A Rare Presentation of an Uncommon Disease - Intracardiac and Pulmonary Plasmacytoma Presenting With Respiratory Symptoms - A Case Report

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Abstract
Multiple myeloma (MM) is part of a spectrum of diseases ranging from monoclonal gammopathy of unknown significance (MGUS) to plasma cell leukemia. It can have a varied presentation ranging from asymptomatic to severely symptomatic depending on the extent of end organ involvement. The extramedullary spread of multiple myeloma is a rare entity resulting from the hematogenous spread of neoplastic plasma cells with a reported incidence of 7% to 18% in newly diagnosed MM (1). MM with extramedullary disease affecting multiple sites is an extremely rare entity (2). We report a case of multiple myeloma with pulmonary and intracardiac plasmacytoma at diagnosis in a 61-year-old male.

Keywords: *Multiple myeloma, Pulmonary plasmacytoma, cardiac plasmacytoma*

Introduction

Multiple myeloma is a plasma cell malignancy characterized by the neoplastic proliferation of plasma cells. It accounts for approximately 2% of neoplastic disease and 13% of hematologic malignanies (3). MM is defined by the presence of bone marrow clonal plasma cells >10% or bony or biopsy proven extramedullary plasmacytoma and by myeloma defining events such as renal insufficiency, hypercalcemia, anemia, osteolytic bone lesions and presence of free light chain in serum (4). The plasma cell proliferation occurs in the bone marrow. However 'bone marrow escape' of a subclone of plasma cells may result in infiltration of soft tissues and formation of extramedullary disease. Previous studies have reported lymph nodes, liver, kidney, skin, breast, chest wall and paraspinal area to be the most frequent sites of infiltration (1,5). Extramedullary disease in MM is associated with aggressive disease with poor prognosis and increased rates of relapse (6).

Pulmonary involvement in multiple myeloma is rare and has been reported in the form of pulmonary infections, pleural effusions, pleural and pulmonary plasmacytomas and pulmonary infiltration (7, 8). In this case report we present a rare case of Multiple myeloma with pulmonary plasmacytoma which initially mimicked lung carcinoma with metastasis. Subsequent investigations confirmed the diagnosis of multiple myeloma and coexisting intracardiac plasmacytoma.

Case Report

A 61 year old male presented to Respiratory unit 2 of National Hospital, Kandy, Sri Lanka with productive cough and pleuritic chest pain for 6 weeks duration. There was no associated hemoptysis or fever. He also complained of significant constitutional symptoms with loss of appetite and loss of weight of 2kg over the preceding two months. There was no past history or contact history of tuberculosis .He also reported of a persistent severe lower back pain which started abruptly 3months ago while he was trying to squat. The pain was sharp and was exacerbated by movement and only partly relieved by rest and immobilization. The patient developed progressive lower extremity weakness and numbness following this event further impairing his mobilization. There was no bladder or bowel incontinence. His systemic review was otherwise unremarkable and the past medical history was negative for any chronic illnesses. He was a smoker with 10 pack years and consumed alcohol minimally.

Examination revealed moderate pallor, finger clubbing and bilateral mild pitting oedema in lower limbs. There was no cervical or axillary lymphadenopathy. Respiratory and cardiac system examinations were standard. There was localized tenderness over the mid back region mainly over the lower thoracic vertebrae.

Neurological examination was remarkable with spastic gait with spastic paraparesis, quadrihyperrelexia and a sensory level at T12.

Initial investigations revealed white cell count of 4000 /microlitre with normal differential, haemoglobin 8.6 g/dl with normochromic normocytic indices and platelet count of 186000/mm3. Erythrocyte sedimentation rate was 135mm/hour and C- reactive protein 21mg/l (normal<10). Sputum for acid fast bacilli and retroviral screening were negative. His chest X ray revealed a rounded peripheral opacity in the right lung with lytic lesions over the right scapula. (Figure 01) A computed tomographic scan of the thorax (CT thorax) revealed a right upper lobe lung mass with pleural and adjacent rib involvement, multiple lytic lesions of cervical and thoracic spine and possible pathological fractures of C6 and T10 vertebrae suggestive of metastatic disease with primary lung carcinoma. (Figure 02)

