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CASE REPORT

An Unusual Case of Myelomatous Pleural Effusion in Nonsecretory Myeloma

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Abstract

Non-secretory MM account for 3% to 5% of the total MM population and total incidence of pleural effusion (PE) in MM is only 6%. In this case, patient developed PE during the disease course emphasizing the rarity of PE in NSMM. Mostly PE is attributed to infective causes, but the presence of myeloma cells in a PE along with the involvement of bone marrow is a very rare scenario that emphasizes the importance of detailed evaluation of such patients by both clinicians and pathologists.

Key Words

Multiple Myeloma, NSMM, Pleural Effusion

Introduction

Multiple myeloma (MM) is a neoplastic clonal proliferation of plasma cells in the bone marrow that produces an abnormal increase in monoclonal proteins (M proteins)/immunoglobulins (Ig) which are subsequently detected in blood and urine. These myeloma cases account for 10% of hematologic malignancies and 1% of all other cancers.[1] Fascinatingly, since 1950s, it has been noted that a very tiny fraction of the myeloma population is functionally non-secreting, that is no identifiable monoclonal Ig is present in serum or urine is electrophoresis. These non-secretory MM (NSMMs) account for 3% to 5% of total MM population. Since NSMM is very rare within the MM population, therefore nothing much has been known about its clinical course or prognosis.[2] In recent past, occurrence of pleural effusion (PE) due to MM was seen in only 1% of cases and non-MM was seen in 6-14% cases. PE is usually benign in MM and mainly occurs due to congestive heart disease, chronic renal failure, hypoalbuminemia, cardiac amyloidosis, pulmonary infarctions, or infections. However myelomatous pleural effusion (MPE) has a malignant pathology and is associated with poor prognosis.[3] Considering peculiarity of the condition, we would like to report a rare case of NSMM associated with myelomatous pleural effusion in a female with hematological plasma cell disorder.

Case Report

A 49-year-old female, presented to OPD with complaints of shortness of breath and palpitations for 15 days. She was tachypneic (respiratory rate 30-32 breaths/min) with decreased air entry and dullness on percussion on the right side. Routine investigations showed reduced hemoglobin (8.4 gm%), hematocrit of 25.5%, total leucocytes count (TLC) 7.6x10^9/L, differential leucocytes count with polymorphs 57%, lymphocytes 33%,

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eosinophils 01%, monocytes 09%. Her platelet counts and erythrocyte sedimentation rate were normal. Although her hemoglobin was low, her serum ferritin was markedly raised (693.4 ng/ml).

Ultrasonography of the whole abdomen/thorax and chest X-ray revealed massive right-sided and mild left-sided pleural effusion. To look for more causes of PE, contrast-enhanced computed tomography chest and abdomen was performed which revealed significant right-sided pleural effusion with partial passive collapse of the right lower lobe. Many lytic lesions were also noted on the sternum, bilateral ribs, and mandible, suggesting a malignant pathology. The pleural fluid aspirated was exudative in nature with serum lactate dehydrogenase levels (102 IU/L), glucose levels (143 mg/dL), and TLC count (1740 cells) with plasma cells predominance. The Cytospin smears from PE showed numerous atypical plasma cells admixed with lymphocytes and few neutrophils in a hemorrhagic background. (Fig. 1 A & B)

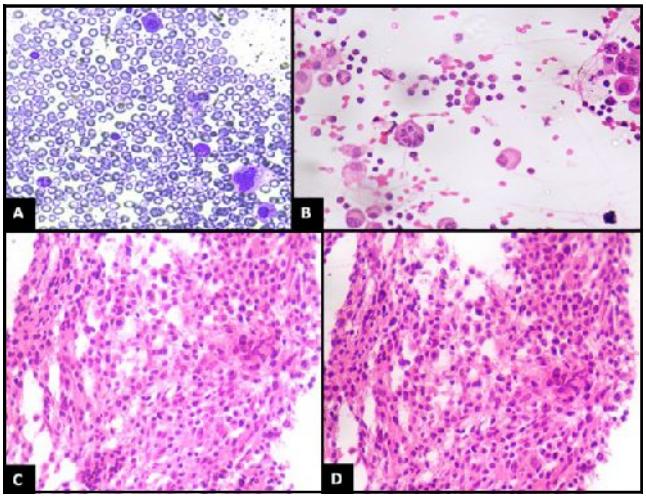

Fig. 1. Multiple atypical plasma cell with eccentric nucleus along with atypical mitotic figures seen on cytospin smears of pleural fluid (A) May-Grünwald Giemsa stain (100X), (B) Hematoxylin & Eosin stain (100X). Bone marrow biopsy shows infiltration by atypical plasma cells involving whole of the marrow. (C&D, 400X)

