

Synchronous presentation of mantle cell lymphoma and plasma cell dyscrasia: A case report

Galani K.S., Gadage V.S., Deshmukh Mahesh, Menon S., Gujral S.

ABSTRACT

Introduction: The coexistence of non-Hodgkin lymphomas (NHL) with plasmacytoma or multiple myeloma (MM) at same anatomical location is reported only as a few case reports. **Case Report:** We report a case of 63-year-old male who presented with nasal obstruction and bilateral cervical and right axillary lymphadenopathy of three months duration. Biopsy from nasal mass revealed diffuse infiltration by atypical lymphoid cells which expressed LCA, CD20, CD5 and cyclin D1 on immunohistochemistry. A diagnosis of extranodal mantle cell lymphoma was rendered. A staging bone marrow aspirate revealed an increase in atypical lymphoid cells and morphologically atypical plasma cells, consisting 15% and 20% of the nucleated non-erythroid cells respectively. The bone marrow biopsy showed small paratrabeular and interstitial nodular aggregates of atypical lymphoid cells which were highlighted by CD20, CD5 and cyclin D1. In addition, CD138 highlighted an interstitial increase in plasma cells which also demonstrated an aberrant expression of CD56 and cyclin D1. This proved the coexistence of mantle cell lymphoma and plasma cell dyscrasia, which is

worth reporting. **Conclusion:** This study clearly demonstrates the unique existence of mantle cell lymphoma (MCL) with plasma cell dyscrasia based on careful morphologic evaluation and a panel of immunostains. A molecular approach to analyze the clonal relationship between the two populations is needed to exclude a composite lymphoma and allow for appropriate treatment.

Keywords: Cyclin D1, Mantle cell lymphoma, Plasma cell dyscrasia, Synchronous

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INTRODUCTION

Clonal plasma cell component is well described in some non-Hodgkin lymphomas (NHL), including marginal zone lymphomas (MZL) and lymphoplasmacytic lymphoma (LPL). It is very rarely seen in other NHL like chronic lymphocytic leukemia (CLL), follicular lymphoma (FL) and mantle cell lymphoma (MCL). The coexistence of NHL with plasmacytoma or multiple myeloma (MM) at same anatomical location is reported only as a few case reports. Occasional case report also mentions metachronous presentation of MCL in a case of MM. Herein, we report a case of MCL with plasma cell dyscrasia (PCD), on bone marrow in a 65-year-old male [1–6].

CASE REPORT

A 65-year-old male complained of neck swelling since last six months, underwent cervical lymph node fine needle aspiration (FNA) which revealed granulomatous lymphadenitis. Based on this, he was treated with antitubercular treatment (ATT) at his local place for three months. Due to persistence of cervical swelling and nasal obstruction, he was referred to our institute. On examination, he had bilateral cervical and axillary lymphadenopathy without hepatosplenomegaly. His complete blood counts (CBC) revealed anemia (hemoglobin: 8 g/dl) with normal total leukocyte ($7.3 \times 10^9/L$) and platelet count $372 \times 10^9/L$. Liver and renal function tests were normal. Serum LDH 232U/l; (normal range: 100–180 U/L) and β_2 microglobulin 9.85 mg/L; (normal range: 0.83–1.15 mg/L) were elevated. Computed tomography (CT) scan of paranasal sinuses showed large lobulated homogeneously enhancing soft tissue density in the nasopharynx and right upper oropharynx, extending across the midline up to the posterior choana and left soft palate. It also revealed multiple soft tissue densities in bilateral orbits. Ultrasonography (USG) of abdomen and pelvis revealed mesenteric, peripancreatic and paraaortic lymphadenopathy. A nasal mass biopsy was performed which showed diffuse infiltration by atypical lymphoid cells expressing LCA, CD20, CD5 and cyclin D1. Mib1 proliferation index was 40%. A diagnosis of extranodal mantle cell lymphoma was rendered. A staging bone marrow aspirate (BMA) and biopsy (Figure 1) revealed mildly hypercellular particles with trilineage hematopoiesis. Few atypical lymphocytes were seen along with an increase in morphologically abnormal plasma cells. Nucleated non-erythroid cell count done on BMA showed 20% abnormal plasma cells and 15% atypical lymphocytes. Background red blood cells (RBC) did not show any rouleaux formation. Plasma cells showed ballooning of cytoplasm with intracytoplasmic grayish blue inclusions (? immunoglobulin collection) and an eccentric nucleus (Figure 2A). Occasional binucleated cells were also noted. Bone marrow sample was not sent for immunophenotyping by flow cytometry (FCM) and thus atypical lymphoid cells were not characterized on aspirate. The bone marrow biopsy showed small paratrabecular and interstitial nodular aggregates of atypical lymphoid cells as well as an interstitial increase in plasma cells (Figure 1A). On immunohistochemistry (IHC), aggregates of lymphoid cells were highlighted by CD20, CD5 and cyclin D1 (Figure 1B–D). On careful examination, cyclin D1 was also positive in plasma cells (Figure 2D). These plasma cells were highlighted with CD138, CD56 and showed lambda light chain restriction (Figure 2B–C). Aberrant CD56 expression proved the neoplastic nature and lambda light chain restriction (Figure 2E) proved the clonal nature of plasma cell component in this case. Mantle cell lymphoma component did not reveal either lambda or kappa light

