Bilateral Pleural Masses and Shortness of Breath Associated With Multiple Myeloma*

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A 62-year-old man was admitted to the hospital with progressive shortness of breath and severe back and left flank pain. A diagnosis of multiple myeloma had been made by means of bone marrow biopsy 4 weeks before presentation.

Physical Examination

Vital signs included the following: temperature, 36.4°C; BP, 170/96 mm Hg; pulse, 96 beats per

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Figure 1. Chest radiograph (frontal view) demonstrates bilateral upper pleural opacities (arrows) and a left pleural effusion (open arrow).

Figure 2. CT scan reveals extensive paravertebral and pleural mass with extension into the spinal canal (solid arrows), paraspinal soft tissues (open arrow), and a left pleural effusion.

Figure 3. CT scan at level of right lower lobe pulmonary artery shows intraluminal filling defect (arrow).
minute; respirations, 20 breaths per minute. There was no tenderness on palpation of the left paraspinal and flank regions. The lungs were clear.

*Laboratory Studies*

Values were as follows: hematocrit, 33% (was 42.6% 3 years previously); WBC count, 6,400/mm³ with normal differential cell count; platelet count, 169,000/mm³; sedimentation rate, 127 mm/h; total protein, 11.6 g/dL. Oxygen saturation level was 98% (with the patient breathing room air). Skeletal survey showed a compression fracture of T12 and osteolytic skull lesions consistent with multiple myeloma. A chest radiograph is shown in Figure 1.

*Hospital Course*

The patient was treated with intravenous morphine for pain relief. On the 2nd day of hospitalization, a CT scan of the chest revealed multiple bilateral pleural and chest wall soft-tissue masses. At the T11 and T12 levels, a pleural mass crosses the midline into the paravertebral and intraspinal space and extends both inferiorly into the retrocrural space and posteriorly through the intercostal space into the left paraspinal musculature (Fig 2). In addition, a scan at the level of the right descending pulmonary artery (Fig 3) reveals an intraluminal filling defect. The ribs appeared normal, and no lymphadenopathy was identified. A transthoracic needle biopsy of the left paraspinal component of the mass was performed.

What is the cause of the pleural masses and of the patient’s dyspnea?
Diagnosis: Extramedullary plasmacytomas; incidental pulmonary embolism.

Multiple myeloma is a neoplastic disorder caused by the proliferation of transformed B lymphoid progenitor cells that give rise to a clone of malignant immunoglobulin-secreting plasma cells, as noted by Kyle. Multiple myeloma may manifest as diffuse bone disease (myelomatosis), as solitary plasmacytoma of bone, or as extramedullary (extraosseous) plasmacytomas (EMPs), as discussed by Kapadia. EMPs are rare and may be primary or secondary to marrow involvement by myeloma. While approximately two thirds of patients with multiple myeloma have extraosseous involvement, typically involving the nasal cavity, paranasal sinuses, and nasopharynx, only 5% of patients with EMP have coexistent multiple myeloma, which was shown by Rao and Venkatesh and by Moulopoulos et al.

EMPs are more commonly associated with an aggressive terminal phase of myeloma or plasma cell leukemia. While patients with multiple myeloma have an average age of 60, those diagnosed with the aggressive form of disease tend to be younger, with a mean age of 50 years. Whereas the most common thoracic disorders associated with myeloma are bone involvement or pulmonary infiltrates secondary to a complicating infectious process, primary thoracic involvement by myeloma occurs in less than 1% of cases (studies by Shin et al and Kintzer et al). Reported patterns of EMP of the thorax include lung masses (investigations by Nonomura et al and Joseph et al), multiple pulmonary nodules (studies by Moulopoulos et al), diffuse reticulonodular infiltration by myeloma cells with amyloid deposition (studies by Shin et al), lymphadenopathy and mediastinal mass (evidenced by Kyle and by Moulopoulos et al), pleural effusion and nodular pleural thickening (manifested by Moulopoulos et al), and tracheobronchial infiltration (seen in the investigation by Shin et al).

