

IgG Myeloma Presenting as a Large Mediastinal Mass and Pleural Effusion

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Summary: Plasmacytoma, a rare form of plasma cell dyscrasia, refers to a neoplastic proliferation of plasma cells. Although the upper aerodigestive tract is the most common site for extramedullary plasmacytoma, mediastinal and pleural involvement is extremely rare. We report the case of a patient who presented with a large mediastinal mass and pleural effusion. Diagnosis was challenging and subsequently made as mediastinal extramedullary plasmacytoma with simultaneous myelomatous pleural effusion. Further investigations revealed multiple myeloma.

Key Words: plasmacytoma, extramedullary, multiple myeloma, mediastinal

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CASE REPORT

A 70-year-old man was referred to our Pulmonary Clinic for evaluation of a mediastinal mass. Two weeks before his visit, he was hospitalized for chest pain. A thorough evaluation revealed no cardiac cause for the chest pain. Computed tomography scan of the chest at the time demonstrated a moderate right-sided pleural effusion, a large mediastinal mass involving the right suprahilar, hilar, and subcarinal areas, with significant mass effect to the esophagus, and numerous pleural-based nodules throughout the right lung, the largest measuring 3.7 cm (Fig. 1). On further review, a chest x-ray from 7 years earlier showed a mediastinal mass. Drainage of right-sided pleural effusion and an endoscopic ultrasound with multiple fine-needle aspiration (FNA) passes were performed and were nondiagnostic.

The patient's history was significant for hypertension, gastroesophageal reflux disease, chronic ob-

structive pulmonary disease, and peripheral vascular disease with stent placement and lumbar spine surgery. He had a history of 50 pack year of smoking cigarettes and drinking a quart of hard liquor every other day for >40 years. He had retired 2 years earlier, after working in a machine shop making farm equipment for 20 years. There was no family history of pulmonary diseases and no history of exposure to asbestos or tuberculosis infection. His physical examination was unremarkable. Routine laboratory work showed mild anemia and stage I chronic kidney disease. Complete blood count, angiotensin-converting enzyme level, lactate dehydrogenase, and metabolic panel were normal. Pleural fluid and FNA sample cultures showed no growth.

Although video-assisted thoracoscopic surgery (VATS) or mediastinoscopy were offered as definitive diagnostic procedures, the patient elected to first undergo bronchoscopy with transbronchial needle aspiration, which also did not reveal malignancy, sarcoidosis, and infections. Subsequently, the patient was taken to the operating room where mediastinoscopy was performed after biopsy specimens collected by bronchoscopy with endobronchial ultrasound were found nondiagnostic. Tissue examination showed many plasma cells with positive CD138, CD38, and CD56 on flow cytometry. Serum immunoglobulin (Ig)G level was elevated with lambda spike on serum protein electrophoresis. Cytogenetic and fluorescence in situ hybridization analyses showed 13q deletion. These findings were consistent with plasmacytoma. A second thoracentesis revealed plasmacytoid cells with similar characteristics confirming the diagnosis of simultaneous myelomatous pleural effusion. Further investigations showed elevated serum β 2 microglobulin level and a normal bone survey. Bone marrow biopsy was found to be normocellular with 6% plasma cells with the same cellular markers as in mediastinal mass, confirming the diagnosis of multiple myeloma. The patient was started on chemotherapy with velcade, melphalan, and prednisone.

DISCUSSION

Extramedullary plasmacytoma (EMP), a rare form of plasma cell neoplasm, constitutes

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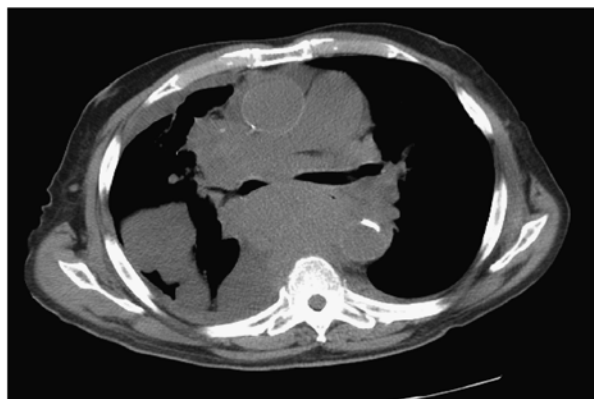


FIGURE 1. Computed tomography scan of the chest revealing a large right hilar mass extending into the subcarinal area and right-sided pleural effusion.

only 4% of all plasma cell tumors.¹ The most common site for extramedullary involvement is the upper aerodigestive tract.² Only 5% of patients with EMP have coexistent multiple myeloma.³ According to Kintzer et al,⁴ in a study of 958 cases of multiple myeloma, intrathoracic EMP was seen in only 11 patients (1%). Although extremely rare, various patterns of thoracic involvement have been reported, including lung mass, multiple pulmonary nodules, diffuse reticulonodular infiltration, lymph node enlargement, mediastinal mass, nodular pleural thickening, and pleural effusion.^{5–8} Concurrent myelomatous pleural effusion and mediastinal EMP as the initial presentation of multiple myeloma, as in our case, is extremely rare. Less than 1% of cases present in this manner, and nearly all are associated with IgA myelomas. To our knowledge, there is only 1 other similar case of pleural and mediastinal involvement of EMP with IgG gammopathy.⁹

Pleural involvement of plasma cells can be associated with adjacent bone, chest wall invasion, and direct implantation of plasma cells into the pleural space.¹⁰ In our case, pleural involvement was likely because of direct invasion of the mediastinal pleura with implantation of plasma cells, as the bone survey and the computed tomography chest did not show bony or chest wall involvement. Other proposed mechanisms of pleural effusion in patients with multiple myeloma include a variety of causes such as infectious complications, nephrotic syndrome, pulmonary embolism, congestive heart failure secondary to amyloidosis, and obstruction of

lymphatic drainage by mechanical effect of mediastinal mass.

Our case showed another rare intrathoracic manifestation of multiple myeloma, that is, mediastinal mass, pleural nodules, and pleural effusion. Because thoracic involvement of multiple myeloma is very rare, it is usually not included in the initial differential diagnosis of patients with mediastinal mass and pleural effusion. Although diagnosis of lung cancer presenting as mediastinal mass (small cell or nonsmall cell) can typically be made easily with endoscopic sampling (FNA through endoscopic ultrasound or endobronchial ultrasound), diagnosis of EMP can be challenging and may require a more invasive approach such as VATS or mediastinoscopy for larger tissue sampling.

It is important to consider thoracic involvement of multiple myeloma in the initial differential diagnosis of patients with a large mediastinal mass. Simple tests such as serum and urine electrophoresis and for serum β_2 microglobulin may give diagnostic clues, and an early aggressive diagnostic approach such as VATS or mediastinoscopy for a larger tissue sampling may make diagnosis easier and faster. Another key to the evaluation is differentiation of EMP with and without coexistent bone marrow involvement (multiple myeloma), as treatment options are different. Although EMP is highly radiosensitive, and surgery can be considered for localized tumors such as those in the gastrointestinal tract, coexistence of multiple myeloma requires treatment with systemic chemotherapy and has poorer prognosis.^{11,12}

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