Primary Pulmonary Hodgkin’s Disease: A Distinct Entity

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ABSTRACT

A 30-year-old lady presented with fever, dry cough and weight loss for the preceding five months. Radiological investigations revealed a solitary nodular lesion in the lingula of the left lung. Guided fine needle aspiration cytology failed to yield any diagnostic material. Bronchoscopic cytology was also not contributory. As a last resort open lung biopsy was done and a diagnosis of Hodgkin’s disease was made. Hilar and para aortic lymph node biopsies showed only reactive change. The final diagnosis was primary pulmonary Hodgkin’s disease. [Indian J Chest Dis Allied Sci 2006; 48: 139-141]

Key words: Lung neoplasms, Hodgkin’s disease, Diagnosis.

INTRODUCTION

Primary pulmonary Hodgkin’s disease (PPHD) is a rare entity and has to be distinguished from the more common intrathoracic nodal Hodgkin’s disease secondarily involving the lung. Kern et al have described the criteria for the diagnosis of PPHD as follows: (i) histological features of Hodgkin’s disease, (ii) restriction of the disease to the lung without hilar lymph node involvement, and (iii) adequate clinical and/or pathologic exclusion of the disease at distant sites.

A case of solitary pulmonary lesion that fulfilled all the criteria for the diagnosis of PPHD is reported. Though the condition usually terminates as a disseminated disease, its original limitation to the lung offers a chance for definitive treatment, and therefore its recognition is of importance.

CASE REPORT

A 30-year-old female presented with dry cough, on and off fever and weight loss of five months duration. Clinically there was no evidence of peripheral lymphadenopathy or hepatosplenomegaly. Respiratory system examination revealed decreased air entry over the left anterior chest.

INVESTIGATIONS

Laboratory investigations revealed a haemoglobin of 9.5 g/dl, total leucocyte count of 20,300/mm³ with neutrophilic leucocytosis and erythrocyte sedimentation rate of 84 mm at the end of first hour.

Chest radiograph (PA view) revealed a homogenous opacity in the left hilar and parahilar region with obliteration of the left heart border. Computerised tomographic scan (CT scan) of the chest showed a well-defined lobulated soft tissue attenuation lesion in the left paramediastinal plane, superior to the hilum and lateral to the pulmonary trunk. Contrast study revealed non-homogenous enhancement. There was no evidence of mediastinal or hilar lymphadenopathy (Figure 1). Air bronchogram was seen in the periphery.

CT guided fine needle aspiration cytology (FNAC) was done thrice. Initially it revealed only blood and mesothelial cells. However, third time it showed the...
presence of lymphoid cells, but a definitive diagnosis could not be made. Bronchoscopy showed oedematous hyperaemic mucosa of the lingular bronchus with no evidence of endobronchial lesion. Bronchial brushing was reported as inflammatory smear. Stain for acid-fast bacilli (AFB) was negative. Ultrasonography abdomen showed no abnormality. A clinical diagnosis of round pneumonia in the lingular lobe possibly tuberculosis was made.

At exploratory thoracotomy a large tumour was seen in the anterior segment of left upper lobe and inferior segment of lingula, adherent to the pericardium, vagus and phrenic nerves. Incisional biopsy of the tumour was done. Enlarged hilar and pre aortic lymph nodes were biopsied. Scrape smear of the lung biopsy was done and stained with rapid haematoxylin and eosin (H & E) stain. The presence of characteristic Reed-Sternberg cells and its variants in a background of lymphocytes, plasma cells, neutrophils and eosinophils enabled a diagnosis of Hodgkin’s disease. Frozen section correlated with the scrape smear diagnosis. H & E stained sections showed lung parenchyma with nodules of tumour tissue separated by broad collagenous fibrous tissue bands. Within the nodules were lacunar cells, mononuclear Hodgkin’s cells and few classical Reed-Sternberg cells with lymphocytes, histiocytes, neutrophils and eosinophils in the background (Figure 2). Necrosis was not seen. Adjacent lung parenchyma showed nodular aggregates of lymphoid tissue in the peribronchiolar and interstitial tissue. The lymph nodes were reactive and showed no evidence of Hodgkin’s disease.

