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Myelomatous Malignant Pleural Effusion: A Rare Case Report

Shirin Jamshidi[1], Farzaneh Akbari[2], Parisa Vedad[3], Fariba Rezaeetalab[4,*], Seyedeh Zahra Banihashemian[5]

[1]Department of Pathology, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran
[2]Department of Internal Medicine, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran
[3]Department of Pathology, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran
[4]Lung Diseases Research Center, Faculty of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran
[5]Faculty of Medicine, Semnan University Of Medical Sciences, Semnan, Iran

Abstract

English:

Background: *Multiple myeloma (MM) is a malignant disorder of plasma cells and includes 1% of all malignancies and 10% to 13% of hematologic cancers. Multiple myeloma, characterized by malignant proliferation of plasma cells, leads to the production of monoclonal immunoglobulins. Pleural involvement by plasma cells in multiple myeloma is rare.*
Case Report: *We report a 61-year-old woman with myelomatous pleural effusion in MM.*
Conclusion: *A few cases of pleural effusion secondary to MM, in literature, with an incidence of less than 1%, were reported.*

Keywords

Multiple myeloma • Pleural • effusion

Pleurezie maligna mielomatoasa –o raportare de caz rara

Rezumat

Romanian:

Mielomul multiplu (MM) este o afecțiune malignă a celulelor plasmatice și include 1% din toate afecțiunile maligne și 10% până la 13% din cancerele hematologice. Mielomul multiplu, caracterizat prin proliferarea malignă a celulelor plasmatice, duce la producerea de imunoglobuline monoclonale. Implicarea pleurală a celulelor plasmatice în mielomul multiplu este rară.
Raportăm cazul unei femei de 61 de ani cu revărsat pleural mielomatos în MM. În literature, au fost raportate câteva cazuri de revărsat pleural secundar MM, cu o incidență mai mică de 1%.

Cuvinte-cheie

mielom multiplu • pleural • revarsat

Introduction

Multiple myeloma (MM) is a clonal B- cell malignancy that leads to malignant plasma cells proliferation. These plasma cells accumulate essentially in the bone marrow. MM is the second most common hematological malignancy (1). The most common clinical symptoms of MM include bone pain, pathological fractures, and anemia which are nonspecific (2). Although MM is a bone marrow-based disease, it may also metastasize outside the extramedullary sites, including

*Corresponding author: Fariba Rezaeetalab
E-mail: rezaitalabf@mums.ac.ir

the pleural cavity, which manifests with pleural effusion. Malignant myelomatous pleural effusion is seriously rare in MM, occurring in less than 1% of cases, and it has poor prognosis. The following case report represented a patient with massive unilateral pleural effusion due to multiple myeloma and plasma cell invasion.

Case Report

A 61-year-old woman with dyspnea referred to the emergency room. The patient had IgG-kappa MM, stage I, which was diagnosed two years ago. She had an orthopedic vertebral column surgery 4 months ago. At the time of admission, the patient had vertebral plasmacytoma and collapse in T12 and L2 vertebrae. She denied having a fever and sweating. Examinations of the jugular vein pressure (JVP), thyroid function, and heart auscultation were all normal. There were decreased breath sounds in the lower lobe of the right lung.

