A Rare Case of Extramedullary Plasmacytoma in the Mediastinum

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A rare case of extramedullary plasmacytoma in the mediastinum is reported. An 80-year-old man was admitted for further examination of a mediastinal tumor. Chest computed tomography (CT) revealed a large mediastinal mass and right interlobar pleural effusion. Needle biopsy under CT guidance established a diagnosis of plasmacytoma. Immunohistochemical staining revealed that the tumor cells were producing monoclonal IgA kappa. Serum immunoelectrophoresis revealed an IgA kappa monoclonal component with a serum concentration of 5,040 mg/ml. The bone marrow aspiration was normal. Bone roentgenogram and bone scintigram showed osteoporosis but no neoplastic lesion.

Key words: mediastinal tumor, myeloma, plasma cell

Introduction

Multiple myeloma is a disseminated malignant disease in which a clone of transformed plasma cells proliferates in the bone marrow, disrupting its normal functions and invading the adjacent bone. Localized plasmacytoma show monoclonal gammopathy, and can occur as a solitary plasmacytoma of bone or as an extramedullary plasmacytoma (1). Neoplastic change in plasma cells results in the following five clinical manifestations: 1) multiple myeloma, 2) solitary plasmacytoma of bone, 3) extramedullary plasmacytoma, 4) plasma cell leukemia, and 5) premyeloma (2).

Extramedullary plasmacytoma is uncommon, and may affect various organs. It is most commonly seen in the upper respiratory tract, and occurrence in the mediastinum is very rare (1, 3, 4). We report a case of extramedullary plasmacytoma in the mediastinum with interlobar pleural effusion.

Case Report

An 80-year-old man was admitted to our hospital for further examination of a mediastinal mass. After falling from his bicycle on January 25, 1990, he complained of chest pain; chest roentgenogram revealed a large mediastinal mass and right pleural effusion without any bone fracture (Fig. 1). At admission, there was no dyspnea nor palpitation, and the chest pain had already disappeared. His past medical history was unremarkable, except for mild hypertension. He was an ex-smoker with a history of 20 cigarettes per day for 20 years. No family history of neoplasmas was noted. A physical examination showed no goiter, lymphadenopathy or hepatosplenomegaly.

Laboratory investigation revealed a hemoglobin level of 11.2 g/dl, and a white blood cell count of 6,600/mm³ with 27% neutrophils, 57% lymphocytes and 11% eosinophils. The erythrocyte sedimentation rate was 138 mm/h. Serum protein electrophoresis showed a monoclonal gammopathy. Immunoelectrophoresis of the serum revealed an IgA kappa monoclonal component without Bence-Jones protein. Serum concentration of IgG was 863 mg/ml; IgM, 58.5 mg/ml; and IgA, 5,040 mg/ml. Bone marrow aspiration from the sternum showed 2.4% plasma cells. No abnormal cells were detected in the clot specimen. Bone roentgenogram showed osteoporotic change in vertebrae. Chest CT revealed a large mediastinal tumor and right interlobar pleural effusion (Fig. 2). Needle biopsy was performed under CT guidance. Histopathological examination revealed proliferation of atypical plasma cells, and...
Plasmacytoma in Mediastinum

neoplasms are multiple myelomas, and less than 10% are localized plasmacytomas (6). Males are affected more frequently than females with a 1.7–12.1 ratio (1, 4, 7). Reports in the Japanese literature of patients with extramedullary plasmacytoma revealed an average age of 5 to 80, with 63% of the patients presenting between the 4th and 6th decades (4). Extramedullary plasmacytoma arises in soft tissue, generally in the upper respiratory tract; occurrence in the mediastinum is very rare. Mori et al reported that of reported cases in Japan 37% of extramedullary plasmacytoma originate in the oral cavity and upper respiratory tract, 13% in lymph nodes, 10% in the stomach, and 5% in the thyroid gland (4). Fifty-four cases of intrathoracic plasmacytomas were reported by Herskovic et al, but of these only seven cases occurred in the mediastinum, and there was no case with pleural effusion (6).

The present case was unique because of its large size, increased paraprotein, and interlobar pleural effusion (8). The cause of interlobar pleural effusion is not clear, but a blockage of normal pulmonary lymphatic flow by tumor invasion might be responsible (8, 9). The increase in paraprotein corresponds to the size of the plasmacytoma. As the tumor size was about 3 x 10 x 10 cm in the present case, the paraprotein increased, without any dissemination. After two years, there was no change in the size of the tumor, and only a small increase in the paraprotein. These findings suggested our case was not clinically malignant.

The treatment for solitary plasmacytoma is determined by the spread of the disease. Localized solitary plasmacytoma is treated by surgery and/or radiation therapy; 98% are sensitive to radiation (1, 7). As described above, the mean survival time of patients with extramedullary plasmacytoma is much longer than in those with multiple myeloma. Fifty percent of patients with multiple myeloma die within 2 years of diagnosis (1). The present patient was considered not to have disseminated disease, as follow-up CT and scintigram revealed no other involvement. And the patient showed no symptoms of disease for over 2 years, despite his refusal of any treatment.

References