Myelomatous Effusion in Plasma-Cell Leukemia: a Case Report

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ABSTRACT

Myelomatous pleural effusion is extremely rare and cases with cavitary involvement are mostly IgA type myeloma. The effusion is thought as a late manifestation in the natural history of multiple myeloma or an expression of the aggressive behaviour of the disease and a very aggressive treatment is indicated. A case of myelomatous effusion presenting as plasma-cell leukemia two years after initial diagnosis of IgG type multiple myeloma is described.

Key Words: Multiple myeloma, Myelomatous effusion, Plasma-cell leukemia

ÖZET

Plazma Hücreli Lösemi ve Myelomatöz Efüzyon: Olgu Sunumu

Myelomatöz efüzyon oldukça nadir görülmekte olup, olgularda kaviter tutulum sıklıkla IgA tip multiple myeloma ile birliktelik göstermektedir. Efüzyon hastalığın doğal sürecinde geç dönemde ortaya çıkan bir bulgu olmakla birlikte, hastalığın saldırgan özelliğinin bir sonucu da olabilir ve agresif bir tedavi yaklaşımı gerektirir. İki yıl önce IgG tip multiple myelom tanısı alarak, myelomatöz plevral efüzyon olarak değerlendirilen ve plazma hücreli lösemi olduğu tesbit edilen olgu literatür eşliğinde sunuldu.

Anahtar Kelimeler: Multiple myeloma, Myelomatöz efüzyon, Plazma hücreli lösemi

INTRODUCTION

Multiple myeloma (MM) is a plasma-cell neoplasm that is characterized by skeletal destruction, renal failure, anemia, hypercalcemia and monoclonal gammopathy (1). Multiple myeloma and related diseases are known to present with various clinical manifestations (2-6). Pleural effusion in multiple myeloma is relatively infrequent and myelomatous pleural effusion is extremely rare. Here we report a case of plasma cell dyscrasia who developed pleural effusion during follow up.

CASE REPORT

A 74-year-old man who was diagnosed with multiple myeloma 2 years prior, was admitted with exertional dyspnea and fatigue. Physical examination revealed moderate respiratory distress with a respiratory rate of 30/minute, mild tachycardia and no breath sounds at left hemithorax with decreased breath sounds at right lung base. Laboratory examination was as follows; hemoglobin 8 g/dl, white blood cell count $6.8x10^9$ /L with 55% immature plasma cells in the peripheral smear, platelets $101x10^9$/L, creatinine 2.4 mg/dl, blood urea nitrogen 29 mg/dl, albumin 3.5 g/dl, globulin 6 g/dl. Chest x-ray showed bilateral pleural effusions. A computed tomography (CT) of chest revealed bilateral massive pleural effusions and nodular densities which were localized on parietal pleura. On echocardiographic examination, ejection fraction was normal with no pericardial effusion.

Two years before his admission, he was diagnosed with multiple myeloma of IgG-kappa type. His disease was refractory to melphalan plus prednisone (MP) and vincristine, doxorubicin and dexamethasone (VAD) combination regimens respectively. He has been on thalidomide 150 mg/day for the last fifteen months.

