ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

https://patsjournal.org

Journal of the Pan African Thoracic Society

Case Report

Multiple myeloma presenting as a lung plasmacytoma: A case report

Filmon Tsegay[1], Ephrem Berhe[1], Hadas Weldezgina[2]

Departments of [1]Internal Medicine and [2]Pathology, Ayder Comprehensive Specialized Hospital, College of Health Sciences, Mekelle University, Mekelle, Ethiopia.

***Corresponding author:**
Ephrem Berhe, M.D,
Head of Nephrology Unit,
Department of Internal Medicine, Ayder Comprehensive Specialized Hospital, College of Health Sciences, Mekelle University, Mekelle, Ethiopia.

eph123ber@gmail.com

Received: 08 August 2022
Accepted: 02 November 2022
EPub Ahead of Print: 19 November 2022
Published: 30 January 2023

DOI
10.25259/JPATS_34_2022

Quick Response Code:

ABSTRACT

Multiple myeloma (MM) is a clonal proliferation of malignant plasma cells mainly affecting the bone marrow. The most common sites of extramedullary dissemination reported in the literature are skin, liver, kidneys, and central nervous system. The presentation of MM with lung plasmacytoma is found to be very uncommon. We report a case of lung plasmacytoma associated with MM. A 65-years-old, non-smoker, female Ethiopian patient presented with 5 months history of dry cough, low grade intermittent fever, fatigue, anemia, hypercalcemia, renal insufficiency and osteolytic lesions in the skull. On a chest X-ray, she had right upper lung opacity with a well-defined margin medially. Fine-needle aspiration cytology from the lung mass lesion confirmed mononucleated and multinucleated plasmacytoid cells. The presentation of MM with lung plasmacytoma is a rare entity. As elderly patients with MM may first present with pneumonia-like presentation, MM or plasmacytoma should be considered as a differential diagnosis in an older patients presenting with thoracic lesion.

Keywords: Multiple myeloma, Lung, Plasmacytoma, Case report, Ethiopia

INTRODUCTION

Multiple myeloma (MM) is a hematologic malignancy characterized by the neoplastic proliferation of a single clone of plasma cells derived from B-cells. It affects mainly the elderly with a median age at diagnosis being 65 years and accounts for about 1% of all types of malignancies and 10% of hematologic malignancies.[1] The typical systemic symptoms of MM are bone pain, malaise, anemia, renal insufficiency, and hypercalcemia. Myeloma cells may be extramedullary located due to extramedullary plasmacytoma or extramedullary dissemination of MM.

At initial presentation, the presence of extramedullary disease is seen in about 7% of patients.[2] To the best of our knowledge, uncommon sites of extramedullary disease are the gastrointestinal and urogenital tracts, central nervous system, lymph nodes, and skin. Association of MM with lung plasmacytoma is found to be extremely rare.[3,4] In a study of 958 cases of MM, six patients presented with an extramedullary plasmacytoma in the lung.[5] Two cases of MM involving the thorax were reported from India.[6] The classic thoracic manifestations of MM are bony involvement of the thoracic cage. However, primary thoracic involvement by myeloma occurs in less than 1% of cases.[7,8] Other manifestations reported were pneumonia, intraparenchymal mass lesions, mediastinal lymphadenopathy, pulmonary nodules, reticulonodular infiltrations, and intrapulmonary calcification.[5-7]

The diagnosis of extramedullary thoracic plasmacytoma is particularly difficult when there is no thoracic vertebral or rib involvement. We present a case of MM with respiratory symptoms as the initial presentation of the disease. Given the rarity of this localization, this report aimed to increase knowledge of this disease among primary care physicians and pulmonologists, to provide a more timely diagnosis.

CASE REPORT

A 65-year-old female, non-smoker with no history of respiratory hospitalization and relatively good past health presented to our hospital in 2021 with a complaint of intermittent dry cough, low-grade intermittent fever, easy fatigability, and loss of appetite of 5 months duration. She also had right lateral pleuritic chest pain associated with profuse night sweating and significant weight loss of 10 kg since the onset of the illness. Her symptom of cough and fever worsened 2 weeks before presentation. Otherwise, there were no other pertinent complaints from other systems.

