Myelomatous Pleural Effusion
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Abstract
Serous effusions in multiple myeloma are uncommon but a myelomatous pleural effusion occurring in these patients is extremely rare. Here we report a rare case of a 38 years lady who was diagnosed to have multiple myeloma and subsequently developed pleural effusion. The myelomatous nature of the effusion was first diagnosed on cytology and subsequently confirmed by a pleural biopsy. The pleural effusion showed an initial response to chemotherapy but subsequently recurred.

INTRODUCTION
Multiple myeloma is a clonal proliferation of plasma cells that primarily affects the bone marrow and skeletal system. Extraosseous involvement of myeloma is usually seen in the reticuloendothelial system. Serous effusions due to multiple myeloma are extremely rare. Accurate diagnosis of this condition is important as it portends a poor prognosis. Morphologic and flow cytometric evaluation are the best methods to diagnose this condition. Here we report a rare case of myelomatous pleural effusion occurring in a 38 years lady with multiple myeloma.

CASE REPORT
A 38 years female presented in September 2001 with intermittent backache in the mid thoracic region of one year's duration. She also complained of pain over the right chest wall and right thigh for six months. The patient did not have any pallor, jaundice, lymphadenopathy or organomegaly. Bony tenderness was felt over the sternum and right lower ribs. Her hemoglobin at this time was 11.25 gm%, total count and platelet counts were normal. The erythrocyte sedimentation rate was increased at 107 mm, serum creatinine was 0.91mg%, blood urea 39 mg% and calcium 8.7 mg%. The protein electrophoresis did not show any M band. Bone scan showed multiple hot spots in the skeletal system. CT Scan of the spine showed a destructive lesion at D6 and D8. MRI showed a collapse of D6 vertebra and marrow changes in D4 and D8. Bone marrow aspiration was done and showed 15% plasma cells. The bone marrow was repeated after 15 days. The marrow picture was suggestive of a plasma cell dyscrasia with 16% immature plasma cells. An FNAC from the rib lesion showed plasma cell neoplasia of bone. Serum electrophoresis repeated at this time showed a dense M band of 1.25mg/dl IgG type of monoclonal protein with lambda light chain restriction. Urine Bence Jones proteins was positive. Urine electrophoresis showed abnormal lambda light chain protein. X-ray spine showed lytic lesions in the right iliac blade, right proximal femur, ala of the right sacrum and collapse of D6 vertebra. Lytic lesions were also seen in the right eight rib. The left cardiophrenic angle was not well visualized on chest X-ray. The patient was started on VAD (vincristine, adriamycin and dexamethasone) regimen. The bone marrow repeated after four cycles of chemotherapy was normal. The serum electrophoresis did not reveal any M band. The patient remained in remission for a year and eight months and received only oral calcium and Inj Aredronate. She came back in May

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Fig. 1 : Pleural fluid cytology shows large atypical plasma cells
2003 with weakness. Her blood counts were normal. Plasma cells were not present in the peripheral blood. The ESR was raised at 130 mm. Protein electrophoresis showed an M band (3.2 gm/dl). A pleural effusion of the left side was detected on chest X-ray. Pleural fluid cytology showed numerous immature and atypical plasma cells admixed with more mature forms. A diagnosis of myelomatous pleural effusion was made. She was started on pulse dexamethasone and thalidomide. Her pleural effusion responded and the M band also improved on this treatment. However the patient could not tolerate this chemotherapy and she was put on melphalan. The effusion recurred within a few months. A pleural biopsy done at this time showed sheets of plasma cells which confirmed the myelomatous nature of the effusion.

**DISCUSSION**

While pleural effusions in multiple myeloma are infrequent, a myelomatous pleural effusion is extremely rare. According to a Mayo Clinic review, the frequency of pleural effusion in multiple myeloma was 6% while there were only 0.8% cases of myelomatous effusions. The effusion generally develops as a late complication of the disease but there have been reports of patients presenting with pleural effusion at the time of diagnosis. Eighty percent of these cases are associated with IgA myeloma followed by those due to IgG. Kim et al have reported a case of myelomatous pleural effusion associated with IgD-λ myeloma. Multiple myeloma in our patient was of IgG-λ type. Serous effusions in multiple myeloma usually indicate a poor prognosis and death within four months to one year.

Pleural effusions in multiple myeloma commonly occur due to congestive heart failure as a result of either amyloidosis or atherosclerotic heart disease. Other causes include pulmonary embolism, chronic renal failure and second neoplasm. The pathogenesis of myelomatous effusions is unknown. It may result from direct pleural involvement by a myeloma or spread from adjacent skeletal lesions or chest wall plasmacytomas.

In our case the pleural effusion may have occurred due to spread from a rib lesion as well as due to pleural involvement. Reactive plasma cells may be present in serous effusions secondary to cardiac surgery, tuberculosis, Hodgkins disease and carcinomatosis. High cellularity with a predominant plasma cell population featuring immature and atypical plasma cells would favor a diagnosis of myelomatous effusion over a reactive one.

A myelomatous effusion is characterized by a high specific gravity, high protein content and large number of plasma cells. Exfoliative cytology is extremely useful for the initial diagnosis. This may be further supplemented by fluid electrophoresis and flow cytometric (FCM) evaluation for plasma cell markers such as CD38, CD138 and light chain restriction. Pleural biopsy may be done when flow cytometry is not available. In a flow cytometric evaluation of serous fluids for DNA ploidy, Palmer et al have reported an aneuploid DNA content in 60% of their cases.

To conclude, myelomatous pleural effusion is extremely rare. It indicates advanced disease and a poor prognosis. Cytologic evaluation is a useful tool in its diagnosis.

**REFERENCES**