

## CASE REPORT

# Pulmonary and nodal multiple myeloma mimicking lymphoma

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**ABSTRACT.** Multiple myeloma is a haematological malignancy characterized by the occurrence of plasma cell tumours within the bone marrow. In advanced multiple myeloma, metastatic deposits outside the bone marrow (extramedullary) are rare. Reported extramedullary sites include the upper respiratory tract and lymph nodes. Parenchymal pulmonary plasmacytoma is exceptionally rare. We describe such a case in a 51-year-old male, who underwent allogenic marrow transplantation. 6 months post-transplantation developing nodal and hilar pulmonary multiple myeloma, the radiological appearances mimicking lymphoma.

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Multiple myeloma (MM) is a systemic disease process primarily involving the bone marrow. In MM the bone marrow is replaced by malignant plasmacytes, which produce monoclonal proteins. This disease process primarily involves the axial skeleton.

Extramedullary plasmacytoma (EMP) are tumours of malignant plasmacytes occurring outside the bone marrow. A solitary plasmacytoma is a single focal tumour of malignant plasmacytes that may occur in or outside the bone marrow.

EMP are uncommon and typically manifest as a solitary plasmacytoma. Solitary plasmacytomas occur most commonly in the upper respiratory tract [1] such as nasal cavities, paranasal sinuses and nasopharynx. Approximately 15% of patients with extramedullary plasmacytoma progress to full multiple myeloma [2].

Localized intrapulmonary plasmacytoma is rare, representing fewer than 5% of all extramedullary plasmacytomas [3]. We describe a case of MM with pulmonary and lymph node involvement. To our knowledge the CT findings of extramedullary pulmonary multiple myeloma have not been previously described.

## Case report

A 51-year-old man with a background history of light chain disease MM stage 4b had previously undergone an allogenic sibling bone marrow transplant. Initially, he made a satisfactory clinical recovery. However, a follow-up bone marrow aspirate performed at 2 months post-transplant revealed some atypical and immature plasma cells raising concern for residual disease.

6 months after transplantation, the patient re-presented to the emergency department with increasing cough and shortness of breath. A chest radiograph was performed which demonstrated bilateral hilar lymphadenopathy and

lung nodules. The findings raised concern for a recurrence and a bone marrow aspirate was performed, which revealed a hypocellular marrow with trilineage engraftment and monoclonal plasma cells inkeeping with recurrence.

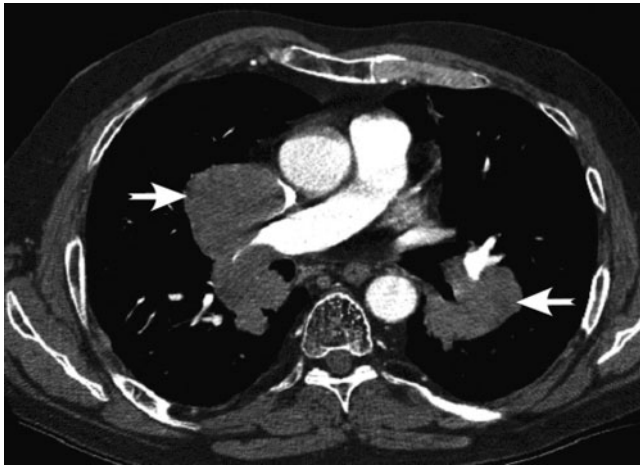
In view of the suspicion of malignancy on the chest radiograph, an intravenous contrast enhanced CT was ordered. This confirmed the presence of extensive bilateral hilar lymphadenopathy (Figure 1a) and multiple lung nodules. The lung nodules were round, smooth or lobulated, and had soft tissue attenuation. They measured from 1.1 cm × 1.0 cm in size (anteroposterior × transverse diameter) in the left upper lobe to 4.9 cm × 4.8 cm diameter in the left lower lobe (Figure 1b).

The initial differential diagnosis, based on the CT findings included lymphoma and post-transplant lymphoproliferative disease. The hospital respiratory and haematology services were consulted, and the patient subsequently underwent a bronchoscopy and transbronchial biopsy. The biopsy samples contained a homogeneous cellular infiltrate with immunophenotypic features of plasma cells, consistent with recurrent multiple myeloma (Figure 2). Despite haematological and intensive care supportive measures, including treatment for opportunistic pathogens, the patient's respiratory function progressively deteriorated over the next 2 weeks. The patient died 16 days after admission of progressive respiratory failure. No post mortem was performed.

