Case Report

Blastoid Variant of Mantle Cell Lymphoma with Extranodal Presentation and Aberrant CD10 Expression

Abstract

Mantle cell lymphoma (MCL) constitutes 3%–10% of non-Hodgkin lymphoma and is characterized by *t* (11:14)(q13;q32). The common presentation is generalized lymphadenopathy with weight loss, infrequently night sweats, and fever. Among histological subtypes of MCL, the blastoid variant of MCL constitutes 10%–15% of all the cases. It is challenging to diagnose the blastoid variant of MCL based on its morphology alone as it mimics large B-cell lymphoma. Hence, the immunophenotyping and molecular studies aid in its correct diagnosis. We report an elderly man diagnosed with blastoid variant MCL. He presented with disseminated soft-tissue and subcutaneous nodules, and showed aberrant CD10 expression. Presentation of the extranodal site and aberrant CD10 expressions carries an overall poor prognosis. CD10-positive MCL can be mistaken for large B-cell lymphoma.

Keywords: *Aberrant CD10, mantle cell lymphoma, soft-tissue nodules*

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Introduction

Mantle cell lymphoma (MCL) constitutes 3%–10% of mature B-cell non-Hodgkin lymphoma. It has a male preponderance, usually occurs in the 6th–7th decades and most patients present with stage III or IV. MCL is known to have an extranodal presentation. The *IGH: CCND1* fusion is hallmark of MCL is characterized by *t* (11;14)(q13;q32) and immunohistochemistry (IHC) showing by CD20+, CD5+ CD23−, and Cyclin D1 positivity. Fluorescence *in situ* hybridization (FISH)/molecular studies is currently considered gold standard for its definitive diagnosis.[1] The morphologic variants of MCL include blastoid, pleomorphic, small cell, and marginal-zone-like variants. It is challenging to diagnose the blastoid variant of MCL based on its morphology alone as it mimics large B-cell lymphoma.[1] The majority of cases of MCL are thought to be caused by the naive pregerminal center B-cell, which characteristically shows positivity for Cyclin D1, pan B-cell marker, CD5, and BCl2. Neither CD23 nor follicular center cell-associated antigens such as CD10 nor BCL6 are present. We present a case of blastoid MCL, which predominantly presented with soft-tissue involvement and also showed CD10 aberrant expression.

Case Report

A 69-year-old male presented to the fine-needle aspiration cytology (FNAC) clinic with multiple swellings over the anterior chest wall, bilateral infrascapular region, and left lower eyelid for a 2-month duration. FNAC from the chest wall and infra-scapular site showed sheets of monomorphic intermediate-sized cells with dispersed chromatin, inconspicuous nucleoli, and scant cytoplasm. Immunocytochemistry showed diffuse positivity for CD79a; overall features were of non-Hodgkin lymphoma of B-cell origin. Peripheral smear and bone marrow were not involved. Aspirate obtained during fine-needle aspiration was processed using flow cytometry and it expressed strong positivity for pan B-cell marker (CD19, CD20, CD22, and CD79a), and weak positivity for CD5, CD10, IgD, and lambda restriction. Negativity for nTdT and CD23 favored the possibility of MCL with aberrant CD10 expression [Figure 1]. Excision biopsy from infrascapular swelling shows diffuse sheets of large-sized atypical lymphoid cells with scant cytoplasm, irregular nuclear membrane, and coarse chromatin. IHC showed positivity for CD20, CD10, and CD5 and negative for CD3 and CD23. The Ki67 labeling index was 80%. Features were consistent with a

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high-grade B-cell lymphoma. With a differential diagnosis of large cell lymphoma such as diffuse large B-cell lymphoma (DLBCL), high-grade follicular lymphoma, and blastoid MCL, a further panel of IHC done showed positivity for Cyclin D1, Bcl6 and negative for MUM1, c-MYC, and TdT [Figure 2]. These findings were consistent with a diagnosis of a blastoid variant of mantle cells with CD10 aberrant expression. FISH done subsequently showed positivity for *t* (11:14)(q13;q32). A postbiopsy fluorodeoxyglucose-positron emission tomography scan revealed abnormal uptake seen in multiple cervical regions and extensive lymphadenopathy of the thoracic,