The physical examination then found a pale patient with a pulse rate of 96/min, respiratory rate of 20/min, blood pressure of 130/80 mmHg, and SpO2 of 90% with atmospheric oxygen. A pulmonary examination was notable for decreased air entry and inspiratory fine crepitation over the right upper lung field. Other physical examinations found no obvious abnormalities. Laboratory blood workup showed a total leukocyte count of 7400 cells/mm^3 with neutrophil of 40%, lymphocyte = 33%, platelets were 385,000 cells/mm^3, hemoglobin of 9.2 g/dL, mean cell volume of 92 fl, mean cell hemoglobin of 32.8 pg, mean cell hemoglobin concentration of 35 g/dL, and erythrocyte sedimentation rate of 88 mm/h. Other laboratory examinations including a kidney function test at the time of presentation showed no obvious abnormalities. Her chest X-ray showed right upper lung opacity with a well-defined margin medially and normal rib cages and clavicles [Figure 1]. She had also a colonoscopy examination for a workup of the anemia and was normal. A sputum microscopic examination was not done as it was a non-productive cough. At this moment, she was admitted and treated empirically for atypical pneumonia with IV antibiotics and discharged with minimal improvement. After 1 month, she presented again to the chest clinic with the same complaint. Hence, ultrasound-guided fine-needle aspiration cytology from the lung parenchymal mass lesion was done. Pathologic examination of this tissue on Giemsa stain smear showed proliferation of plasma cells displaying eccentrically located nuclei with abundant bluish cytoplasmic perinuclear Hof, some of which lost their cytoplasm and appear bare nuclei with a hemorrhagic and proteinaceous background [Figure 2a].

Binucleation, multinucleation, and mitosis were seen on the Giemsa stain. The presence of plasmacytoid cells both

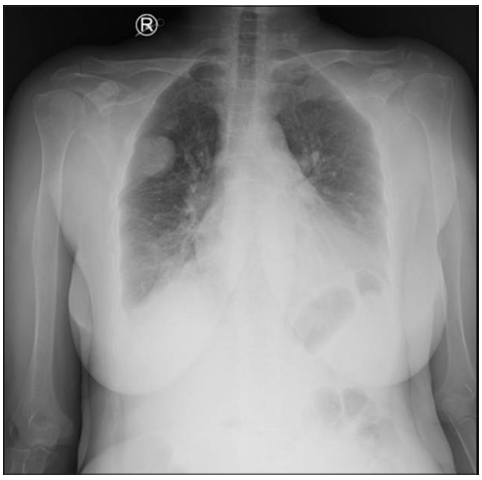

Figure 1: Chest X-ray (PA view) showing right upper lung opacity with well-defined margin medially.

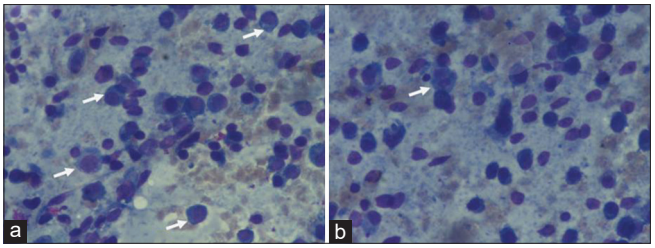

Figure 2: FNAC from a lung mass lesion shows both mononucleate (a) and multinucleate (b) plasmacytoid cells. The white arrows indicate the proliferation of plasma cells displaying eccentrically located nuclei with abundant bluish cytoplasmic perinuclear Hof, some of which lost their cytoplasm and appear bare nuclei with a hemorrhagic and proteinaceous background. Giemsa stain. HP view. FNAC: Fine-needle aspiration cytology, HP: High-power field.

mononucleate and multinucleate per high-power field view with characteristic multiple plasmablasts having abundant cytoplasm and perinuclear clearing was depicted [Figure 2b].

A subsequent X-ray of the skull showed multiple osteolytic lesions [Figure 3]. Workups done days later in the line of MM revealed a total protein of 12.6 g/dL, erythrocyte sedimentation rate increased to 102 mm/h, serum creatinine progressively increased to 2.3 mg/dl, serum total calcium of 15.1g/dL, and serum albumin ranging from 2 to 2.5g/dL.