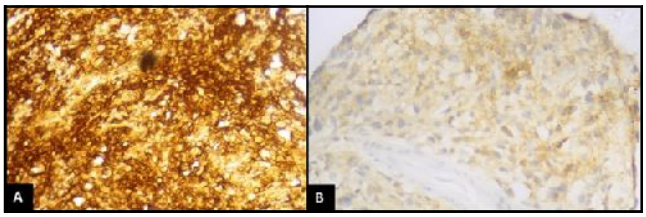

Fig. 2. IHC staining show (A) positivity for CD138 (400X) and light positivity for Lambda (400X)

Further, bone marrow examination (BME) was performed which exhibited massive BM infiltration by 49% of atypical plasma cells. (Fig. 1 C & D) The morphology was representative of many immature and large bizarre forms of plasma cells; however, confirmation was done by immunohistochemistry (IHC) on a paraffin-embedded block of BM biopsy which was positive for CD 138, lambda light chain, and negative for kappa light chain on atypical plasma cells (Fig. 2). For further work-up, a sample for serum protein electrophoresis (SPE) was also sent which revealed an increase in albumin levels (70.2%) and a decreased alpha 2 (4.7) and gamma levels (7.2) with no myeloma band. To confirm the presence of NSMM, serum-free light chain assay (SFLC) was done, which was increased for both free kappa, 155 mg/L, and free lambda, 10.5 mg/L, with a raised free kappa to lambda ratio of 14.76. Thus, a definitive diagnosis of NSMM with MPE was made, and chemotherapy was initiated.

Despite beginning outpatient chemotherapy, her condition worsened and she had numerous readmissions before her untimely death four months after the initial diagnosis.

Discussion

As per the literature, MM accounts for 1% of all malignancy and 2nd most common after lymphoma. It mostly affects the bone marrow but can also infiltrate extramedullary tissues.[1] About 6-10% of individuals with MM have PE, which may indicate thoracic involvement.[1] The incidence of extramedullary disease (EMD) has increased in recent years, most likely as a result of improved imaging tests sensitivity. EMD evidence strongly indicates a bad prognosis, and MPE is not an exception. It has a less than 4-month overall survival rate and a progression-free survival rate even with vigorous therapy.[3] The hematogenous spread is main mechanism behind causing the disease to proceed and significantly worsening the prognosis. The presence of these myeloma cells in a pleural effusion is used to diagnose MPE.[4] Pleural effusions in MM patients are frequently caused by a variety of conditions namely hyperviscosity or amyloidosis-induced congestive heart failure, renal failure, hypercoagulable state or plasma embolization, etc. Thus, MPE origin must be established based on the presence of plasma cells which can easily be identified on

Romanowsky stains in pleural fluid.[5]

Even with the existence of specific criteria, MPE diagnosis is still difficult. The diagnosis of MPE is made in around half of these patients by pleural fluid cytology or pleural biopsy, despite a blind pleural biopsy being a risky technique owing to MPE's uneven effects on the [6] According to Kim *et al.*, these effusions frequently reoccur within months despite rigorous treatment with systemic chemotherapy and radiation.[7]

NSMM is a subset of patients with multiple myeloma who do not have monoclonal proteins in their serum or urine; in 85% of these patients, Ig synthesis has been demonstrated by cytoplasmic M-proteins within their plasma cells. Complete blood count, serum protein electrophoresis, immunofixation electrophoresis, sFLC assay, and quantitative Ig levels should all be included in primary laboratory investigations, followed by bone marrow studies. Clinical signs, normal serum and urine protein electrophoresis, immuno-electrophoresis, and presence of 10% monoclonal bone marrow plasma cells are used to evaluate the diagnosis of NSMM.[8] The International Myeloma Working Group defines NSMM as the absence of M protein in serum or urine, bone marrow plasmacytosis, and associated organ or tissue impairment. Our patient met the criteria based on the findings of a bone marrow biopsy, which revealed the presence of 45% atypical plasma cells. Her IHC indicated positivity for CD138 and lambda light chain markers and no M- band was seen on SPE.

The precise clinical characteristics and prognosis of disease are unclear due to the low incidence of NSMM and patients's ineligibility for clinical trials. The patient's overall health, prognosis, and suitability for hematopoietic stem cell transplantation will all influence the course of treatment.[9]

Conclusion

In this case, the patient developed PE during the disease course emphasizing the rarity of PE in NSMM. Mostly PE is attributed to infective causes, but the presence of myeloma cells in a PE along with the involvement of bone marrow is a very rare scenario that emphasizes the importance of detailed evaluation of such patients by both clinicians and pathologists.

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