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Case Series

Plasma cell rich effusions and its concurrence with plasma cell neoplasms- A case series

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Abstract

Plasma cell neoplasms are clonal hematologic malignancies that represent 10% of all hematologic neoplasms. Incidence increases with advancing age and early diagnosis is rewarding due to great treatment amenability. Diagnosis of this malignancy requires a combination of clinical, biochemical, immunoelectrophoretic and bone marrow findings each with its due. Plasma cell rich effusions that do not form a part of diagnostic algorithm was identified as an incidental finding in 1 case of suspected plasma cell neoplasm and another confirmed case of plasma cell leukemia at our center is being described in this article. The value of plasma cell rich effusion as an associated finding in aggressive cases of plasma cell myeloma is being described here in correlation with the available literature. Immunocytochemical workup for kappa and lambda light chains could not identify e restriction and proved inconclusive in both the cases. The article aims to add to the existing literature on the same topic.

Keywords: Plasma cell rich effusion, Kappa and lambda, Plasma cell neoplasms.

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1. Introduction

Plasma cell neoplasm encompasses a spectrum of hematological neoplasms predominantly affecting elderly with prevalence increasing with age among plasma cell neoplasms, the most common entity is plasma cell myeloma and constitutes 10% of the category of hematological neoplasms.[1]

The entity of plasma cell myeloma was first described by Dr. Samuel Solly (London) in a patient with bone pain, fractures and proteinuria in 1844.[2] Henry Bence Jones identifies the unusual and heat precipitated protein in 1847 excreted in the urine of the patients of this condition that was later named in honour of its discoverer; it was in the late 1800s and early 1900s, the neoplastic cells of this entity were identified plasma cells.[3] The related conditions of MGUS, plasmacytoma, smouldering myeloma were discovered around the time of 1950s. In the decade of 1980s to 1990s newer diagnostic techniques evolved (like SPEP, immunofixation, bone marrow biopsy).

The criteria established by the International Myeloma Working Group (IMWG) for multiple myeloma and related plasma cell neoplasms are the corner stone for diagnosis of the plasma cell neoplasms which was last updated in 2014. The updated criteria has brought forth myeloma defining events, a major drift from the earlier classification and has made radical changes in the diagnosis of this entity of plasma cell myeloma. If any of these events like 60% or more plasma cells in bone marrow biopsy, serum involved / uninvolved free light chain ratio 100 or greater and more than one focal lesion on MRI at least 5 mm in size would point to a diagnosis of multiple myeloma.[4] The diagnosis of this entity requires a combination of serological, radiological, immunoelectrophoretic and bone marrow findings. If all of the essential criteria are not fulfilled, then the diagnosis may

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get redirected to another entity of the diagnostic spectrum of plasma cell neoplasms like monoclonal gammopathy of undetermined significance. M protein spike in immunoelectrophoresis is a mandatory criterion for the diagnosis and if not present, then diagnosis becomes non secretory myeloma.

Effusions of any serous cavity is not a diagnostic criterion of plasma cell myeloma and is a very rare occurrence according to published literature.[5] At our centre, we have reported two cases of plasma cell rich pleural effusion; one being a case of suspected plasma cell myeloma and another of plasma cell leukemia at our centre.

2. Case Presentations

The first case is of a 76 year old female who presented with breathlessness of acute onset which upon evaluation was due to a right sided pleural effusion. The effusion was tapped and sent for cytology; the patient had a significant history of suspicion of plasma cell myeloma and was evaluated for the same; the patient had a suspicious m spike on immunoelectrophoresis but the bone marrow examination could not be done as the patient expired in a few days. A previous bone marrow done 2 years before was inconclusive.

The second case is of a 63 year old female who was a diagnosed case of plasma cell leukemia and was on chemotherapy with drugs bortezomib, lenalidomide, daratumumab (bld regimen)[6] then she was started on injection carfilzomib as the chemotherapy drug. Following chemotherapy patient presented with breathlesness and on examination had right sided moderate to severe pleural effusion that was tapped and sent for cytological examination.

Cytological analysis of pleural fluid from the first patient showed a plasma cell rich effusion; immunocytochemical test on the cell block preparation yielded positivity for CD138 but kappa and lambda staining was inconclusive due to low cellularity effusion fluid cytology of the second patient showed a plasma cell rich effusion with plasmablasts; immunocytochemistry showed strong diffuse CD138 positivity but kappa and lambda in this case also came out inconclusive.

3. Diagnosis

The first case was given a diagnosis of plasma cell rich effusion and the second one was given a diagnosis of plasma cell rich effusion with plasmablasts consistent with myelomatous effusion.