The abdomen was soft without organomegaly. A thyroid, liver, and kidney function test, as well as serum electrolytes, were all normal. After serum protein electrophoresis for the patient, monoclonal gammopathy was identified. Detection of Bence Jones protein in the urine (U/A) was negative. In the complete blood count (CBC) normocytic anemia with Hb=8.1 g/100cc, WBC= 2.9 billion cells/L (PMN: 52.1%, Lymph= 35.5%) and platelet=146000/L was noted. PPD test was negative. On further evaluation, CRP was positive, serum protein= 11.7 gr/dL, LDH=362 U/L and alkaline phosphatase 115 U/L. Considering the patient's symptoms, including dyspnea, chest x-ray was obtained at the time of admission on which heart size was normal and massive right pleural effusion was observed. According to prior orthopedic surgery vertebral device was seen (Figure 1). Lung computed tomography (CT) scan confirmed pleural effusion (Figure 2). This patient underwent diagnostic thoracocentesis to test the pleural fluid. The fluid was turbid and bloody. Cytological examination of the pleural fluid showed a hemorrhagic fluid with mesothelial proliferation, and some plasmacytoid cells. In order to rule out mesothelioma and confirm pleural involvement by neoplastic plasma cells, immunohistochemistry staining was performed on the cell block slide. Neoplastic cells were CD138+, Kappa+, Calretinin+, and Lambda–, therefore, malignant myelomatous pleural effusion was approved (Figures 3-7). Pleural fluid was negative for TB (smear and culture) as well as polymerase chain reaction (PCR) for BK was negative. Also adenosine deaminase (ADA) activity was normal. The patient received systemic chemotherapy and she was treated with bortezomib, dexamethasone, and lenalidomide. After a while, due to the patient's financial problems, her prescribed drugs were changed to bortezomib, thalidomide and dexamethasone. The medications had no effect on the course of the disease and unfortunately the patient died 4 months after diagnosis.

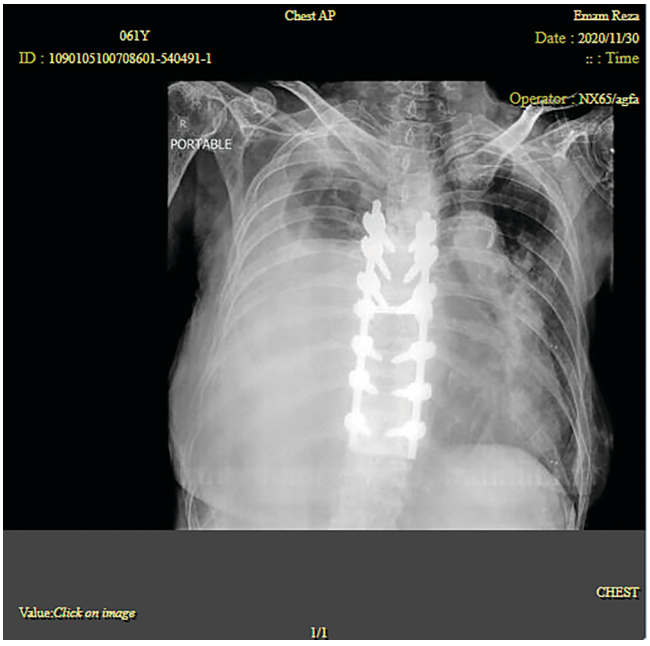

Figure 1. Chest X-ray, right massive pleural effusion.

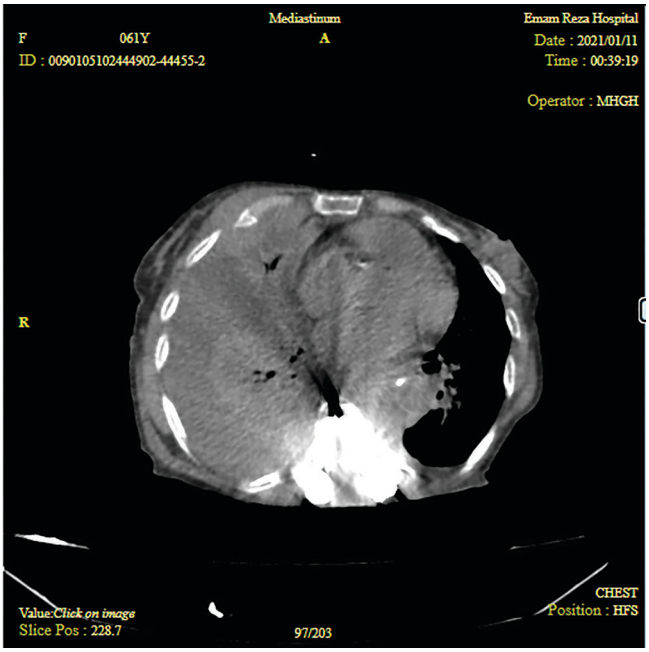

Figure 2. CT of the thorax showed unilateral sided pleural effusions in the right side.

