Diffuse primary plasmacytoma of the lung

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Abstract—Primary plasmacytoma of the lung is a rare tumor, thus presenting a diagnostic challenge to the clinician. So far, approximately 20 cases have been verified by immunohistochemistry. We describe an elderly patient presenting with progressive dyspnea on exertion, dry cough, weight loss and malaise. The main finding on plain chest radiography was a diffuse infiltration of pulmonary parenchyma in the lower parts of both lungs and in the middle part of the right lung. The histology of the open lung biopsy of the right middle lobe revealed massive and diffuse infiltration by well differentiated plasma cells with extracellular deposits of amyloid. The plasma cells and amyloid expressed a monoclonal lambda light chain. No monoclonal spike was shown by serum and urine immunoelectrophoresis. A skeletal survey and bone marrow biopsy specimen excluded a disseminated disease and a diagnosis of extramedullary plasmacytoma was made. The patient was considered for VI courses of VMCP chemotherapy after which a complete regression on chest roentgenography was evident. Almost five years after the diagnosis the patient is still alive without any evidence of disease recurrence or dissemination.

Key words: Primary pulmonary plasmacytoma; diffuse; chest X-ray; chemotherapy.

INTRODUCTION

Extramedullary plasmacytomas are plasma cell tumors usually localised in the head and neck region, paranasal cavities and upper respiratory tract [1]. Primary plasmacytoma of the lung (PPL) is a relatively rare tumour [2]. The first case was reported by Gordon and Walker in 1944. So far, more than twenty cases have been reported. Radiotherapy and chemotherapy could affect the natural course of the tumor by slowing the progression into myeloma or even eradicate the disease [3–5]. We report a patient with diffuse infiltration of lung with tumor tissue, contrary to

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most cases where pulmonary plasmacytoma arise as nodular and distinctly separated from the surrounding tissue [6].

CASE REPORT

A 65-year old male, with smoking habits, presented at our clinic in December 1994 with a 4-month history of progressive malaise, weight loss and dry cough. He had been hospitalized twice before, in June and August 1994, for bilateral bronchopneumonia. On admission, he was afebrile, eupnoic, with no hepatosplenomegaly, adenopathy, clubbing or bleeding. There was no bony tenderness either. An auscultatory finding showed wheezing and bilateral crepitations more to the right, in the lower third of the lungs. A clear heart murmur (2/6) was heard above the apex. Chest X-rays revealed marked reticulonodular opacities in the lower parts of both lungs and in the middle part of the right lung (Fig. 1). Initial fiberoptic bronchoscopy was nondiagnostic. Open surgical lung biopsy of the right lung was performed revealing on histology a massive and diffuse infiltration by monomorphic plasma cells which had partially destroyed pulmonary parenchyma (Fig. 2). The plasma cells were rather mature looking with rare mitotic figures and minimal stroma. Rare epithelial granulomas were observed in the tumor tissue. Amorphous eosinophilic deposits, stained positively with Congo red and with morphological characteristics of amyloid were found around thickened blood vessel walls. The immunoperoxidase staining method (alkaline phosphatase, LSAB+, Dakopaths, Denmark) detected in both tumor cells and amyloid the presence of solely lambda light chains of immunoglobulins. Once the diagnosis of extramedullary plasmacytoma had been established histologically, the appropriate staging for multiple myeloma was performed. Except for slight, normocytic normochromic anemia (Hb 11.7 g/dl), the blood count was normal. Sedimentation rate was 68 mm/h
BUN, creatinine, uric acid, transaminases, alkaline phosphatase, LDH and electrolytes were normal. Serum protein electrophoresis showed polyclonal increase of immunoglobulins-30 g/l IgG (normal: 3.5 g/l–18.5 g/l). No monoclonal spike was detected by serum and urine immunoelectrophoresis. Bone marrow aspirate showed a normal differential count with 1.5% mature plasma cells. Immunophenotyping of bone marrow mononuclear cells by EPICS detected no proliferative processes within the B-lymphocyte lineage. Skeletal survey, including plain roentgenograms and CT, failed to demonstrate osteolytic lesions in all portions of the axial skeleton and proximal long bones. Brain and liver-spleen scans were all normal. The biopsy of rectal mucosa for amyloid was negative. According to this survey, the diagnosis of PPL in IE stage was assessed. The patient was treated with vincristine, melphalan, cyclophosphamide and prednisone (VMCP). After six cycles of chemotherapy, a complete regression of radiographic changes in the lungs was evident. The patient is alive and feels well without any evidence of disease progression and dissemination, five years after diagnosis.