DISCUSSION

Our patient presented with a lung lesion, progressive fatigue, significant weight loss, anemia, hypercalcemia, renal insufficiency, and osteolytic lesions in the skull, all of which are consistent with the presence of MM. The most common presenting symptoms and signs of MM include bone pain (66%) anemia (73%), fatigue related to anemia (32%), weight loss (12%), elevated serum creatinine (48%), lytic bone

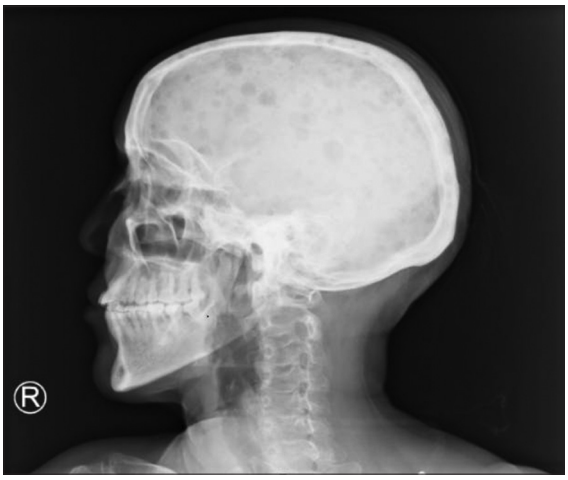

Figure 3: Skull X- ray (lateral view) showing multiple different sizes punched out osteolytic lesions.

lesions (79%), and hypercalcemia (13%).[1] Interestingly, this reported case initially presented with respiratory symptoms and right upper lung zone opacity. Ultrasound-guided cytology from the lung parenchymal lesion revealed mononucleated and multinucleated plasmacytoid cells consistent with plasma cell infiltration. Bone marrow aspirate/biopsy was not done in this patient as the diagnostic criteria for MM in this patient had been already met.[9]

The 2014 International Myeloma Working Group updated criteria for the diagnosis of MM are as follows.[9]

Both criteria must be met

a. Clonal bone marrow plasma cells ≥10% or biopsy-proven bony or extramedullary plasmacytoma
b. Any one or more of the following myeloma defining events:

Evidence of end-organ damage that can be attributed to the underlying plasma cell proliferative disorder, specifically:

i. Hypercalcemia: Serum calcium >1 mg/dL higher than the upper limit of normal or >11 mg/dL
ii. Renal insufficiency: Creatinine clearance <40 mL/min or serum creatinine >2 mg/dL
iii. Anemia: Hemoglobin value of >2 g/dL below the lower limit of normal or a hemoglobin value <10 g/dL
iv. Bone lesions: One or more osteolytic lesions on skeletal radiography, CT, or PET-CT
v. Clonal bone marrow plasma cell percentage ≥60%
vi. Involved: Uninvolved serum FLC ratio ≥100 (involved FLC level must be ≥100 mg/L)
vii. >1 focal lesion on magnetic resonance imaging studies (at least 5 mm in size)

These clinical, laboratory, and pathologic findings of our patient were consistent with the diagnosis of MM with the involvement of the lung parenchyma. Similar to our case, a report by Saha *et al*. reported cases of MM who presented with pneumonia-like respiratory symptoms associated with a mass lesion arising from the apex of the left lung.[6] In another report by Prasad *et al*., a case of MM with plasmacytoma of the lung had similar sign and symptom constellations except the lytic lesions were in the thoracic cage unlike our case, where the lytic lesions were evidenced in the skull.[7]

The diagnosis of extramedullary thoracic plasmacytoma is particularly difficult when there is no thoracic vertebral or rib involvement. Besides, the radiologic findings in the lung parenchyma are non-specific mimicking primary or metastatic carcinoma or lymphoma.[10-12]

We reported a case of MM in which a lung lesion was an initial clinical presentation of the disease. Such presentation is not reported from Ethiopia to the best of our knowledge. Pulmonary parenchyma is an uncommon site of extramedullary involvement in MM; only isolated cases with histological proof have been reported in the literature.

After the diagnosis of plasmacytoma, extensive investigation for MM is crucial as the prognosis of patients with pulmonary dissemination is poor. Extramedullary involvement is associated with aggressive disease, leading to shorter overall survival and progression-free survival.[13] It is difficult to recommend a specific treatment strategy; however, experts recommend that it should be treated as high-risk MM.[14] Treatment of MM depends on patients' age and prognostic factors. Induction and maintenance therapy are two key steps in the management of newly diagnosed MM patients.[15] Regardless of the treatment regimen or initial response to treatment, the disease follows a high relapsing rate in the majority of the patients.