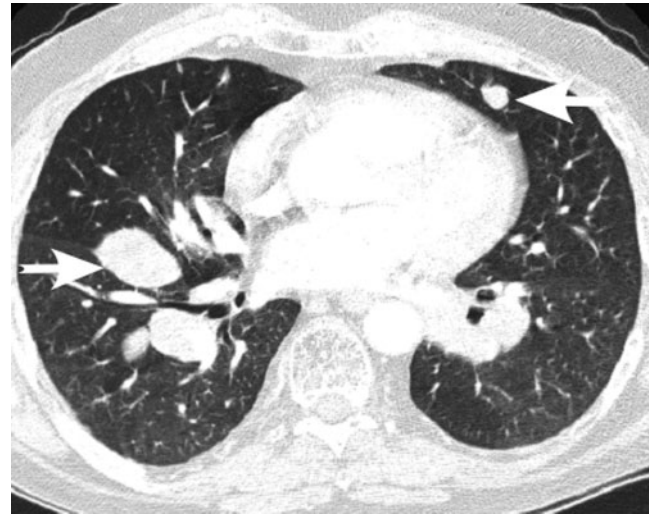
## Discussion

The occurrence of extramedullary disease is uncommon in MM. In one study of a group of 432 patients with MM, only 19 (4.4%) patients were identified as having extramedullary plasmacytoma (EMP) [4]. In the same study, a total of 48 solitary plasmacytomas were identified in 19 patients with extramedullary disease. The most common sites were lymph nodes in 10 (20.8%), the pleura in 8 (16.6%) and soft tissues in 6 (12.5%). Only

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(a)



(b)

**Figure 1.** A 51 year-old man with extramedullary myeloma. (a) Mediastinal windows: CT image shows large bilateral hilar lymph nodes (right arrow) compressing the pulmonary artery and bronchus intermedius. (b) Lung windows: CT image shows bilateral lung nodules and masses (Hounsfield range  $65 \pm 12$ ) (arrows).

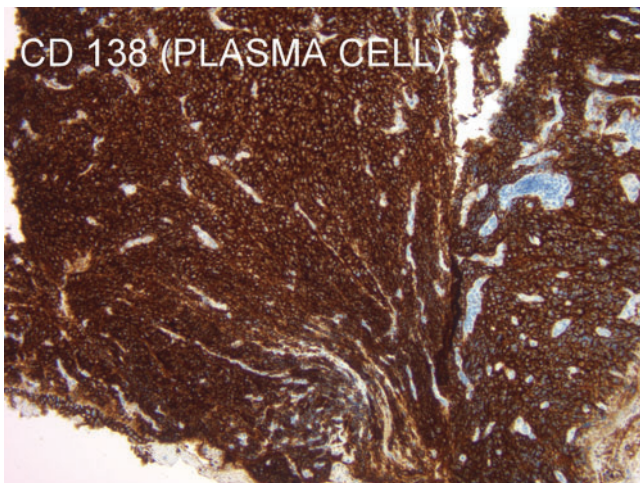
3 (6.2%) lesions occurred within the lung parenchyma. The prognosis of patients with pulmonary multiple myeloma is poor [5]. This contrasts with reports of long survival rates with primary pulmonary plasmacytomas of the lung [6].

The most typical thoracic manifestations of multiple myeloma are bony involvement of the thoracic cage or a pulmonary infiltrate secondary to infection [7]. One author has recently described 13 patients with multiple myeloma and pulmonary complications. Six (46%) patients had pneumonia, 2 (15%) had mass lesions, 2 (15%) had multiple nodular lesions and 3 (23%) had interstitial infiltrates [7]. Other described manifestations of myeloma in the lung include; a diffuse reticulonodular pattern [8] or pulmonary calcification [9]. Amyloid, often seen with multiple myeloma has several manifestations in the lung. Nodular parenchymal disease (multiple well-defined nodules), tracheobronchial (focal or diffuse

thickening of the airway) and diffuse parenchymal (also termed alveolar septal) amyloidosis [10]. The CT findings in our patient of bilateral hilar adenopathy with round pulmonary nodules are unusual for MM, and resemble more those of lymphoma and post-transplant lymphoproliferative disorder (PTLD).

Immunophenotyping is a technique used to differentiate individual cell lineages. This is done by attaching surface antibodies to cell specific surface antigens. Myeloma has been shown to stain positively with CD138 [11]. Both bronchoscopic biopsy samples stained strongly positive for CD 138 plasma cell markers. Also, in PTLD we would expect immunophenotypic features of B cell lymphoma and Epstein Barr virus (EBV) positivity, both of which were absent.

In conclusion, although the CT features and clinical history in this case would strongly support PTLD or lymphoma, the biopsy and immunophenotyping revealed a very unusual manifestation of multiple myeloma.



**Figure 2.** Medium power view ( $20 \times$ ) of bronchoscopy biopsy infiltrate, with CD 138 plasma cell marker, exhibiting a strongly positive brown stain. These features are characteristic of multiple myeloma.

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