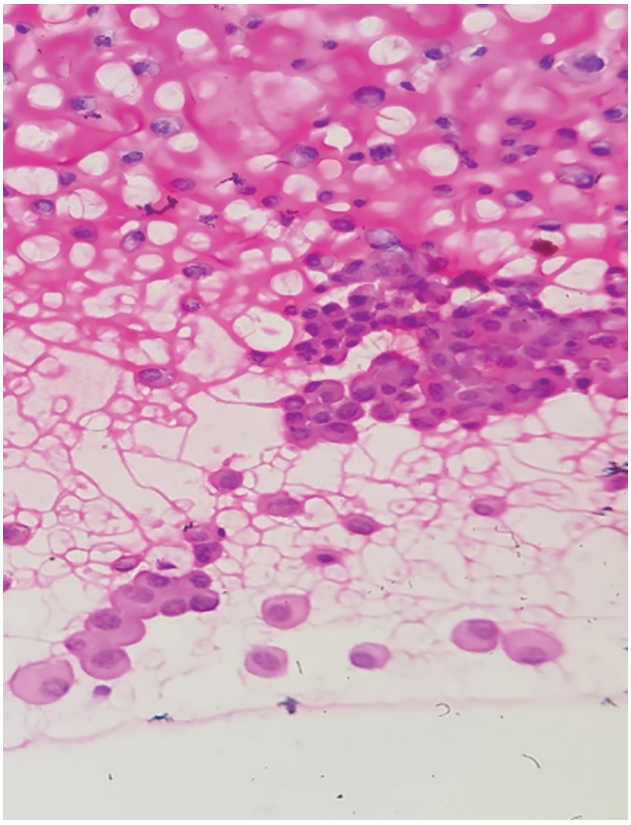

Figure 3. Pleural fluid smear shows plasmacytoid cells with abundant cytoplasm and eccentric nuclei.there are some inflammatory cells in the background.

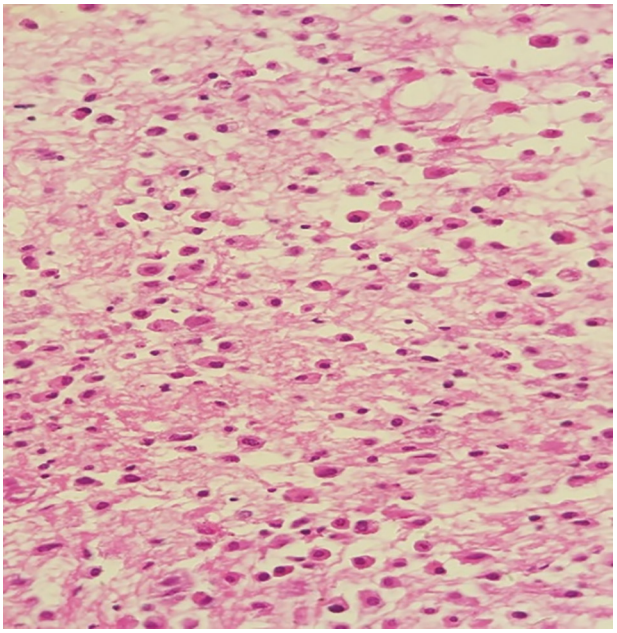

Figure 4. Cell block sections reveal numerous plasmacytoid cells with abundant cytoplasm and eccentric nuclei.