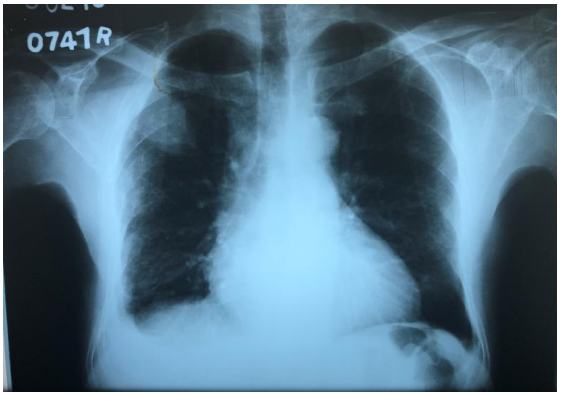

Figure 01: Chest radiography revealing a peripheral opacity (thick arrow) in the right lung with lytic lesions in the right scapula (thin arrow)

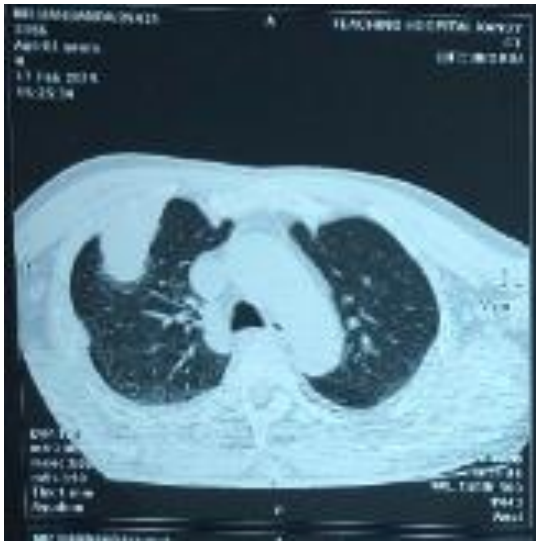

Figure 02: Computed tomographic scan of the thorax revealed right upper lobe lung mass with pleural involvement

Liver transaminases, serum bilirubins, renal functions and electrolytes were normal. Further investigations revealed total serum protein was 8.4g/dl with reversed albumin and globulin ratio, albumin 3.3g/dl and globulin 5.1g/dl. Alkaline phosphatase was 130U/L, serum total calcium level was 2.72 mmol/l and phosphorus 1.52 mmol/l. Urine full report showed 3+ protein urea. His skeletal survey revealed multiple punched out lytic lesions in the skull, bilateral humeri, thoracic vertebrae and T10 wedge fracture. (Figure 03) The biopsy of the lung lesion revealed diffuse sheets of plasma cells including binucleated forms suggestive of plasmacytoma.(Figure 05) Serum protein electrophoresis showed a monoclonal band in the gamma region (M prtein-6g/dl) and urinary examination for Bence Jone's protein was positive. Beta-2 microglobulin was 3560ng/ml. Bone marrow biopsy revealed markedly suppressed erythropoiesis and

granulopoiesis with 95% of total nucleated marrow cells formed by abnormal plasma cells compatible with multiple myeloma. (Figure 04) Thus multiple myeloma with pulmonary palsmacytoma was diagnosed on the basis of investigations. Fiber-optic bronchoscopy and bronchoalveolar lavage (BAL) fluid were examined for the presence of monoclonal plasma cells which was negative.