chain restriction (Figure 2F). Additional workup for plasma cell dyscrasia was done. Skeletal survey did not reveal any lytic lesions. Serum globulin levels (3.8 g/dl) were within normal range 2.8–4.3 g/dl. Serum electrophoresis and immunoprotein studies were not performed. The final diagnosis of Mantle cell lymphoma with Plasma cell dyscrasia was offered. Patient was staged IV BEx (Ann Arbor staging for NHL) and started on R-CEOP (rituximab, cyclophosphamide, vincristine, prednisolone) protocol. On his recent follow-up after three months, he is clinically stable. However, there is persistent anemia with hemoglobin of 8.4 g/dl.

DISCUSSION

The co-existence of mantle cell lymphoma and plasma cell dyscrasia involving the bone marrow is a very rare occurrence. The index case showed presence of extranodal mantle cell lymphoma involving nasopharynx without the plasma cell component at that site. However, both the components in the bone marrow were morphologically and immunohistochemically distinct. Morphologically abnormal plasma cells with ballooning of cytoplasm and aberrant expression of cyclin D1 and CD56 along with lambda light chain restriction clinched the neoplastic and clonal nature of plasma cells. Though the presence of cyclin D1 both in MCL and plasma cells does not prove their different clonal nature, the expression of CD56 only in plasma cells proves their neoplastic nature and supports the distinct nature of synchronous neoplasms as against the clonal differentiation of MCL with clonal plasma cell differentiation. Unfortunately, bone marrow aspirate was not submitted for cytogenetic studies in index case.

Very few cases of MCL with plasmacytoma have been reported in literature previously [1, 5]. These patients were usually more than 60 years of age, with upper respiratory tract being the most common site of involvement as in the index case [1, 3, 5].

Mantle cell lymphoma (MCL) with clonal plasma cell differentiation/component, within the same tissue biopsies, however, has been described in very few studies [2, 7]. Young et al. reported a case of a nodal MCL with clonal plasma cell differentiation based on kappa-light chain restriction only in plasma cell component and not in MCL, similar to index case [2]. However, in his study demonstration of t(11;14) by FISH (fluorescent in situ hybridization) in both the components suggested that the clonal plasma cells are a part of MCL rather than a second lymphoproliferative disease [2]. Composite presentation of immunophenotypically distinct MCL and extramedullary plasmacytoma in a single anatomical site without plasma cell component in bone marrow has been reported in literature as some case studies [1, 5]. It is important to evaluate this plasma cell component for clonality and also its neoplastic nature using a wide range of IHC panel. Demonstration of cyclin D1 in both

