

# A Rare Case Presentation: Multiple Myeloma with Extramedullary Plasmacytoma Mimicking Lung Tumor

Elvi Aprillia Karamoy<sup>1</sup>, Yesicha Kurniawati<sup>2</sup>, I Wayan Sunaka<sup>3</sup>, Ni Made Dwita Yaniswari<sup>4</sup>,  
Wayan Wahyu Semara Putra<sup>5</sup>

<sup>1</sup> Intern of Internal Medicine Department in Wangaya Regional General Hospital, Denpasar, Bali, Indonesia

<sup>2</sup> Intern of Pulmonology and Respiratory Medicine Department in Wangaya Regional General Hospital, Denpasar, Bali, Indonesia

<sup>3</sup> Internist of Internal Medicine Department in Wangaya Regional General Hospital, Denpasar, Bali, Indonesia

<sup>4,5</sup> Pulmonologist of Pulmonology and Respiratory Medicine Department in Wangaya Regional General Hospital, Denpasar, Bali, Indonesia

**Abstract:** Multiple myeloma (MM) is a hematological malignancy characterized by clonal proliferation of plasma cells in the bone marrow. Mostly, clonal proliferation of plasma cells is limited to the bone marrow, the occurrence of the extramedullary disease is very uncommon in MM. And approximately 80–90% of extramedullary lesions occur in the head and neck. Only in rare cases, the malignant plasma cells of multiple myeloma had infiltrated the lung parenchyma. Hereby, we present a case of a 53-year-old-male diagnosed with an extramedullary disease of multiple myeloma (EMD). The patient complained of fatigue, nausea vomiting, and bone pain. Chest radiograph discovered opacity in the left lung, transthoracic biopsy showed an extramedullary plasmacytoma, and bone survey discovered multiple lytic lesions.

**Keywords:** multiple myeloma, extramedullary disease, plasmacytoma, lung tumor, CRAB criteria

## 1. Introduction

Multiple myeloma (MM) is a systemic malignant disease of the blood, characterized by uncontrolled growth of monoclonal plasma cells in the bone marrow which affects multiple organ systems.<sup>[1,2,3,4]</sup> It is an incurable and associated with end-organ damage consisting of anemia, renal insufficiency, bone lesions, and/or hypercalcemia (CRAB criteria).<sup>[2,7,8,12]</sup>

Almost all patients with multiple myeloma evolve from an asymptomatic pre-malignant stage termed monoclonal gammopathy of undetermined significance (MGUS) that progresses to smoldering multiple myeloma (SMM), and finally to symptomatic multiple myeloma (MM).<sup>[12,13]</sup> The diagnosis of both MGUS and smoldering myeloma requires that no 'CRAB' features be present.<sup>[7,8]</sup>

Multiple myeloma accounts for 1% of all cancers and the 2<sup>nd</sup> most common hematologic malignancy (approximately 10% of all hematologic malignancies).<sup>[1,2,3,11,14]</sup> It was seen twice as much in men than women.<sup>[5,10,13]</sup> The median age at diagnosis is 65 years and the current 5-year survival is approximately 46.6%.<sup>[10,13,14,15]</sup>

In 2014, the International Myeloma Working Group (IMWG) updated the diagnostic criteria for MM. The diagnosis of multiple myeloma requires 10% or more clonal plasma cells in the bone marrow examination or a biopsy-proven plasmacytoma plus evidence of one or more multiple myeloma defining events (MDE): CRAB (C is elevated calcium, R is renal insufficiency, A is anemia, and B is lytic bone lesions) features felt related to the plasma cell disorder. The laboratory diagnostic parameters including clonal bone

marrow plasma cells are greater than or equal to 60%, serum-free light chain (FLC) ratio greater than or equal to 100 provided involved FLC level is 100 mg/L or higher, or more than one focal lesion on magnetic resonance imaging (MRI).<sup>[8-13]</sup>

Tumor burden in multiple myeloma has traditionally been assessed using the Durie–Salmon Staging (DSS) and the International Staging System (ISS).<sup>[1,7,13]</sup>