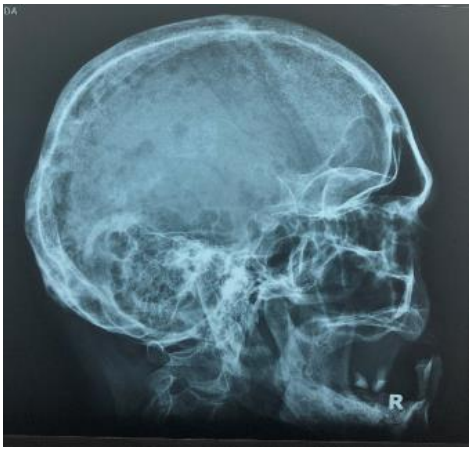

Figure 03: Skull radiograph revealing multiple punched out lytic lesions

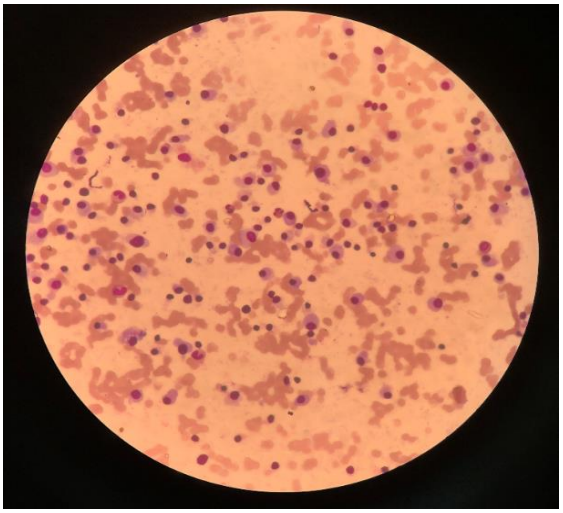

Figure 04: Bone marrow aspiration showing infiltration by abnormal plasma cells

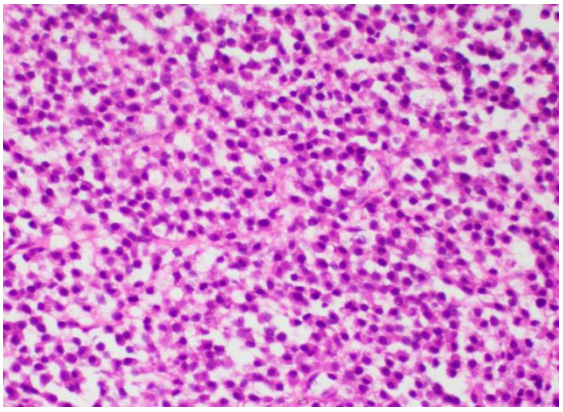

Figure 05: The biopsy of the lung lesion revealed diffuse sheets of plasma cells suggestive of plasmacytoma

A 2D echocardiogram arranged for further assessment revealed a right atrial mass measuring 47mm x28mm obstructing the right ventricular in flow without inferior vena cava involvement. Repeated blood cultures for microorganisms consistent with infective endocarditis were negative.

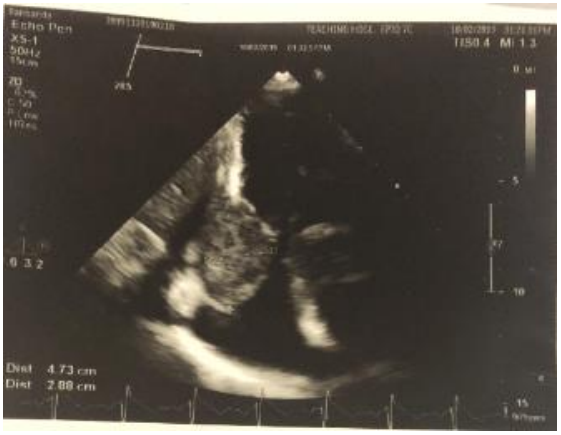

Figure 06: 2D echocardiogram revealing a right atrial mass

Considering the existing MM with the extramedullary disease the cardiac mass was presumed to be an intracardiac plasmacytoma. However, definitive histological diagnosis of the cardiac lesion could not be done as the patient did not consent. The patient was referred for oncological management. The repeat 2D echocardiogram after initial chemotherapy revealed a significant reduction of the intracardiac mass, suggesting it is a palsmacytoma. However the patient succumbed to the illness a few months into treatment.

Discussion

Extramedullary palsmacytoma (EMP) is a solitary mass of neoplastic monoclonal plasma cells. It accounts for about 3% of plasma cell malignancies and the majority is found in the upper aerodigestive tract and head and neck region (9). By definition, EMP does not cause end organ damage such as CRAB (hypercalcemia, renal failure, anemia, and osteolytic bone lesions) and does not cause plasma cell infiltration of the bone marrow. Extramedullary dissemination of multiple myeloma is a rare entity and results from the hematogenous dissemination of neoplastic plasma cells outside the bone marrow. Commonly affected organs are liver, lymph nodes, spleen, thyroid, adrenal lung, pleura, pericardium and skin. Pulmonary parenchymal and cardiac involvements are rare with only few cases reported in literature so far (1, 7). Coexisting pulmonary and intracardiac plasmacytomas at diagnosis of multiple myeloma is an infrequent occurrence and to the best of our knowledge has not been previously reported. Extramedullary disease in MM is associated with poor prognosis and still remains a treatment challenge (1).

