Myelomatous pleural effusion: A case report

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Abstract- Multiple myeloma is a common hematological malignancy. Serous effusions are described uncommonly in myelomas. A myelomatous effusion however, is extremely rare and carries a poor prognosis. We report a case of multiple myeloma with leukemic transformation and simultaneous pleural fluid involvement in a 66-year old patient who was receiving chemotherapy. The patient developed exceptional dyspnea, dry cough and fatigue prior to admission. Radiological investigations revealed a massive pleural effusion with partial collapse of the lung. Pathological examination of the fluid showed plasma cells and plasmablasts with abnormal forms, consistent with a myelomatous pleural effusion. Hematopoietic neoplasms are associated with body cavity effusions but rare etiologies such as myeloma should also be considered as the treatment protocols differ in these cases.

Index Terms- Myelomatous pleural effusion, Plasmablast.

I. INTRODUCTION

Multiple myeloma is a clonal proliferation of plasma cells that primarily affects the bone marrow and the skeletal system. Extraosseous involvement of the reticuloendothelial system is not uncommon. It comprises about 1% of all malignancies as well as 10% of all the hematopoietic neoplasms. The mean age at diagnosis is around 65 years[1]. Malignant pleural effusions due to myeloma are extremely rare[2]. The incidence is 1-2% of all cases of multiple myeloma[3,4]. An accurate diagnosis of this condition is imperative as it portends a poor prognosis.

We present a case of malignant pleural effusion arising in a 66 year old female patient with long standing multiple myeloma; cytologically typed as a myelomatous effusion.

II. CASE REPORT

A 66 year old female patient, a known case of myeloma, presented to the outpatient department with a history of progressive lower backache, generalized weakness and joint pains. She also had a concomitant history of type2 diabetes mellitus, hypertension and hypothyroidism. Complete hemogram showed an anemic profile (Hb: 9gm% and ESR: 104 mm/hour) with mild leucocytosis (11500 /mm^3) and thrombocytopenia (platelets: 84000 /mm^3).The differential counts showed an increase in plasmacytoid lymphocytes and plasma cells with occasional plasmablasts. A repeat bone marrow examination showed myeloma not in remission.

Six days after admission, the patient developed a rapid onset dyspnea and chest pain. The radiological investigations comprising of a chest radiograph and a CT scan showed a massive pleural effusion of the left lung with partial collapse of the underlying lung parenchyma (fig.1).

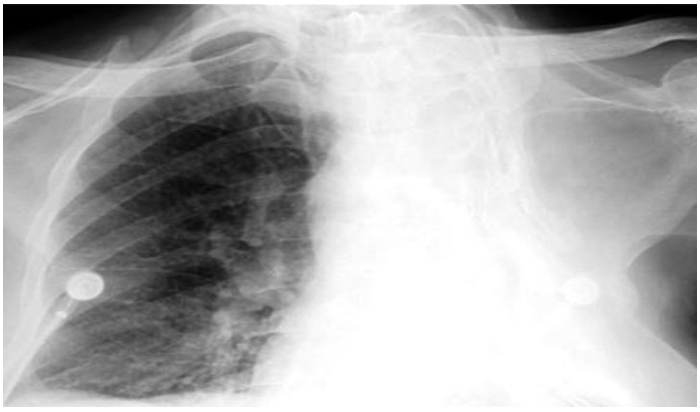

Fig 1: Chest radiograph showing a massive left sided pleural effusion and partial collapse of the lung parenchyma.

Serum electrophoresis also suggested a rapid disease progression. The follow up smear revealed a leukemic transformation of myeloma, with plasmablasts numbering 32 %. A thoracocentesis intended for both therapeutic and diagnostic purposes was carried out and the pleural tap was analyzed in our department.

The pleural fluid was heavily blood stained with features suggestive of an exudate (pH: 7.9, total protein: 3.2gm/dL, glucose: 142mg/dL and LDH: 2043U/L).The WBC counts were 12000 /mm^3 and the RBC count was 0.27x10^6 /mm^3.

The smears studied showed a marked increase in plasma cells and lymphoid cells against a background of hemorrhage (fig. 2). Numerous plasmablasts were seen with prominent nucleoli and a basophilic cytoplasm (fig 3).

