Case Report

Primary Pulmonary Hodgkin’s Disease—Report of two cases

Tz. BOSHNAKOVA* V. MICHAILOVA†, M. KOSS‡, Ch. GEORGIEV*, T. TODOROV*
AND M. SARBINOVA§

*Department of Pathology, †Department of Pediatric Thoracic Surgery, §Department of Pediatric Oncohematology, Medical University Sofia and ‡Department of Pulmonary and Mediastinal Pathology and Armed Forces Institute of Pathology, Washington, U.S.A.

Introduction

Primary pulmonary Hodgkin’s disease (PPHD) is a distinct entity and denotes involvement of the lung without hilar adenopathy or disseminated disease. Fewer than 100 cases of PPHD have been reported. (1–6,8,10).

The authors present two cases of PPHD with endobronchial involvement in young men illustrating the difficulties in the clinical and biopsy diagnosis.

Case 1

A 15-year-old boy presented with 6-week history of cough and fever, treated with antibiotics unsuccessfully. There were no enlarged peripheral lymph nodes and the test of the physical examination was normal. There were no biochemical or haematological abnormalities. Chest X-ray results showed nodular lesion in the right lower lobe. Computer scan revealed a tumour mass in posterior basal segment of the right lung. The mediastinum was not remarkable. Bronchoscopy showed plaque like infiltrate in the mucose of the lower part of the right lower bronchus. The biopsy specimen revealed bronchial mucose infiltrated by lymphocytes and eosinophils, findings suggestive for eosinophylic granuloma. Right thoracotomy was undertaken. A fairly hard, slightly nodular tumour was found to occupy most of the right lower lobe. The palpable mediastinal lymph nodes were moderately enlarged and measured from 5–15 mm. Several of the nodes were removed on frozen section, they were found to be with reactive changes only. In view of the fact that the tumour was entirely confined to the right lower lobe a lobectomy was performed. Grossly a solid lobulated fleshy grayish yellow tumour mass occluded the right lower lobar bronchus, occupied the major portions of the lobe and measured 10 × 5 cm. There were focal areas of necrosis and cavitations. Histologically the tumour in the lung was characterized by marked fibrosis and presence of nodular areas of lymphoid tissue infiltrated by typical Reed–Sternberg cells, atypical mononuclear cells, eosinophils and neutrophils. In the adjacent lung parenchyma there was lymph folicullar hyperplasia. The hillar and mediastinal lymph nodes showed benign reactive changes only. The immunoprofile of the tumour included focal positive staining of the atypical cells for L 26 (B-cell marker) Ki-1 and Leu M 1 and negative staining for LCA and Cam 5.2.

The histological and immunohistochemical features of the case were typical of that expected in Hodgkin’s disease-nodular sclerosing type. The patient was treated with conventional chemotherapeutic and radiotherapeutic protocols for Hodgkin’s disease and had favourable response with 3 years remission till the moment.

Case 2

A 42-year-old man had a 4-month clinical history suspicious for tuberculosis and bilateral pneumonia. He was treated by tuberculostatics and antibiotics without significant effects. At the admission to the hospital he had small haemoptysis, fever and was severely dyspnoeic. No enlargement of peripheral lymph nodes was found. There were no biochemical or haematological abnormalities. Chest X-ray revealed diffuse reticular infiltrates with shadow in the right lower lobe measured 6 cm in diameter. Bronchoscopy revealed signs of chronic inflammation of bronchial mucose. Transbronchial biopsy showed histological findings of interstitial lymphocytic pneumonitis and pneumosclerosis. The physical examination failed to disclose any evidence of lymphadenopathy. The patient died due to progressive pulmonary failure with final clinical diagnosis: idiopathic diffuse interstitial fibrosis, chronic obstructive pulmonary disease, bilateral pneumonia and arterial hypertonia. Grossly a neoplastic mass measured 6.1 × 4.5 × 4 cm occupied the lower part of the right lower lobe. The hilar lymph nodes were not particularly enlarged. Histologically masses of lymphoid tissue were seen to incorporate the right lower lobe main bronchus consisted...
of lymphocytes, Reed–Sternberg cells, atypical monocellular cells, eosinophils and neutrophils. The findings were consistent with PPHD-mixed cellularity type. Alveolar interstitium of both lungs was diffusely infiltrated by lymphocytes. The mediastinal and hilar lymph nodes and the spleen were with reactive changes only.

Discussion

Presentation of PPHD with endobronchial involvement is very uncommon. (3). Both our cases demonstrated lesion of the lung with endobronchial involvement of the right lower lobar bronchus and lack of changes of hilar or mediastinal lymph nodes. Both biopsies taken at bronchoscopies of the patients were inconclusive due to a lack of Reed–Sternberg cells and atypical mononuclear cells in the specimens. In the lungs of both patients there were histological signs of benign lymphoproliferative lesions accompanying PPHD-follicular hyperplasia in the first case and interstitial lymphocytic pneumonitis in the second. A hyperactivity of the lymph tissue in the lungs might be suspected, followed by development of Hodgkin’s lymphoma. Bilateral pneumonia had lead to the fatal outcome of the second patient. Histologically the differential diagnosis of both cases included infectious and non-infectuous granulomatous disease and non-Hodgkin’s lymphoma.


References

Table 2: Ann Arbor Staging Classification of Hodgkin’s Lymphoma. Details of the lymph nodal regions are depicted in Figure 3. Laterality is utilized in defining the stage when the following regions are involved and should be denoted with “right” or “left” categorization: Cervical, supraclavicular, occipital and pre-auricular. Infraclavicular. Axillary and pectoral. Epitrochlear and brachial. Hilar.