperative injury of the left phrenic nerve (Figure 3). Patient did not have any respiratory symptoms and refused to perform plication of the diaphragm.

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## Discussion and Conclusions

Primary pulmonary lymphomas are uncommon, comprising less than 1% of lung cancers and fewer than 1% of malignant lymphomas, and accounting for only 3.6% of extranodal lymphomas.<sup>4</sup> Nodular sclerosis (NS) PPHL is the most common variety and comprises 60-70% of PPHL.<sup>5</sup>

The criteria for the diagnosis of PPHL include: i) histological features of Hodgkin's lymphoma, ii) restriction of the disease to the lung with or without minimal hilar lymph node involvement and iii) adequate clinical and/or pathological exclusion of the disease at distant sites.<sup>1,4-7</sup> Our case meets each criteria as mentioned above.

In the largest report, PPHL showed a slight female preponderance (1.4:1 F:M), with a bimodal age distribution (<35 and >60 years). The most common presenting symptoms are weight loss, fever, night sweats and dry cough. Dyspnea and hemoptysis are also common. 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 present as a solitary mass, alveolar consolidation or multiple nodules. Cavitory pulmonary lesions have a wide differential diagnosis.<sup>6</sup> The present case emphasizes that no radiological appearance is pathognomic for PPHL. The principal radiographic differential diagnosis of

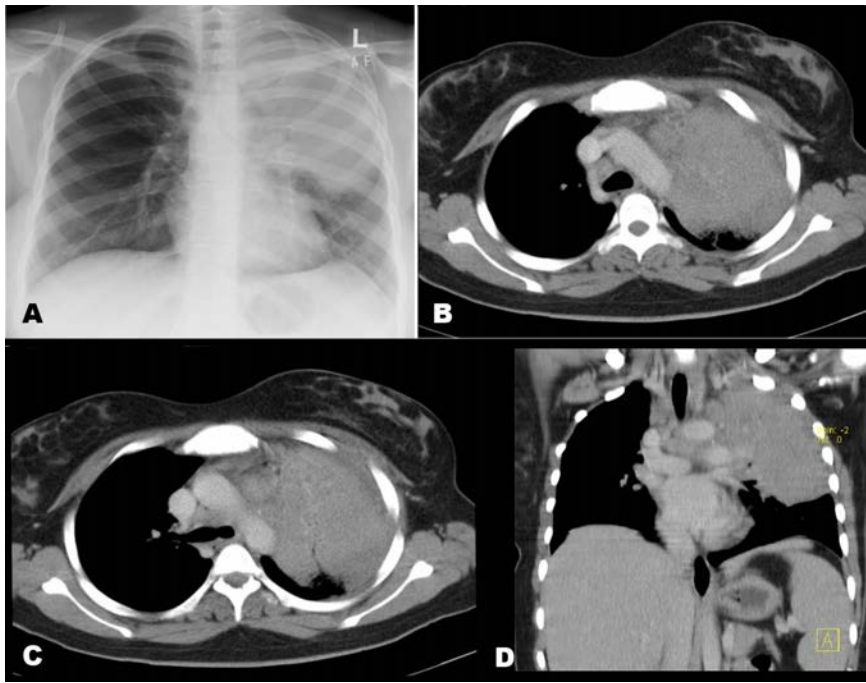


Figure 1. A) Chest x-ray showing a homogenous opacity occupying the left upper lung zone. B-C) Axial and reconstruction computed tomography images are showing consolidation and collapse of the left upper lung lobe and lingula with air bronchogram.