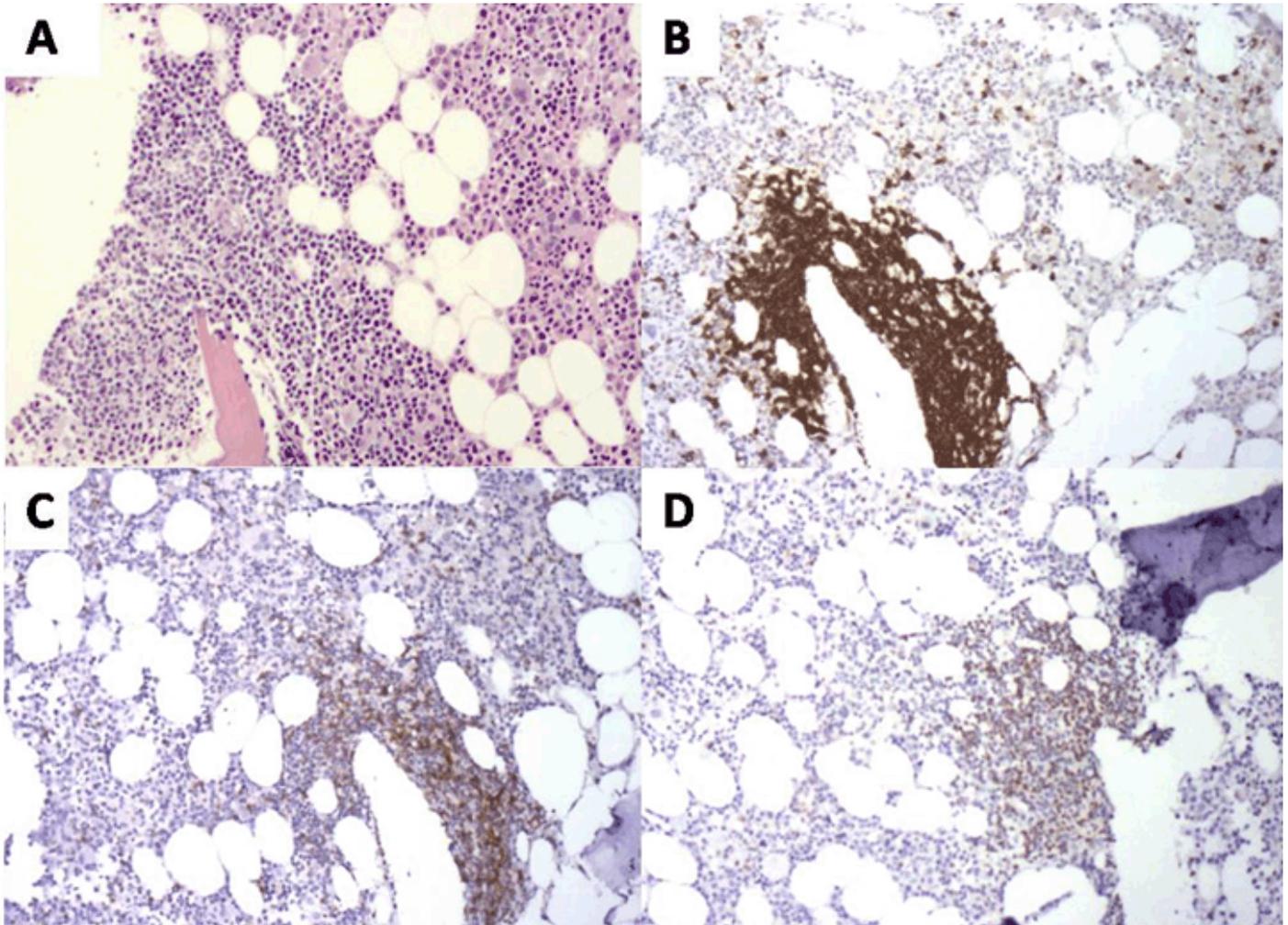


Figure 1: (A) Bone marrow biopsy showing paratrabecular lymphoid cell aggregates with interstitial increase in plasma cells (H&E); (B) CD20, (C) CD5, (D) cyclin D1 highlights paratrabecular lymphoid cell aggregates of mantle cell lymphoma.