On immunohistochemistry Reed-Sternberg cells and its variants were CD15 and CD30 positive. A final diagnosis of primary pulmonary Hodgkin’s lymphoma (nodular sclerosis) was made.

Bone marrow examination was done before initiating chemotherapy and was found to be normal. Following chemotherapy, the lung lesion regressed and the patient is free of the disease for almost a year.

DISCUSSION

The entity of PPHD without lymph node involvement is exceedingly rare. Less than 100 cases are reported so far. Radin2 in his review of 60 cases recorded in the world literature and adding one of his own case, has described this lymphoma as affecting women more frequently than men and that it shows bimodal age distribution (<35 yrs and > 60 yrs). Dry hacking cough is the most common presenting symptom. It typically involves the superior portions of the lung. Radiologically, it appears as a solitary mass or multinodular disease. Inhomogeneity or cavitation of these lesions is common. Compared to non-Hodgkin’s lymphoma, Hodgkin’s disease more often presents as a nodular lesion3. Rarely, it manifests as a diffuse infiltrate along the lymphatic routes. Sometimes an interesting pattern may be noted at the periphery of nodular masses: pneumatic consolidation of architecturally intact air spaces by fibrin, fibroblasts in a mucopolysaccharide-rich matrix, and a cellular infiltrate of Hodgkin’s disease. Since the presentation of this disease is non-specific, and as non-invasive tests are rarely revealing, diagnosis requires open thoracotomy and lung biopsy.

Histopathologically, it is most often-nodular sclerosis type followed by mixed cellularity type. Bronchi are frequently involved, with resultant obstructive changes distally, but presentation, as endobronchial lesion is rare4. Bronchoscopy and bronchial cytology is unrevealing in most of the cases5. Boshnakova et al reported that bronchoscopic biopsies were inconclusive in their two cases with endobronchial involvement, due to lack of Reed-Sternberg cells. However, bronchoscopy is recommended to exclude infectious diseases and other neoplastic disorders like bronchogenic carcinoma.

The differential diagnosis includes tuberculous and fungal granulomas, Wegener’s granulomatosis, Langerhans histiocytosis, metastatic carcinomas and Non-Hodgkin’s lymphoma particularly T-cell lymphomas. If extensive necrosis and granulomas are seen, the presence of Reed-Sternberg cells help to exclude infectious granulomas and Wegener’s granulomatosis. Large number of eosinophils may cause Langerhans histiocytosis to be considered, but Langerhans cells lack the malignant characteristics of Reed-Sternberg cells. Undifferentiated carcinomas may show the presence of Reed-Sternberg like cells, but neutrophils are frequently seen and eosinophils are seldom present. In difficult cases immunohistochemistry resolves the diagnostic problem. Exclusion of other types of lymphoma may be impossible on routine histology. Some forms of T-cell lymphomas may simulate Hodgkin’s disease by showing extreme degree of cellular pleomorphism and a background of reactive inflammatory cells. Immunohistochemistry is required for a definitive diagnosis.
in such cases for a confirmatory diagnosis4.

Factors, which correlate with poor prognosis, include older age group, “B” symptoms, bilateral disease, multilobe involvement, penetration of the pleura and cavitations2. The staging and treatment of these lymphomas according to the extent of pulmonary involvement are recommended, as radiotherapy or combination chemotherapy may be effective in appropriately selected cases2.

To conclude, a case of PPHD presenting as a solitary nodular lesion with “B” symptoms is presented for its rarity and the difficulties encountered in arriving at the diagnosis. Open lung biopsy is necessary for a definite diagnosis. Scrape cytology is complimentary to frozen section.

REFERENCES