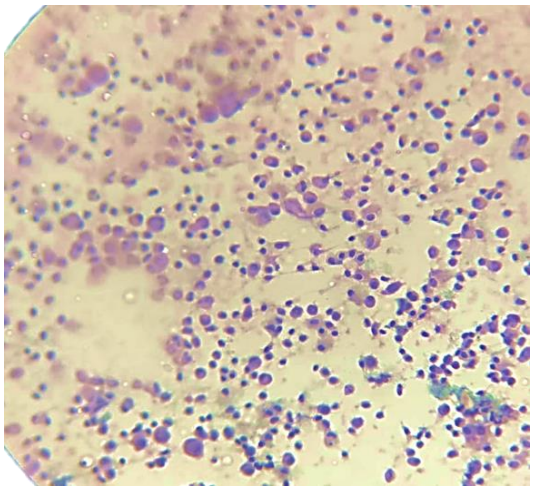

Figure 1: Case 1- High power image of plasma cell rich effusion (20x)

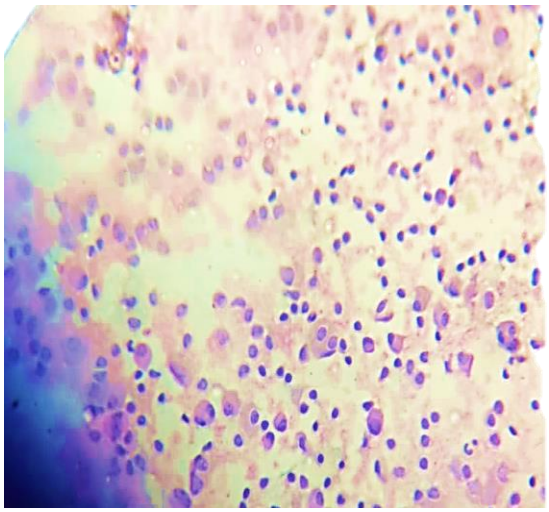

Figure 2: Case 1- High power image with binucleate forms (40x)

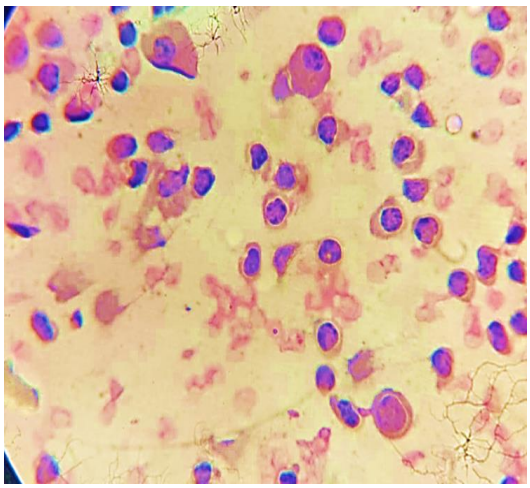

Figure 3: Case 1- Oil immersion image of effusion (100x)