Mostly, the clonal proliferation of plasma cells is limited to the bone marrow. However, in some cases, they can migrate/spread outside the bone marrow, which is recognized as an extramedullary disease of multiple myeloma (EMD).<sup>[20]</sup> The occurrence of the extramedullary disease is very uncommon in MM. Only about 7-18% of MM patients with extramedullary disease (EMD) at the time of initial diagnosis.<sup>[13,17,19]</sup> And approximately 80–90% of extramedullary lesions occur in the head and neck.<sup>[16,18]</sup> Only in rare cases, the malignant plasma cells of multiple myeloma had infiltrated the lung parenchyma.<sup>[16,17]</sup>

## 2. Case Report

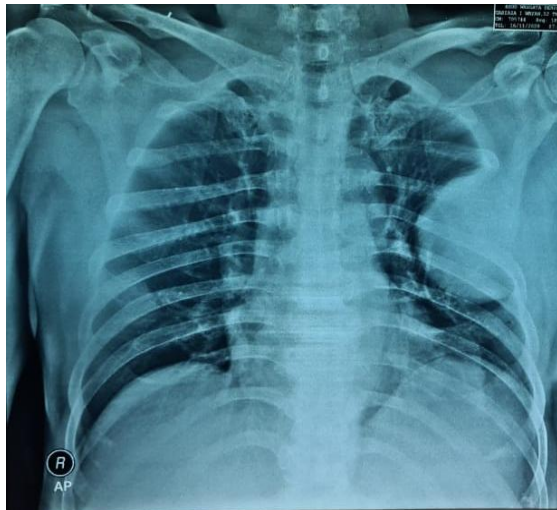
A 53-year-old man came to the Emergency Room (ER) with complaints of nausea and vomiting 2 days prior. The patient also complained of progressing fatigue, general weakness, bone pain, and significant weight loss for a one-year duration. There was no history of cough, dyspnea, breathlessness.

One month prior to the hospital admission, the patient was diagnosed with diabetes mellitus type 2 with 500 mg metformin as a daily medication.

On admission, the patient was conscious with a normal vital signs (blood pressure of 112/72 mmHg, pulse rate 90 bpm, respiratory rate of 18 x/minutes, and axillary temperature of 36,7<sup>0</sup>C).

Anemic conjunctiva was observed during physical examination. Dullness and decreased vesicular sound on the mid-lateral left lung were also noted. Routine hematology examinations revealed low hemoglobin (9,6 g/dL), decreased kidney function (blood urea nitrogen 98 mg/dL and creatinine serum of 6,3 mg/dL), and hypoglycemia (random blood glucose 60 mg/dL). The blood calcium value was 11,0 mg/dL.

Chest radiograph discovered opacity in the left lung [Figure 1]. A chest computed tomographic (CT) without contrast was performed due to decreased renal function and revealed a necrotic heterogeneous solid mass in the left hemithorax with the destruction of the left lateral fourth rib [Figure 2]. Multiple lytic lesions were found at the 4<sup>th</sup> and 5<sup>th</sup> right ribs, 3<sup>rd</sup> left rib, corpus vertebrae thoracolumbar and both scapula.



**Figure 1:** Chest radiograph showed a mass in the left lung or the left pleura.

The bone survey discovered multiple lytic lesions at the skull [Figure 3] and pubic bone right-left [Figure 4].



**Figure 2:** Chest CT scan showed a necrotic heterogeneous mass in the left hemithorax with the destruction of the lateral fourth rib.

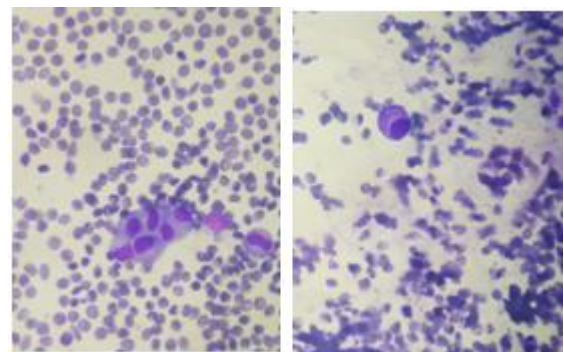


**Figure 3:** The bone survey showed multiple lytic lesions at skull



**Figure 4:** The bone survey showed multiple lytic lesions at pubic bone right-left.

Transthoracic biopsy with chest CT scan guiding confirmed an extramedullary plasmacytoma [Figure 5].