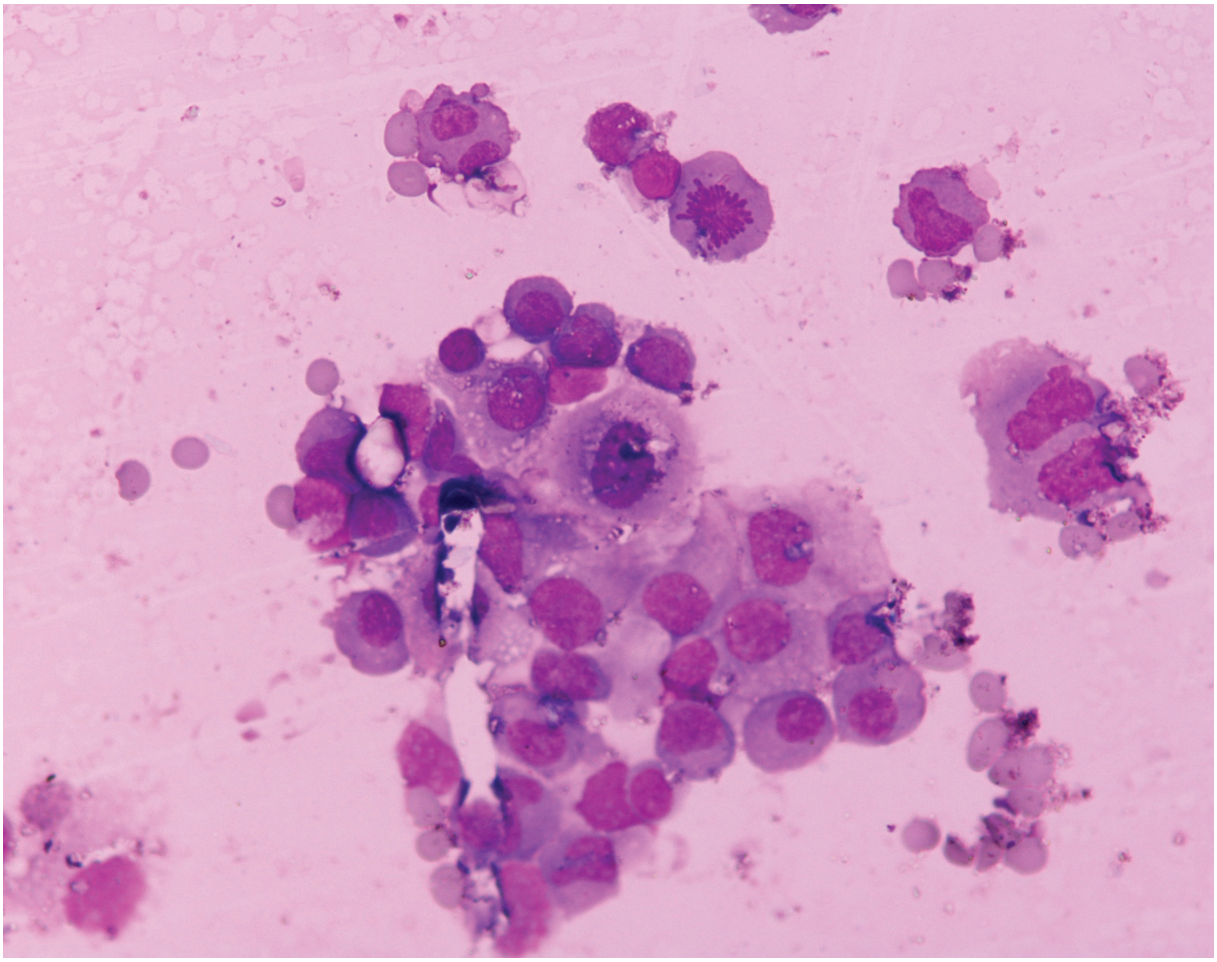
A thoracentesis was performed and 1000 cc of serohaemorragic fluid was removed. Cytospin preparations of the fluid showed plasma cells (Figure 1). Flowcytometric analysis of the pleural fluid revealed that 99% of the cells were expressing CD 38. Therefore, the patient was accepted as plasma-cell leukemia with myelomatous pleural effusions. He was elected to receive supportive care with large volume thoracentesis and transfusions because he refused further treatment.

DISCUSSION

Pleural effusions in multiple myeloma occur in about 6% of patients, commonly due to non-spesific manifestations of the disease such congestive heart failure, pulmonary embolism, amyloidosis and chronic renal failure (7,8). In the presence of pleural effusion in a myeloma patient, secondary causes other than myelomatous involvement should be excluded (9). Cytologic examination of cells obtained from the pleural effusion, immunophenotyping by flow cytometry, as well as molecular analysis by polymerase chain reaction applied to cytology specimens can contribute to differential diagnosis. Obtained findings sometimes need to be confirmed by pleuroscopy or thoracoscopy with biopsy (9).

