

A Case of Mediastinal Extramedullary Plasmacytoma Associated with Multiple Myeloma¹

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Extramedullary plasmacytoma is a rare manifestation of multiple myeloma, and involvement of the mediastinum by extramedullary plasmacytoma is very rare. We report here on a rare case of a large mediastinal extramedullary plasmacytoma and several pleural nodules with pleural effusions in a 45-year-old male patient with multiple myeloma that involved the thoracic spine and the calvarium. The mediastinal extramedullary plasmacytoma manifested on CT as an 11 × 4.5 cm-sized, relatively homogeneous, mildly enhancing, anterior mediastinal mass with several pleural nodules, and this simulated malignant lymphoma or malignant thymic epithelial tumor.

Index words : Multiple Myeloma

Plasmacytoma

Mediastinum

Pleura

Tomography, X-Ray Computed

Extramedullary plasmacytoma (EMP) refers to a neoplastic proliferation of plasma cells arising from non-osseous sites, while the neoplastic plasma cells in multiple myeloma patients usually involve the medullary space of bones. Extramedullary plasmacytoma may be primary or secondary to marrow involvement by myeloma. Extramedullary plasmacytoma arising from the mediastinum is known to be extremely rare (1). Herein, we present a rare case of mediastinal EMP that manifested

as a large anterior mediastinal mass associated with several pleural nodules and bilateral pleural effusions in a patient with multiple myeloma that involved the thoracic spine and the calvarium.

Case Report

A 45-year-old man was referred to our hospital for the evaluation of his severe back pain that he'd suffered with for one week. The spine magnetic resonance (MR) scan (Genesis Signa, GE Healthcare) revealed a soft tissue mass involving the T12 vertebral body (Fig. 1A). The vertebral mass showed homogeneous signal intensity similar to that of muscle on both the T1- and T2-weighted images. Mild heterogeneous enhancement of the mass was noted after intravenous gadolinium administration. The expansile mass extended into the central canal and it compressed the spinal cord without definite cortical destruction.

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At that time, a routine chest radiograph revealed bilateral mediastinal widening that was suggestive of a large mediastinal mass (Fig. 1B). The subsequent computed tomographic (CT) scan (LightSpeed VCT, GE Healthcare) revealed an 11 × 4.5 cm-sized, anterior mediastinal mass that showed homogeneous iso-attenuation compared with that of muscle. The mass showed relatively homogeneous enhancement following intravenous contrast administration (Fig. 1C). Additionally, several pleural nodules and small-sized bilateral pleural effusions were also noted. The provisional differential diagnoses included malignant lymphoma, malignant thymic epithelial tumor with pleural seeding and small cell carcinoma with spinal metastasis. The patient underwent CT-guided, percutaneous needle aspiration biopsies of the anterior mediastinal mass and the vertebral mass. Histopathologically, both of them proved to be plasma cell myelomas (Fig. 1D). The results of immunohistochemical staining were strongly positive for lambda light chains, but not for kappa light chains.

After confirming the diagnosis, the patient received a series of related evaluations for multiple myeloma, including serum and urine protein electrophoresis, a bone survey and a bone marrow biopsy. Protein electrophoresis and immunofixation of the serum showed 2.0 g/dL of lambda light chain monoclonal protein. There was 10,240 mg of protein in his urine collected during a 24-hour period. Electrophoresis of the urine protein showed a monoclonal band in the beta-globulin region. The monoclonal protein concentration in our patient was 80.1%.

A bone marrow biopsy showed marrow involvement by neoplastic plasma cells (this comprised 80% of the total cellularity) with decreased trilineage hematopoietic cells, as is generally seen in patients with multiple myeloma. Immunohistochemical staining for anti-lambda antibody demonstrated monoclonality. A survey for skeletal lesions was also performed. The skull radiograph revealed two localized osteolytic areas in the high frontoparietal bone and the left parasagittal parietal bone, respectively. On brain MR, two calvarial plasmacytomas with intra- and extracranial extensions were found, and these plasmacytomas showed homogeneous iso-signal intensity on the T1-weighted images and slightly high signal intensity on the T2-weighted images. The lesions showed slightly heterogeneous enhancement after intravenous gadolinium administration (Fig. 1E).

Because the patient presented with severe back pain,

he received emergency local external beam radiotherapy to the spinal lesion (21 Gy in seven fractions) and this was followed by dexamethasone medication. He is now receiving chemotherapy with cyclophosphamide and dexamethasone for systemic treatment.