The patient illustrated developed another indirect thoracic manifestation of multiple myeloma, that of pulmonary thromboembolism. Thrombosis is recognized as the most frequent complication of malignant disease. Proposed mechanisms of thromboembolic disease in patients with malignancy include a hypercoagulable state induced by tumor cells and their products, prolonged recumbency after cancer surgery, and thrombosis related to chemotherapy or hormonal therapy (seen in studies by Donati). Patients with multiple myeloma often have additional risk factors of prolonged bed rest from back pain (due to thoracic vertebral compression fractures) and hyperviscosity associated with elevated serum protein concentration, and the presence of amyloidosis may also be important in the pathogenesis of this complication. An estimate of the incidence of thromboembolism based on a myelomatosis trial is about 3%, while pulmonary embolism accounted for about 3% of all deaths, shown by Catovsky and colleagues. Our patient had received radiation therapy for intractable pain related to a T12 compression fracture secondary to multiple myeloma and had been at bed rest until the time of admission.

The detection of unsuspected pulmonary embolism on a contrast-enhanced CT scan occurs in 0.5% of patients. A recent retrospective study found that although the majority of patients with incidental emboli have risk factors for thromboembolic disease, only 23% of patients were scanned primarily for this indication, as discussed by Verschakelen and coworkers. It is expected that widespread use of a spiral CT scan, which provides superior rendering of central (ie, segmental or larger) pulmonary arteries during peak contrast enhancement, will further enhance the diagnosis of incidental emboli.

The development of aggressive (anaplastic, undifferentiated) myeloma with extramedullary disease is associated with rapid progression and resistance to treatment, with a median survival of 1.5 months, shown by Nonomura and others. In a review of 10 cases of pulmonary plasmacytoma in the aforementioned study, half of the patients developed generalized myelomatosis within 16 months and died within 4 years. Early diagnosis of EMP and institution of systemic therapy may prevent further progression of the disease.

The diagnosis of extramedullary thoracic plasmacytoma is particularly difficult when there is no thoracic vertebral or rib involvement. The radiologic appearance is nonspecific, with findings on a CT scan or MRI mimicking those of primary or metastatic carcinoma, sarcoma, neuroendocrine or neuroectodermal tumor, and lymphoma, as discussed by Nonomura et al. Pulmonary plasmacytoma often yields negative cytologic study results on sputum samples and bronchoscopic examination results are also normal. Transthoracic needle biopsy or thoracotomy specimens are frequently necessary for diagnosis. Immunohistochemical staining of biopsy specimens may aid in diagnosis (Nonomura et al evidence this). In this patient, a CT-guided aspiration biopsy of the left paraspinal mass showed abundant atypical plasma cells diagnostic of EMP.

The present patient had extensive myelomatosis as demonstrated by bone marrow biopsy, skeletal survey, and CT scan findings with biopsy specimen confirmation. The spinal involvement was managed; radiation therapy provided improvement in back pain. He opted not to undergo bone marrow transplantation and received chemotherapy, including vincristine, carmustine, cyclophosphamide, and melphalan. The pulmonary embolism was treated with
intravenous heparin and oral coumadin with improvement in pulmonary symptoms and no episodes of recurrent emboli after 3 months of follow-up.

**Clinical Pearls**

1. *In a patient with multiple myeloma and lung, mediastinal, or pleural opacities, EMP should be considered.*

2. A CT scan or MRI is useful in detecting extramedullary disease in patients with multiple myeloma, but the findings are nonspecific and may mimic other primary or metastatic neoplasms.

3. A CT-guided biopsy is a useful technique in the diagnosis of thoracic EMP and can help guide treatment.

4. Pulmonary embolism, appearing as intraluminal filling defects within central pulmonary arteries, can be detected incidentally on contrast-enhanced spiral CT. The pulmonary arteries should be carefully evaluated on every chest CT examination, particularly in patients with malignant neoplasms or other underlying risk factors for thromboembolic disease.

**Suggested Readings**


Donati MB. Cancer and thrombosis. Haemostasis 1994; 24: 128-31


Shin MS, Carcelen MF, Ho KJ. Diverse roentgenographic manifestations of the rare pulmonary involvement in myeloma. Chest 1992; 102:946-48