After the diagnosis was established, our patient was put on ferrous sulfate and folic acid supplementation for the anemia and referred for definitive chemotherapeutic treatment to the capital city of Ethiopia, as access to cancer medications is not available in the Tigray region, due to the ongoing war and siege imposed in the northern part of Ethiopia.[16]

CONCLUSION

The presentation of MM in the lung as extramedullary dissemination is a very rare entity. As elderly patients might have a pneumonia-like presentation, it should be entertained in the differential diagnosis in older patients presenting with thoracic lesions. Etiologic identification of pulmonary mass lesions found in patients with MM is necessary to ensure adequate and timely therapy.

Availability of data and materials

All relevant data are within the manuscript file.

Authors' contributions

Filmon Tsegay and Ephrem Berhe reviewed the patient's clinical data and wrote the initial draft of the manuscript. Hadas Weldezgina reviewed the clinical data and interpreted the pathology findings. Ephrem Berhe reviewed the initial draft, performed the literature search, and finalized the manuscript. All authors read and approved the final revised manuscript for submission.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Kyle R, Gertz M, Witzig T, Lust J, Lacy M, Dispenzieri A, *et al*. Review of 1027 patients with newly diagnosed multiple myeloma. Mayo Clinic Proc 2003;78:21-33.
2. Varettoni M, Corso A, Pica G, Mangiacavalli S, Pascutto C, Lazzarino M. Incidence, presenting features and outcome of extramedullary disease in multiple myeloma: A longitudinal study on 1003 consecutive patients. Ann Oncol 2010;21:325-30.
3. Damaj G, Mohty M, Vey N, Dincan E, Bouabdallah R, Faucher C, *et al*. Features of extramedullary and extraosseous multiple myeloma: A report of 19 patients from a single center. Eur J Haematol 2004;73:402-6.
4. Rai S, Sridevi H, Acharya V, Lobo F, Kini J. Pulmonary plasmacytoma in multiple myeloma: A rare case of extramedullary spread. Egypt J Bronchol 2015;9:293-5.
5. Kintzer J. Thoracic and pulmonary abnormalities in multiple myeloma. A review of 958 cases. Arch Intern Med 1978;138:727-30.
6. Saha R, Bhattacharya A, Deb J, Nayak P, Das B. Multiple myeloma presenting as thoracic plasmacytoma-two rare cases. Egypt J Chest Dis Tuberculosis 2014;63:267-71.
7. Prasad R, Verma SK, Sodhi R. Multiple myeloma with lung plasmacytoma. Lung India 2011;28:136-8.
8. Shih L, Dunn P, Leung W, Chen W, Wang P. Localised plasmacytomas in Taiwan: Comparison between extramedullary plasmacytoma and solitary plasmacytoma of bone. Br J Cancer 1995;71:128-33.
9. Rajkumar SV, Dimopoulos MA, Palumbo A, Blade J, Merlini G, Mateos M, *et al*. International Myeloma Working Group updated criteria for the diagnosis of multiple myeloma. Lancet Oncol 2014;15:e538-48.
10. Moulopoulos L, Granfield C, Dimopoulos M, Kim E, Alexanian R, Libshitz H. Extraosseous multiple myeloma: Imaging features. AJR Am J Roentgenol 1993;161:1083-7.
11. O'Sullivan P, Müller N. Pulmonary and nodal multiple myeloma mimicking lymphoma. Br J Radiol 2006;79:e25-7.
12. Kushwaha R, Kumar S, Mehra S, Prasad R. Pulmonary and nodal multiple myeloma with a pleural effusion mimicking bronchogenic carcinoma. J Cancer Res Ther 2009;5:297-9.
13. Sevcikova S, Minarik J, Stork M, Jelinek T, Pour L, Hajek R. Extramedullary disease in multiple myeloma-controversies and future directions. Blood Rev 2019;36:32-9.
14. Touzeau C, Moreau P. How i treat extramedullary myeloma. Blood 2016;127:971-6.
15. Kumar SK, Callander NS, Adekola K, Anderson L, Baljevic M, Campagnaro E, *et al*. Multiple myeloma, Version 3.2021, NCCN clinical practice guidelines in oncology. J Natl Compr Cancer Netw 2020;18:1685-717.
16. Makoni M. Conflict in tigray impeding basic care for patients with cancer. Lancet Oncol 2022;23:842.

How to cite this article: Tsegay F, Berhe E, Weldezgina H. Multiple myeloma presenting as a lung plasmacytoma: A case report. J Pan Afr Thorac Soc 2023;4: 50-3.