Discussion

Multiple myeloma is a neoplastic disorder that's caused by the proliferation of transformed B lymphoid progenitor cells that give rise to a clone of malignant immunoglobulin-secreting plasma cells (2). Multiple myeloma usually manifests as a diffuse bony disease (myelomatosis), but it can sometimes present as a solitary plasmacytoma of a bone or as extramedullary (extraosseous) plasmacytomas (3). According to Kintzer *et al.* (1) in a study of 958 cases of multiple myeloma with thoracic involvement, intrathoracic EMP was seen in only eleven of all the patients (1%). Furthermore, the EMP arising from the mediastinum is extremely rare. Although rare, various patterns of thoracic involvement of multiple myeloma have been reported, including a lung mass, multiple pulmonary nodules, diffuse reticulonodular infiltration, lymph node enlargement, a mediastinal mass, nodular pleural thickening and pleural effusion, and tracheobronchial infiltration (4). To the best of our knowledge, only a few cases of mediastinal plasmacytoma have been reported in the medical literature (5, 6).

The reported radiologic findings of EMP are generally well-defined soft-tissue masses on CT, and these masses are isointense to muscle and white matter on T1-weighted MR images and they are iso- to hyperintense to muscle and white matter on the T2-weighted MR images, with heterogeneous enhancement (7).

Our case showed another rare intrathoracic manifestation of multiple myeloma, that is, pleural nodules with pleural effusions. Pleural effusions are occasional findings in patient with multiple myeloma, and they occur in approximately 6% of patients (1, 8). The proposed mechanisms of pleural effusion in patients with multiple myeloma include a variety of causes such as infectious complications, nephrotic syndrome, pulmonary embolism and congestive heart failure secondary to amyloidosis (8). Although rare, pleural involvement with myeloma cells from the adjacent bone, a pulmonary plasmacytoma, and direct implantation by myeloma cells can be other causes (9). In addition, lymphatic drainage obstruction by a mediastinal mass or

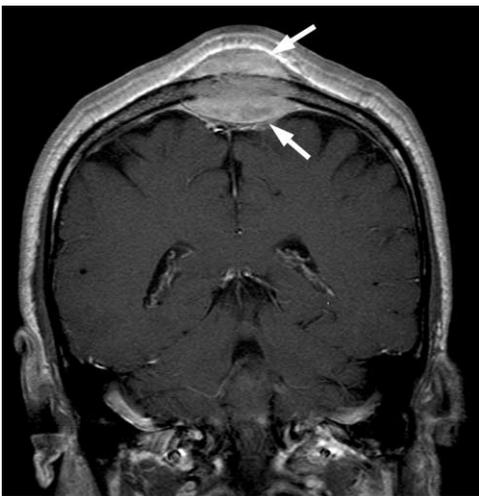
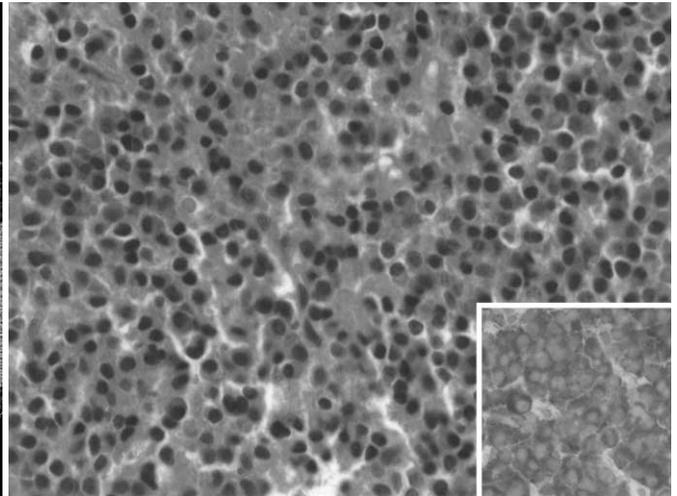
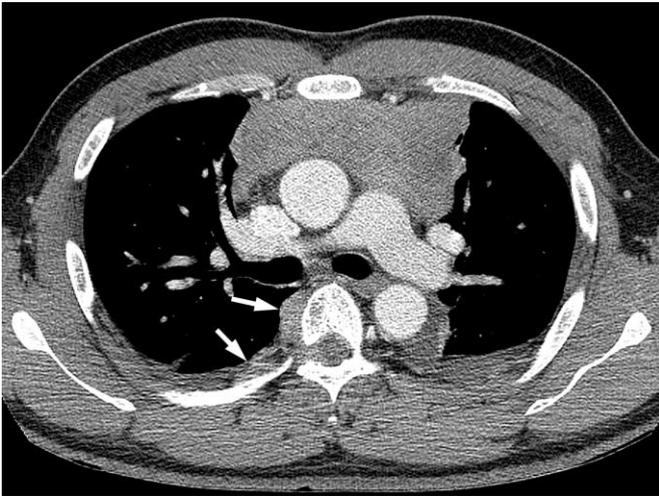
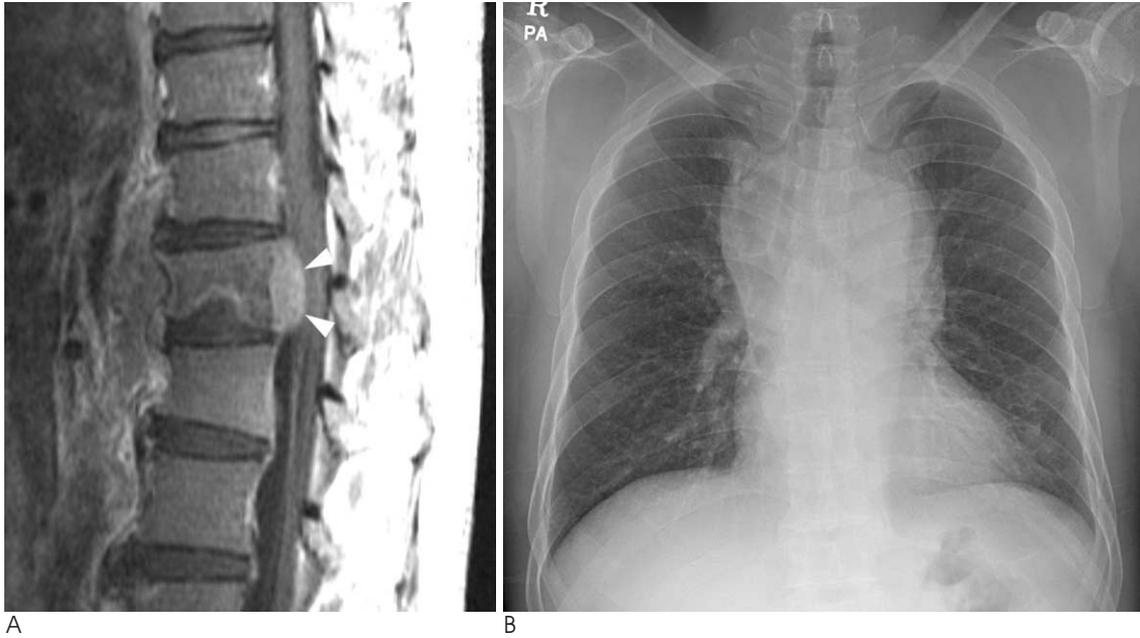