**Figure 5:** Transthoracic biopsy showed an extramedullary plasmacytoma

The patient was admitted to the hospital and treated by internist and pulmonologist with D10% 20 drips per minute, 2 flash of D40% until blood glucose level more than 100 mg/dL, esomeprazole 40 mg injection every 12 hours, ondansetron 4 mg injection every 8 hours, cefoperazone 1 gr injection every 12 hours, morphine-sulfate (MST) 10 mg tablet every 12 hours per oral, the anti-diabetic drug was postponed due to low blood glucose.

After twelve days of hospitalization the patient's condition improved, there were no complaints of nausea vomiting. The patient was referred to the oncology department of the provincial hospital for definitive treatment of multiple myeloma.

### 3. Discussion

We report a unique and rare case of multiple myeloma, with the presence of plasmacytoma in the lung, which is thought to be a lung tumor.

In this case, the patient came with complaints of progressive fatigue, weakness, significant weight loss. The patient was nauseated and vomiting for 2 days prior. He often felt pain all over the body for a year prior.

MM patients often complain of non-specific symptoms, such as fatigue and bone pain. Anemia was present in 73% of patients and this was also associated with fatigue in 32% of patients, bone pain in 58%, weight loss in 25%, osteolytic bone lesions in about 80% of patients, pathological fractures in 30%, hypercalcemia in 15%, elevated serum creatinine level in 20%.<sup>[3,10]</sup>

Chest radiograph discovered opacity (mass) in the left lung which turned out to be a biopsy showing plasmacytoma, and hematology examinations met the signs of CRAB, namely: calcium 11,0 g/dL, serum creatinine 6,3 mg/dL, hemoglobin 9,6 g/dL. The bone survey showed lytic lesions on the head and pubis. This has met the criteria of Multiple myeloma but needs to be confirmed further with a biopsy of bone marrow and urine and blood electrophoresis.

Clonal proliferation of plasma cells is most commonly found in bone marrow, however in some cases they can migrate/spread outside the bone marrow, which is recognized as an extramedullary disease (EMD). The spread of extramedullary plasmacytoma in the lungs is extremely rare. Only about 7-18% of MM patients with the extramedullary disease (EMD) at the time of initial diagnosis.<sup>[13,17,19]</sup> Magnetic resonance imaging (MRI), positron emission tomography (PET)/ computed tomography (CT) imaging may be very useful and should be performed in all patients suspected of extramedullary involvement.<sup>[19,20]</sup>

Supportive treatments are needed in a patient with multiple myeloma. Pain is common in myeloma and often requires opiate analgesia plus NSAIDs. Radiotherapy may help control pain due to localized bony lesions. Hypercalcemia should be managed with fluids and bisphosphonates (pamidronate, zoledronate, or clodronate). Prophylactic antibiotics and antiviral may be used, depending on the treatment.<sup>[1,3]</sup>

There are many active drugs to treat MM in addition to alkylators and corticosteroids. Thalidomide, lenalidomide, and pomalidomide are termed immunomodulatory agents (IMiDs). Bortezomib, carfilzomib, and ixazomib are proteasome inhibitors (PIs). Elotuzumab, daratumumab, and isatuximab are monoclonal antibodies (mAbs). Panobinostat is a deacetylase inhibitor.<sup>[9,10,11,20]</sup>

Initial treatment consists of bortezomib, lenalidomide, dexamethasone (VRD). In eligible patients, initial therapy is given for approximately 3–4 months followed by autologous stem cell transplantation (ASCT).<sup>[6,10,13]</sup> The presence of EMD was associated with poor overall survival.<sup>[19,20]</sup>

### 4. Conclusion

Extramedullary disease (EMD) of multiple myeloma is a rare case. The definitive diagnosis of extramedullary myeloma is difficult if it is seen from the patient's clinical and radiology alone. Further tests are needed to help confirm a diagnosis such as a biopsy, electrophoresis, and the common tetrad of multiple myeloma: CRAB (Calcium elevated, Renal Failure, Anemia, Bone Lesions). EMD was associated with a poor prognosis.

### 5. Author Contribution

All authors contributed equally.

### 6. Conflict of interest

There is no conflict of interest in this case report.

### 7. Acknowledgement

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