 DOI: 10.18231/2394-6792.2017.0032

Multiple myeloma presenting as bilateral pleural effusion: a rare case presentation with review of literature

Rigvardhan[1,*], Prashant Sengupta[2], Nishant Taur[3], Vasu Vardhan[4]

[1]Assistant Professor, [2]Associate Professor, [3]Resident, Dept. of Pathology, Command Hospital, Lucknow, Uttar Pradesh, [4]Consultant, Dept. of Pulmonary Medicine, Military Hospital (CTC), Pune, Maharashtra

***Corresponding Author:**
Email: vardhanrig@yahoo.com

Introduction

Multiple myeloma (MM) is a clonal proliferation of plasma cells that primarily affects the bone marrow and skeletal system. It mainly affects bone marrow although extramedullary tissues especially reticuloendothelial system may be infiltrated. Serous effusions due to MM are rare.(1) Accurate diagnosis of this condition is important as it portends a poor prognosis with median survival time < 4 months. It is particularly rare (<1%) for MM patients to present as myelomatous pleural effusion (MPE), especially pleural effusion as presenting sign.(2,3) Bilateral myelomatous pleural effusions are even more rare.(2) We describe a case of MM who presented initially as bilateral pleural effusion and was managed in our hospital. *To the best of our knowledge, this is the fourth case in literature where bilateral myelomatous pleural effusion was the presenting feature of multiple myeloma*.

Case Report

A sixty-nine year old male presented with one-month history of fever and breathlessness. The patient also gave history of generalized bodyache, weakness and loss of weight. There was no history of cough, hemoptysis or chest pain. On admission in our hospital he looked ill and was using accessory muscles of respiration. The temperature was 100.6°F, pulse 110/min, respiration 30/min and blood pressure of 90/54 mm Hg. Examination revealed signs of bilateral pleural effusion. Hemoglobin was 7.8 gm/dL; white blood count 8100/cumm with normal differential count and platelets were 355,000/cumm, ESR 120 mm in first hr., blood urea nitrogen 17.1 mg/dL, creatinine 2.4 mg/dL, bilirubin 3.0 mg/dL, aspartate aminotransferase 178 IU/L, alanine aminotransferase 231 IU/L, sodium 137 meq/L and potassium of 4.9 meq/L. His total protein was 8.8 gm/dL and albumin was 2.8 gm/dL. Blood and urine cultures were sterile. Chest X-ray showed bilateral pleural effusion (Fig. 1a). Pleural fluid examination showed hemorrhagic fluid with glucose 87 mg/dL, protein 6.7 gm/dL, albumin 2.4 gm/ dL and LDH of 678 IU/L. There were 3000 cells/cu mm which were predominantly plasma cells (Fig. 1b) including binucleate forms (inset Fig. 1b) comprising 43% of differential count. Cell block was made from pleural fluid which revealed plasma cells (Fig. 1c) and immunohistochemistry with CD138 confirmed presence of plasma cells. Pleural fluid Gram stain, fungal and Acid Fast Bacilli smears were negative. The pleural fluid cultures were sterile. X-ray skull and spine showed multiple punched out lesions suggestive of multiple myeloma. Bone marrow examination revealed 35% plasma cells consistent with the diagnosis of multiple myeloma. Serum protein electrophoresis showed monoclonal spike in gamma region. Serum immunofixation electrophoresis showed monoclonal band in the gamma region corresponding to IgG-kappa immunoglobulin. Patient's condition progressively deteriorated and his renal parameters became worse. Due to deranged renal parameters and poor general condition he was given modified chemotherapy with cyclophosphamide and methylprednisolone. His condition did not improve and patient succumbed to illness. The stay of the patient in our hospital was 4 days and extensive investigations could not be done due to deteriorating general condition of the patient.

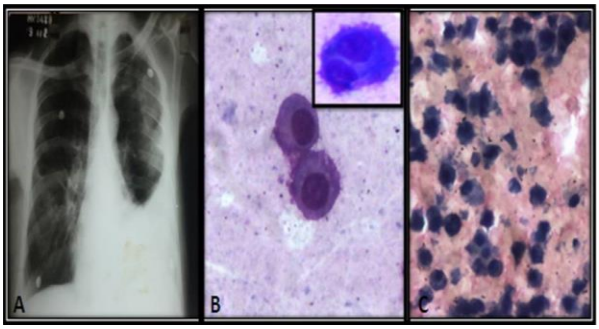

Fig. 1(a): X ray chest showing bilateral pleural effusion (left >right), (b): Pleural fluid showing plasma cells (Leishman stain; 40x); inset showing binucleate plasma cell, (c): Cell block preparation of pleural fluid showing plasma cells (Hematoxylin and eosin stain; 20x)