Fig. 1. A 45-year-old male patient with multiple myeloma associated with an extramedullary plasmacytoma in the anterior mediastinum.
 A. Spinal MRI shows a soft tissue mass (arrowheads) involving the T12 vertebral body on the gadolinium-enhanced T1-weighted sagittal image. Note the compression of the adjacent spinal cord by the epidural mass.
 B. The posteroanterior chest radiograph shows bilateral mediastinal widening, which is suggestive of a large mediastinal mass.
 C. The postcontrast CT scan reveals an 11 × 4.5 cm-sized, anterior mediastinal mass that shows mild contrast enhancement. Additionally, several small pleural nodules (arrows) with small bilateral pleural effusions are also noted.
 D. Photomicrograph of the biopsy specimen obtained from the anterior mediastinal mass shows neoplastic myeloma cells, and this represents an extramedullary plasmacytoma. (H & E stain, ×200) (Inset: The tumor cells are strongly positive for lambda light chains on immunohistochemical staining).
 E. The gadolinium-enhanced T1-weighted coronal image of brain MRI reveals a slightly heterogeneous, enhancing calvarial plasmacytoma with intra- and extracranial extensions (arrows).

lymphadenopathy may result in pleural effusions (1). Although the cause of pleural effusion was not clear in our case, lymphatic drainage obstruction by the mediastinal mass might have been responsible.

In the differential diagnosis of an anterior mediastinal mass with several pleural nodules and multiple osteolytic lesions in the axial skeleton, a possibility of EMP associated with multiple myeloma should be considered in addition to malignant lymphoma, malignant thymic epithelial tumor and small cell lung cancer.

In summary, we report here on a case of a large mediastinal extramedullary plasmacytoma and several pleural nodules with pleural effusions in a patient with multiple myeloma that involved the thoracic spine and the calvarium.

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다발성 골수종에 병발된 전종격 골수의형질세포종의 증례보고¹

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골수의형질세포종은 다발성골수종의 드문 형태로서, 특히 흉부 종격의 병변 발생은 매우 드물다. 이 증례보고에서는 12번 흉추와 두개골에 종양을 형성한 45세 다발성골수종 남자에서 큰 전종격종괴와 몇 개의 흉막결절 및 흉막삼출로 발현된 골수의형질세포종의 영상의학적 소견을 기술하고자 한다. CT 상 전종격종괴는 11 × 4.5 cm 크기로서 비교적 균질하고 경도의 조영증강을 보였고, 몇 개의 흉막결절과 흉막삼출도 동반되어 있었다. 이러한 소견들은 림프종이나 악성 흉선종과 유사하므로 감별이 필요하였다.