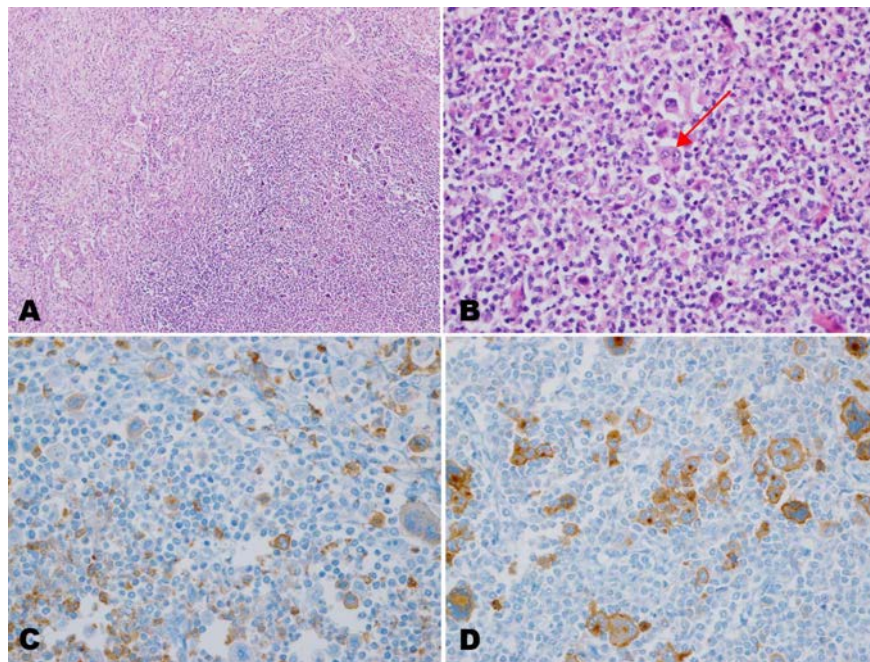


Figure 2. A) Lung Hodgkin lymphoma, large atypical Reed Sternberg like cells in background of inflammatory cells infiltration surrounded by fibrous septa on  $\times 200$  magnification. B) Lung Hodgkin Lymphoma, diagnostic binuclear Reed Sternberg cells with eosinophilic inclusions like nucleoli on  $\times 400$  magnification. C) Hodgkin lymphoma, immunohistochemical analysis showing clusters of Reed Sternberg cell react (cells react) with CD15 on  $\times 400$  magnification. D) Hodgkin lymphoma, immunohistochemical analysis showing clusters of Reed Sternberg cell reacts (cells react) with CD30 on  $\times 400$  magnification.

primary Hodgkin's lymphoma includes pseudolymphoma, lymphocytic interstitial pneumonia, lymphoid granulomatosis, bronchioloalveolar carcinoma, metastasis, and cryptogenic organizing pneumonia.<sup>8</sup>

The Ann Arbor pulmonary lymphoma staging system was used for classification. Stage IE: lung only, could be bilateral; Stage II 1E: lung and hilar lymph nodes; Stage II 2E: lung and mediastinal lymph nodes; Stage II 2EW: Lung and chest wall or diaphragm; Stage III: lung and lymph nodes below the diaphragm; Stage IV: diffuse.<sup>9</sup>

According to Ann Arbor staging system, a patient with PPHL would be either stage IE (involvement of a single extra nodal site) or IIE (localized involvement of extra nodal site and its contiguous lymph node chain).<sup>10</sup>

Hence the clinical presentation in primary pulmonary HL is non-specific, open-lung biopsy is needed in the majority of cases. Occasionally, FNA coupled with IHC stains is helpful. The microscopic appearances are typical of classic HL usually of either nodular sclerosis or mixed cellularity type, and IHC staining's are confirmatory.<sup>11</sup>

Because of the low incidence of primary pulmonary lymphoma, a high index of suspicion is required to diagnose on time and avoid unnecessary delay. As demonstrated by our case, even in the presence of classical B-symptoms, the clinical and radiological data was inconclusive for accurate diagnosis of pulmonary lymphoma. The condition suggested acute necrotizing pneumonia and lung gangrene with obstruction of blood supply to the affected lobe as a cause of failure to respond to medical treatment. This was actually our indication for surgical management and lobec-



Figure 3. Follow up chest x-ray, six years after the surgery and finishing the chemo and radiotherapy.

tomy. The correct diagnosis was made after surgical resection. Our experience demonstrates that suspicious lesions should undergo open lung biopsy or surgical resection for ultimate diagnosis as supported in literature as well.<sup>7,12-14</sup>

### Learning points

The key points of the present paper are: PPHL is a rare disease; nodular sclerosis PPHL is the most common variety; open lung biopsy is often required for definitive diagnosis.

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