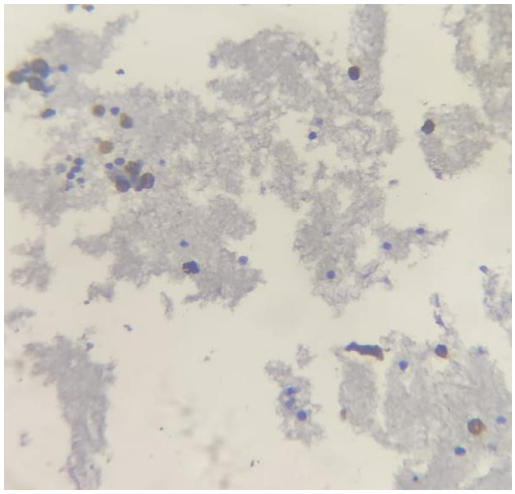

Figure 4: Case 1- IHC image of cd138 showing scattered cell positivity

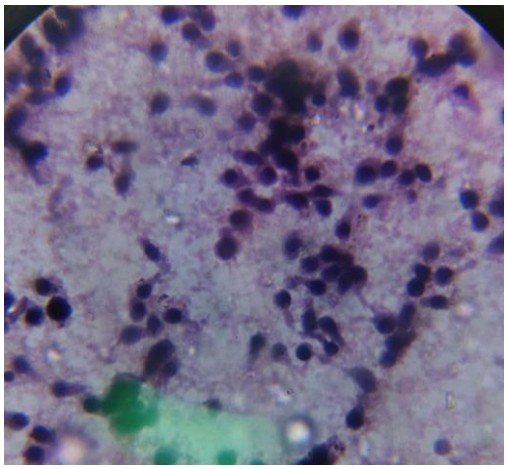

Figure 5: Case 2- 20x image showing plasma cells

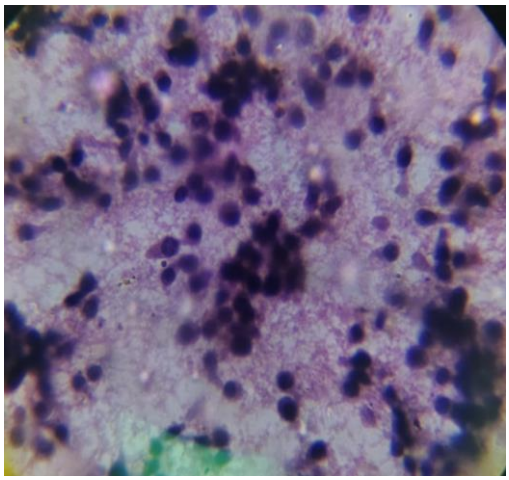

Figure 6: Case 2- 40x image

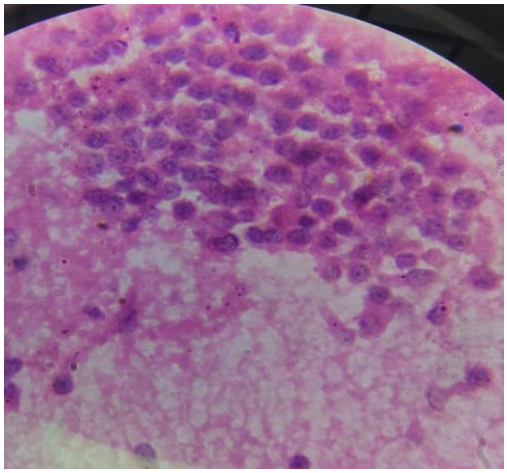

Figure 7: Cell block image 40x image showing plasma cells (Occasional Plasmablasts)

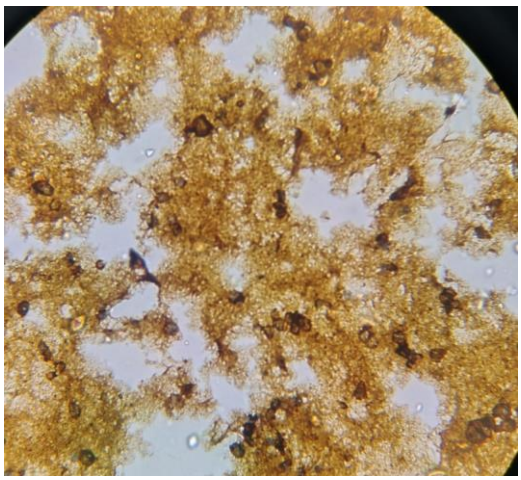

Figure 8: Case 2- CD138 ihc at high power

4. Discussion

Both the above patients had an unfavourable prognosis.[6] The first patient expired within a week of the diagnosis of plasma cell rich effusion while the second one expired after 3 months while undergoing the therapeutic chemotherapy regimen. Plasma cell rich effusion in known or suspected cases of plasma cell neoplasms is a very rare event and is directly correlated with an adverse prognosis and aggressive disease. It is hypothesized that the neoplastic cells can reach the pleural spaces by direct infiltration or can spread from adjacent skeletal lesions or via lymphatic obstruction; in any of the above mentioned cases, the disease becomes aggressive or advanced.[7]

The effusions in myeloma patients can also be non-myelomatous, related to systemic effects or complications.[8] They can even be unrelated to plasma cell myeloma and may represent reactive effusions due to infectious causes such as tuberculosis, viral, fungal, or parasitic infections, or autoimmune conditions like SLE, post-vaccination reactions, chronic inflammatory diseases, or even post-surgical effusions.

The effusion in the second case was diagnosed as myelomatous effusion (owing to the presence of plasmablasts), whereas the first case was diagnosed as plasma cell–rich effusion, since clonality could not be established.

5. Conclusion

Plasma cell myeloma is a hematological neoplasm that constitutes a major proportion of hematological neoplasms reported from our center and possibly the most prevalent in the local community from available data. The concurrent occurrence of plasma cell rich effusion was an incidental finding in the 2 cases discussed here. Myelomatous effusion is associated with aggressive disease and poor prognosis. Early diagnosis is critical and life-saving in most cases of plasma cell myeloma owing to its treatment amenability and response to chemotherapy. The association with poor prognosis was evident in both of our cases and is in support of the available literature regarding the same.

6. Source of Funding

None.

7. Conflict of Interest

None.

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