Discussion

In about 6% of patients with MM, pleural effusion may be a sign of thoracic involvement.(1) Oudart and his colleagues had summarized the six etiologic factors which cause pleural effusion in MM, including congestive heart failure due to amyloidosis, nephritic syndrome secondary to renal tubular infiltration with paraprotein and development of glomerular damage, chronic renal failure, direct infiltration of pleural fluid

from adjacent tissues, hypoalbuminemia, pulmonary embolism, secondary neoplasm, lymphatic drainage obstruction by tumor infiltration, infection and pleural myelomatous involvement.[4] According to published reports, left-sided pleural effusion is most common.[5,6] However, bilateral MPE is extremely rare and only three cases have been reported so far.[2,7]

Given its rarity, a high index of suspicion is required for diagnosis as only about 1% of pleural effusions can be attributed to myelomatous involvement in MM.[1] We believe MPE should always be considered in MM patients presenting with pleural effusions because MPE is associated with an aggressive clinical progression of MM with median survival of 2.8 months despite aggressive local and systemic treatment.[8]

As regards the diagnosis of MPE is concerned, several methods have been described. The best means is the cytological identification of malignant plasma cells as in our case. Plasma cells from the pleural fluid in our case showed typical eccentric nuclei with prominent nucleoli and basophilic cytoplasm. Plasma cells were confirmed on immunohistochemistry with CD138. Reactive plasmacytosis, as seen in tuberculosis and Hodgkin's lymphoma, is usually accompanied by neutrophilic leukocytes, lymphocytes, reactive mesothelial cells, which seldom exceed 15-20% of the cells and have few or no abnormal features.[9] As in our case, the diagnosis is most often revealed by pleural cytology. Protein electrophoresis is another option but is unreliable since most effusions in MM tend to be hemorrhagic.[3,8] Pleural biopsies are less commonly used as patchy pleural involvement in MM leads to low diagnostic yield.[5]

As the majority of the literature published on the subject is in the form of case reports and small series, the relationship between tests/treatment and prognosis is not known. However, as prognosis is poor, palliation is considered the mainstay of treatment and includes chemotherapy pleurodesis and supportive care.[1] There is no chemotherapeutic regimen of choice and many have been used over the time with no distinct advantage of one over other.[10]

To conclude MPE is extremely rare signifying poor prognosis. Consideration of MPE aids in early diagnosis and treatment in this aggressive disease. Cytological identification of plasma cells in the pleural effusion is the mainstay of diagnosis and can easily be done.

Conflict of interest

All authors have none to declare.

References

1. Zhang LL, Li YY, Hu CP, Yang HP: Myelomatous pleural effusion as an initial sign of multiple myeloma – a case report and review of the literature. J Thorac Dis 2014;6:E152–E159.
2. Xu XL, Shen YH, Shen Q, et al. A case of bilateral pleural effusion as the first sign of multiple myeloma. Eur J Med Res 2013;18:7.
3. Adrianzen Herrera, Scott M, Varma M. Myelomatous pleural effusion presenting with extreme hyperferritinemia and severe inflammatory response. Case Rep Oncol 2015;8:349–355.
4. Oudart, JB, Maquart FX, Semouma O, Lauer M, Arthuis-Demoulin P, Ramont L: Pleural effusion in a patient with multiple myeloma. Clin Chem. 2012; 58:672–674.
5. Alshati M, Kumar R, Kannan S: Dysnoea, massive effusion and lytic rib lesion as initial presentation of multiple myeloma in a young man. Can Respir J 2013;20:253–255.
6. Kintzer JS, Rosenow EC, 3rd, Kyle RA. Thoracic and pulmonary abnormalities in multiple myeloma. A review of 958 cases. Arch Intern Med 1978;138:727-30.
7. Dhingra KK, Singhal N, Nigam S, et al. Unsuspected multiple myeloma presenting as bilateral pleural effusion - a cytological diagnosis. Cytojournal. 2007;4:17.
8. Cho YU, Chi HS, Park CJ, Jang S, Seo EJ, Suh C: Myelomatous pleural effusion: a case series in a single institution and literature review. Korean J Lab Med 2011;31:225–230.
9. Gogia A, PK Agarwal PK, Jain S, Jain KP. Myelomatous Pleural Effusion. JAPI 2005;53:734-736.
10. Kamble R, Wilson S, Fassas A, et al: Malignant pleural effusion of multiple myeloma: prognostic factors and outcomes. Leuk Lymphoma 2005;46:1137–1142.