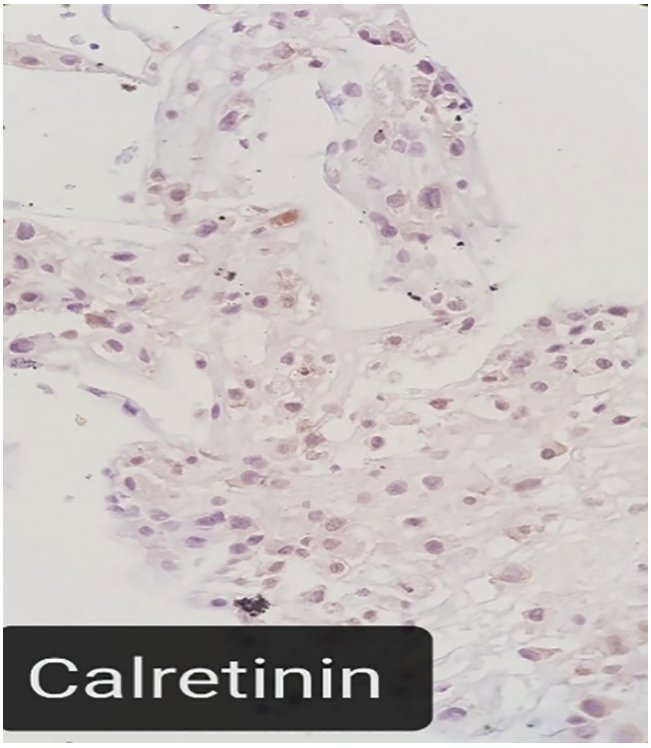

Figure 5. Calretinin is negative in most plasmacytoid cells.

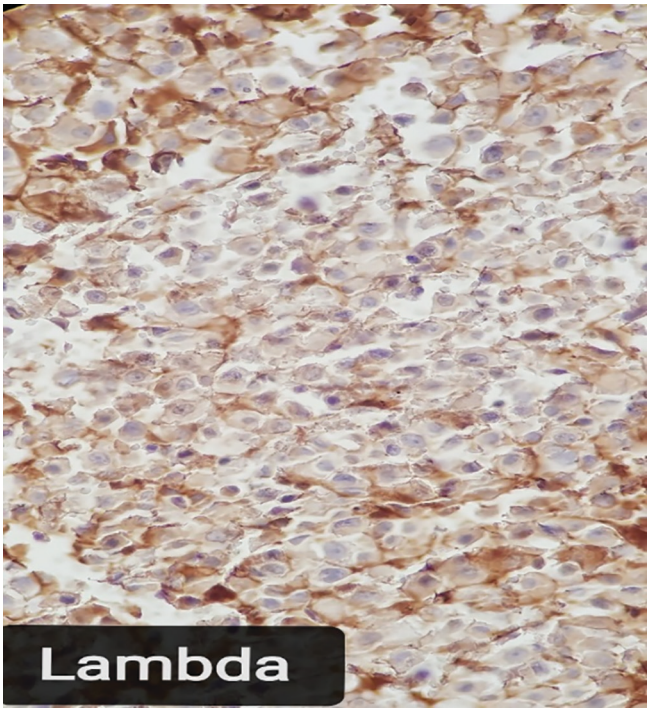

Figure 6. Immunohistochemical staining is negative for lambda light chain in most plasma cells.