This patient presented predominantly with respiratory symptoms with chest radiogram evidence of a mass lesion in the right upper lobe. The background history of smoking and finger clubbing raised the possibility of primary lung carcinoma with bone metastasis. However histological examination of the pulmonary lesion revealed the diagnosis of MM, emphasising the importance of histological examination as routine imaging can sometimes be misleading. Pulmonary plasmacytoma although rare, should always be considered in differential diagnosis in appropriate clinical setting. In this patient the intracardiac mass lesion was diagnosed as an incidental finding while performing routine 2D echocardiogram as part of the general assessment. Primary tumors of the heart in general are rare entities, and benign cardiac myxomas account for up to 50% - 70% of all primary tumors (10). Sarcoma accounts for the majority of malignant primary cardiac tumors while melanoma, leukaemia and lymphoma make up the most of cardiac metastasis. Although cardiac biopsy is essential to confirm cardiac plasmacytoma it was not possible in this case as the patient did not consent for biopsy. However with the background history of multiple myeloma with very high plasma cell burden and biopsy proven pulmonary plasmacytoma, cardiac plasmacytoma was considered as the most likely diagnosis. As a tissue diagnosis was not possible, the reduction in size of the cardiac tumour in response to chemotherapy was taken as further supportive evidence of a cardiac plasmacytoma.

Conclusion

We present a case of multiple myeloma with pulmonary and cardiac dissemination which presented primarily with respiratory symptoms. This case illustrates the importance of considering plasmacytoma, although a rare entity, in the differential diagnosis of pulmonary masses and the value of histology and other routine investigations in establishing the diagnosis.

Consent

Informed written consent was obtained from the patient for publication of this case and accompanying images.

Conflict of Interest

None declared

Authors' contribution DM made the clinical diagnosis and supervised the manuscript drafting. AB, SD, SS and LB drafted the first manuscript, reviewed the literature and involved in direct management of the patient. All authors read and approved the final manuscript.

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References

i. *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 1,003 consecutive patients. Ann Oncol 2009;21(2):325-30*

ii. *Weinstock M, Aljawai Y, Morgan EA, et al. Incidence and clinical features of extramedullary multiple myeloma in patients who underwent stem cell transplantation. Br J Hematol. 2015;169:851-8.*

iii. *National Cancer Institute. Surveillance epidemiology and end results program. SEER stat fact sheets: Myeloma. 2013; http://seer.cancer.gov/statfacts/html/mulmy.html*

iv. *National Comprehensive Cancer Network. NCCN Clinical Practice Guidelines in Oncology, Multiple Myeloma. Version 3 2019. Available at http://www.nccn.org/professionals/physician_gls/pdf/myeloma.pdf.*

v. *S.Z. Usmani, C. Heuck, A. Mitchell, J. Szymonifka, B. Nair, A. Hoering, et al.Extramedullary disease portends poor prognosis in multiple myeloma and is over-represented in high-risk disease even in the era of novel agents Haematologica, 97 (2012), pp. 1761-1767*

vi. *Beksac M, Seval GC, Kanellias N, Coriu D, Rosiñol L, Ozet G, Goranova-Marinova V, Unal A, Bila J, Ozsan H, Ivanaj A. A real world multicenter retrospective study on extramedullary disease from Balkan Myeloma Study Group and Barcelona University: analysis of parameters that improve outcome. haematologica. 2020 Jan 1;105(1):201-8.*

vii. *Sullivan PO, Muller NL. Pulmonary and nodal multiple myeloma mimicking lymphoma. Br J Radiol. 2006;79:e25–7.*

viii. *Niţu M, CriȘan E, Olteanu M, Călăraşu C, Olteanu M, Popescu MR. Lung Involvement in Multiple Myeloma - Case Study. Curr Health Sci J. 2014;40(4):274–276. doi:10.12865/CHSJ.40.04.08*

ix. *Alexiou C, Kau RJ, Dietzfelbinger H, Kremer M, Spiess JC, Schratzenstaller B, et al. Extramedullary plasmacytoma: Tumour occurrence and therapeutic concepts. Cancer. 1999;85:2305–14*

x. *Reynen K. Frequencyof primary tumors of the heart. Am J cardio. 1996;77*