Sasser et al analyzed 56 cases of myeloma with the involvement of the serous cavities in the literature (10). The sites of involvement included the pleural cavity in 30, the peritoneal cavity in 14, the pericardial cavity in 2, and the central nervous system in 10 cases (10). In another review of 958 cases of multiple myeloma, 58 patients had pleural effusions and pleural involvement was documented in 8. Over 50% of the cases with cavitary involvement are of Ig A type (11), while Ig A myeloma represents approximately 25% of all types of multiple myeloma. Of note, our patient had Ig G type myeloma. Multiple myeloma associated with myelomatous pleural effusion has a very poor prognosis with reported length of the survival of less than four months (5). The effusion is thought as a late manifestation in the natural history of multiple myeloma or an expression of the aggressive behaviour of the disease and a very aggressive treatment is indicated (9,12).

It was thought that a major determinant in the development of myelomatous effusion is the production of large quantities of immunoglobulin by malignant plasma cells within the pleural cavity which raises the colloid osmotic pressure of the fluid to such a level that normal absorption can not take place (13). It is also likely that disturbed homing behaviour of myeloma cells may cause extramedullary tumor formation at unusual sites, such as liver, lung, peritoneal and pleural cavities and therefore may also cause a leukemic form of the disease in the circulation, especially during the end stage of the disease (14).



Picture 1. Plasma cells on cytospin preparation of pleural fluid. Wright stain. Original magnification x50.

In our case, the patient had a history of multiple myeloma for two years. He did not have findings of congestive heart failure and also his echocardiographic examination normal. He was afebrile and there were not any infiltrative lesions at thorax CT. Diagnosis was made by examining cytospin preperations and flowcytometric analysis of thoracentesis material. Our case also conforms to the findings seen in end stage disesase with refractoriness to the treatment and presentations with plasma cell leukemia and myelomatous effusion.

In conclusion, myelomatous pleural effusion and plasma cell leukemia may be seen as agressive and usually late manifestations of end stage multiple myeloma.

REFERENCES

1. Kyle R A, Rajkumar V. Multiple myeloma. N Engl J Med 351:1860-1873,2004.
2. Mitchell MA, Horneffer MD, Standiford TJ. Multiple myeloma complicated by restrictive cardiomyopathy and cardiac tamponade. Chest 103:946-947, 1993.
3. Inoue Y, Chua K, McClure RF, et al. Multiple myeloma presenting initially as a solitary pleural effusion later complicated by malignant plasmacytic ascites. Leuk Res. 29:715-718, 2005.
4. Kim YM, Lee KK, Oh HS, et al. Myelomatous effusion with poor response to chemotherapy. J Korean Med Sci 15:243-246,2000.
5. Meoli A, Willsie S, Fiorella R. Myelomatous pleural effusion. South Med J 90:65-68, 1997.
6. Ben Ghorbel I, Ksontini I, Houman MH, et al. Bilateral myelomatous pleurisy: a case report. Rev Med Interne 22:1134-1135, 2001.

7. John CH, May LV. Pleural effusion in multiple myeloma. Cancer 44:1150-1154, 1979.
8. Quinquenel ML, Moualla M, Le Coz A, et al. Pleural involvement of myeloma. Apropos of 2 cases. Rev Mal Respir 12:173-174, 1995.
9. Michael G. Alexandrakis, FH. Passam, et al. Pleural Effusions in Hematologic Malignancies. Chest 125:1546-1555, 2004.
10. Sasser RL, Yam LT, Li CY. Myeloma with involvement of the serous avities. Cytologic and immunochemical diagnosis and literature review. Acta Cytol 34:479-485, 1990.
11. Kintzer JS, Rosenow EC III, Kyle RA. Thoracic and pulmonary abnormalities in multiple myeloma: a review of 958 cases. Arch Intern Med 138:727-730, 1978.
12. Caroline C, Louis CW. Response of myelomatous pleural effusion to chemoterapy. Chest 80:765-766, 1981.
13. Wing CK, Stephen CL, Paul K. Kappa light-chain myeloma with pleural involvement. Chest 86:494-496, 1984.
14. Van Riet I. Homing mechanisms of myeloma cells. Pathol Biol 47:98-108, 1999.

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