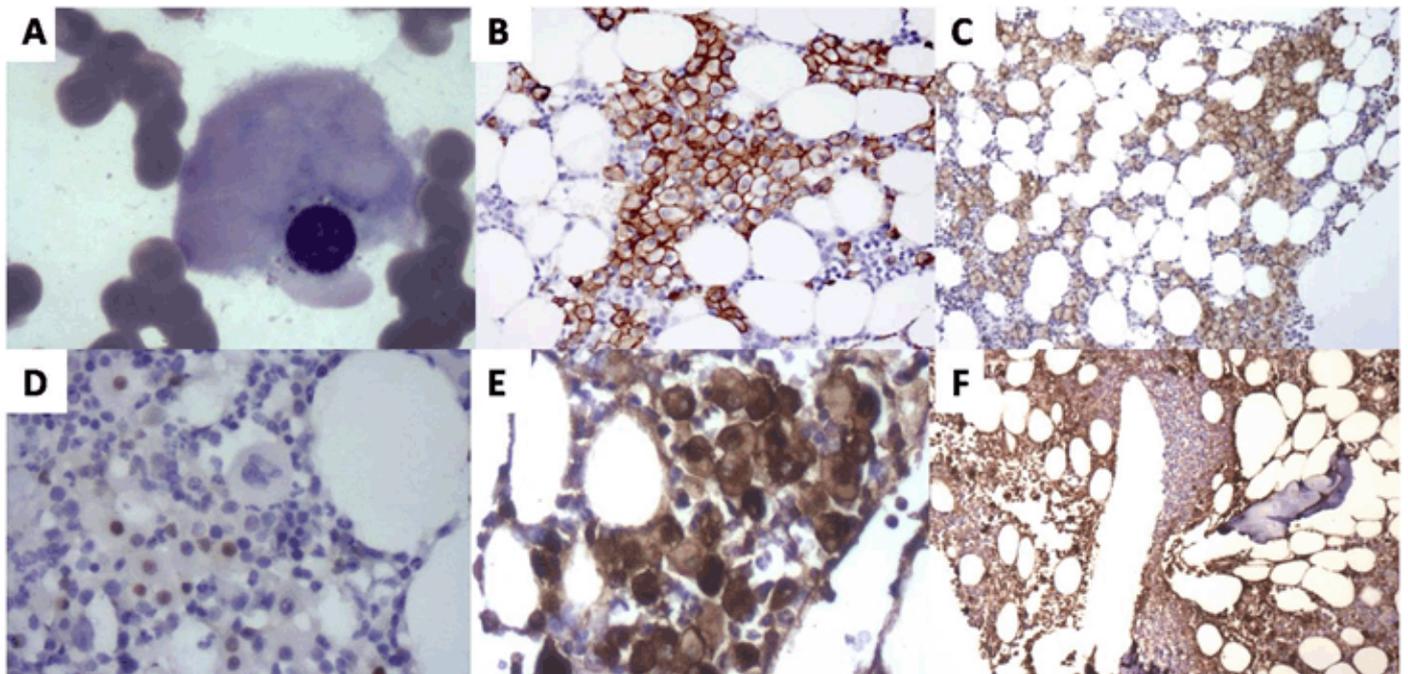


Figure 2: (A) Plasma cell showing intracytoplasmic collection of immunoglobulins on bone marrow aspirate; B to F: Bone marrow biopsy IHC: (B) CD138, (C) CD56, (D) Cyclin D1 highlights neoplastic plasma cells; lambda light restriction is seen in plasma cells (E) but not in paratrabecular aggregates of mantle cell lymphoma (F).

MCL and PCD may prove the clonal nature of the disease but not the neoplastic nature of plasma cells. Expression of cyclin D1 is always aberrant in hematolymphoid neoplasms, including MCL, multiple myeloma (MM) and hairy cell leukemia (weak expression) [3, 6, 8]. Almost 100% cases of MCL express cyclin D1 (cyclin D1 negative MCL-very rare) and cyclin D1 positivity correlates with t(11;14) [6, 9]. However, only 3–50% cases of MM show cyclin D1 expression and this is associated either with t(11;14) in 3–14% [3] or with extra copies of chromosome 11 in 16% [3, 6]. CD56 expression defines the neoplastic nature of plasma cells as against normal plasma cells and is aberrantly expressed in 67–79% cases of plasma cell myeloma [3]. However, unlike in the index case, none of the studies in literature discussing the coexistence of MCL and plasmacytoma have used CD56 by immunohistochemistry (IHC) to demonstrate the neoplastic nature of plasma cells [1, 2, 5, 7]. In this case, ballooning of plasma cells on morphology suggested intracytoplasmic accumulation of immunoglobulins, which was confirmed by IHC and it indicated the non-secretory nature of MM which was further reinforced by the normal globulin levels. However serum electrophoresis to demonstrate the 'M' band was lacking in the index case.

Plasma cell dyscrasia being an incidental and asymptomatic finding in this case, the primary aim of treatment was chemotherapy for MCL. However, there are certain drugs like bortezomib (proteasome inhibitor), lenalidomide/thalidomide (immunomodulating drugs) which have a proven action in both MCL and PCD individually and thus may be used for such rare coexistence [6].

CONCLUSION

In conclusion, our study clearly demonstrates the unique existence of mantle cell lymphoma (MCL) with plasma cell dyscrasia based on careful morphologic evaluation and a panel of immunostains. A molecular approach to analyze the clonal relationship between the two populations is needed to exclude a composite lymphoma and allow for appropriate treatment. At present time, an array of genetic and molecular investigations is under way to explain the relationship between the presence of t(11;14)(q13;q32) and the origin of various hematologic neoplasms. We believe that the present case might offer additional insights to this fascinating topic of composite neoplasms which in turn may yield new avenues for both MCL and myeloma therapy.

Author Contributions

Komal S. Galani – Acquisition of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Vijaya S. Gadage – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Mahesh K. Deshmukh – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Santosh Menon – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Sumeet Gujral – Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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