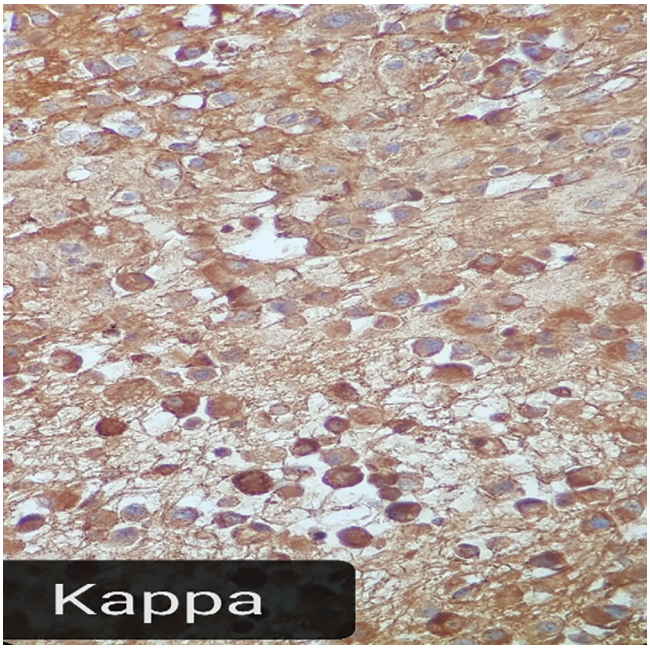

Figure 7. Immunohistochemical staining shows diffuse cytoplasmic kappa light chain positivity in plasma cells.

Discussion

Pleural effusions are not common in patients with MM (approximately 6%) (3). Most of these cases are due to secondary causes like congestive heart failure, due to hyperviscosity or amyloidosis, renal failure, nephrotic syndrome, pulmonary embolism, pneumonia (4). Cardiac and renal functions of our patient were normal, and all secondary causes of pleural effusion in MM were ruled out.

Pleural effusions are rarely involvement with plasmacytoma in MM, with an incidence of less than 1% and was expressed myelomatous pleural effusion (MPE) (5). MPE usually associated with poor prognosis and short survival, left pleura is more commonly affected, and it represented equally between females and males (6). In previous studies, 80 percent of MPE originate from IgA MM (7). Interestingly, our patient had pleural effusion in the right pleura and the patient involved with IgG- kappa type MM.

To confirm MPE, there have been suggested three diagnostic criteria that include: 1) In pleural fluid cytology, detection of atypical plasma cells; 2) on pleural fluid electrophoresis, identification of a monoclonal protein, and 3) for histological confirmation, a pleural biopsy specimen or autopsy (6). In terms of diagnosing, cytological analysis of the pleural fluid is the most effective method. In our patient, cytological and immunohistochemistry examination confirmed the presence of malignant MPE. The pathogenesis of MPE is unknown. Disease invasion from nearby skeletal lesions, chest wall plasmacytoma spread; tumor infiltration of the pleura; and mediastinal lymph node involvement are the mechanisms proposed for the pathogenesis of MPE (7). Since the patient had a vertebral plasmacytoma at the time of referral, it can be concluded that the mechanism of this disease was extension from chest wall plasmacytomas. Although various invasive methods are used to treat this disease, most patients die after a few months. According to studies, the average survival time of these patients after diagnosis is 4 months (8). Therapeutic thoracentesis must be used to alleviate the effects of the effusion. Kamble et al. (9) demonstrated that systemic chemotherapy combined with chest tube drainage or pleurodesis had excellent palliative effects. The British Thoracic Society pleural disease guideline cover the diagnosis, treatment, and follow-up of patients with malignant pleural effusion, including the use of thoracentesis, pleural biopsy, and pleurodesis.The BTS's guidelines provide a comprehensive approach to the management of malignant pleural effusion, including MPE, taking into account the patient's quality of life. Most responses to the pleurodesis are transient and recurrent(10). The importance of meylomatous pleural effusion is the negative effect on the course of the disease and inverse outcome (4).

So, accurate, effective treatment and diagnostic methods for this disease have not been provided so far. Most studies related to this disease are case reports and more clinical studies are needed.

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Conflict of Interest

The authors declare no conflict of interest.

Informed Consent

Informed consent was obtained.

Author Contributions

JS and VP obtained and diagnosed the pathology images and edited the manuscript. AF,RF and BSZ wrote and provided expertise. AF finalized edition. All authors have read and approved the manuscript.

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Institutional Review Board Statement

Not applicable.

Data Availability Statement

Data used to support the findings of this study are available from the corresponding author upon request.

Conflicts of Interest

The authors declare no conflict of interest.

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