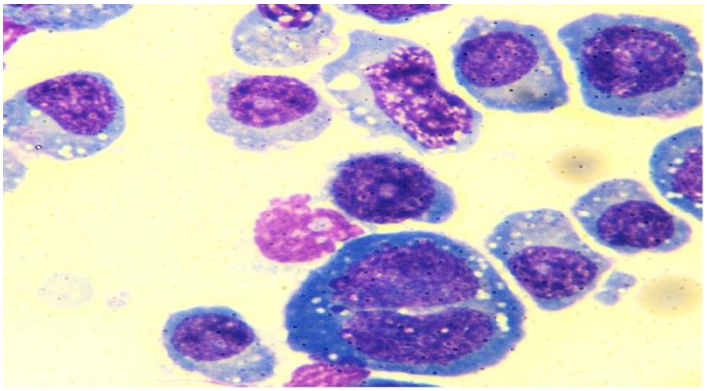

Fig 2: Plasma cells and lymphocytes in the pleural fluid. (Leishman 400x)

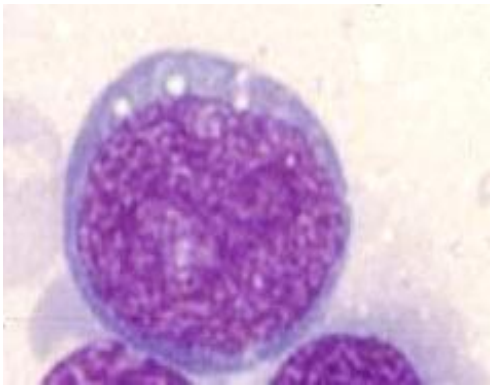

Fig 3: Plasmablast with reticular, immature chromatin and basophilic cytoplasm (Leishman ; 400x)

Few large abnormal lymphoid cells with dense chromatin, irregular nuclear contours and cytoplasmic immunoglobulin secretions were also noted .We gave a diagnosis of a myelomatous effusion with a request for subsequent follow up.

The brief follow up period after thoracocentesis was uneventful and a pleurodesis with bleomycin was planned. At the time of submission of this report, the patient was discharged against advice.

III. DISCUSSION

Pleural effusion in multiple myeloma is a rare event, reported in approximately 6% of cases[5].The first described case was reported in 1994 by Rodriguez and colleagues[5]. A vast majority of these cases are not attributed to myeloma but to related pathologies such as heart failure secondary to amyloidosis, pulmonary embolism, chronic renal failure, a second neoplasm, nephrotic syndrome[5, 6]. A myelomatous effusion has seldom been reported in literature and is said to occur in less than 1% of all cases with approximately 80% of these being in IgA disease[4]. A Mayo Clinic review of 958 cases with multiple myeloma included 58 cases with pleural effusion. However only 0.8% were found to have effusions due to myeloma. In this series the most common cause of effusion was congestive cardiac failure due to amyloidosis[7].

Several probable mechanisms have been postulated for myelomatous pleural effusion: invasion from adjacent skeletal lesions, extension from chest wall plasmacytomas, tumor infiltration of pleura and mediastinal lymph node involvement causing lymphatic obstruction[6]. Rarely, a pleural effusion can be secondary to a myelomatous ascites[8]. Our patient had a leukemic transformation of myeloma and thus a pleural cavity involvement.

As far as the diagnosis of myelomatous pleural effusion is concerned, several methods are available. The cytological examination of pleural fluid for malignant plasma cells still continues to remain the most widely used technique. In our case, plasma cells showed typical basophilic cytoplasm with large eccentric nuclei. Few plasmablasts with prominent nucleoli were also seen, some with cytoplasmic immunoglobulin vacuoles.

Apart from exfoliative cytology, ancillary techniques such as fluid electrophoresis and flow cytometric evaluation for plasma cell markers such as CD38, CD138 and light chain restriction[9,10]. Other methods include a pleural biopsy with immunohistochemical markers.

The differential diagnosis includes reactive plasmacytosis, as seen in tuberculosis, viral infections and Hodgkin lymphoma. They are associated with neutrophils, reactive mesothelial cells and lymphocytes and the plasma cells seldom exceed 15-20%, lacking abnormal features[3].

Myelomatous pleural effusion has been thought as a late manifestion in the natural history of multiple myeloma or an expression of the aggressive behavior of the disease. Regardless, it portends a poor prognosis with reported length of survival less than four months after onset[9]. The mainstay of treatment is systemic chemotherapy as myelomatous effusion is dependent on excess production of monoclonal protein. In this case, the patient's condition improved immediately after thoracocentesis but continued to have progressive dyspnea and fatigue. At the time of submitting this article, no subsequent follow-up was available.

IV. CONCLUSION

Myelomatous pleural effusions are a rare occurrence. It indicates an advanced disease and a poor prognosis for the patient. Cytologic examination continues to be an effective method to diagnose these cases and to rule out other causes of body cavity effusions. Ancillary techniques also help in this regard.

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