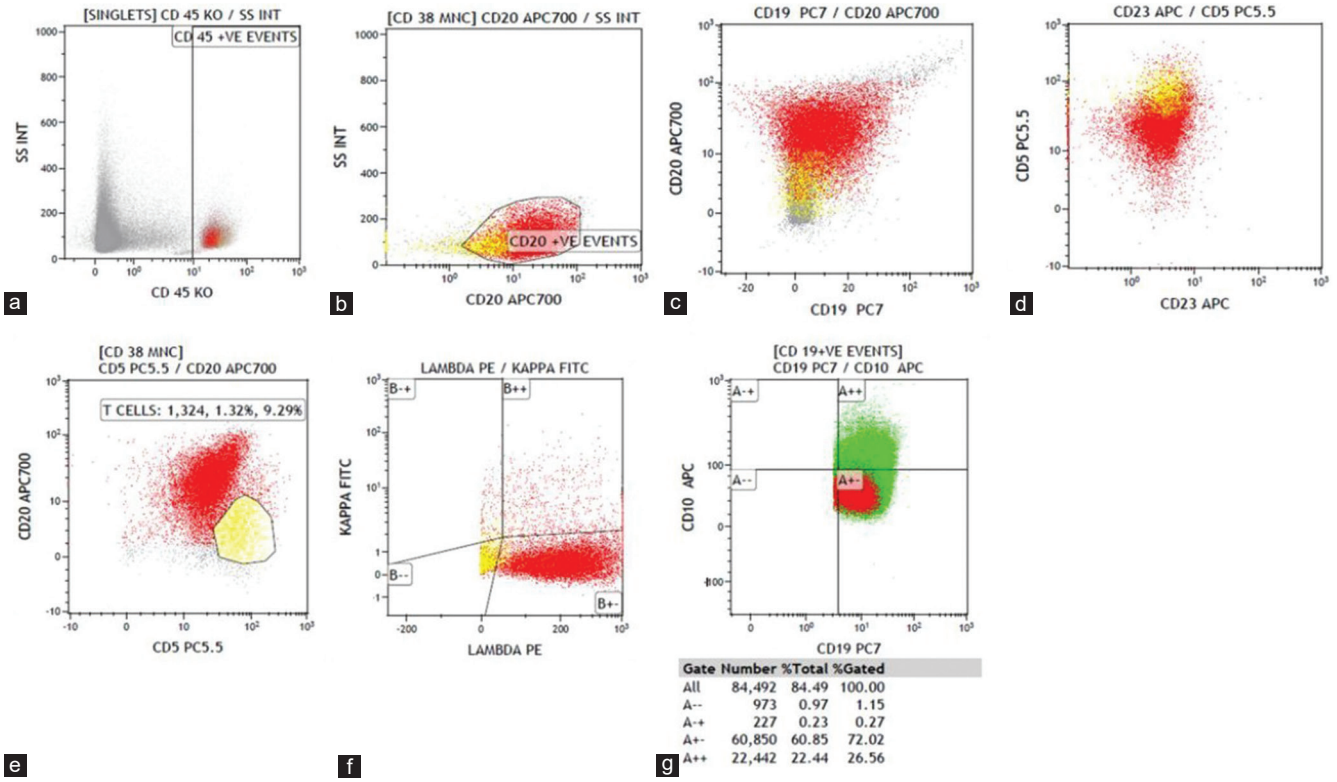

Figure 1: Flow cytometry showing CD20+, CD19+, CD5+, CD23−, CD10+ (weak), lambda restricted. (a) CD45 versus SSC, (b) CD20 versus SSC, (c) CD19 versus CD20, (d) CD5 versus CD23, (e) CD5 versus CD20, (f) Kappa versus lambda, (g) CD19 versus CD10

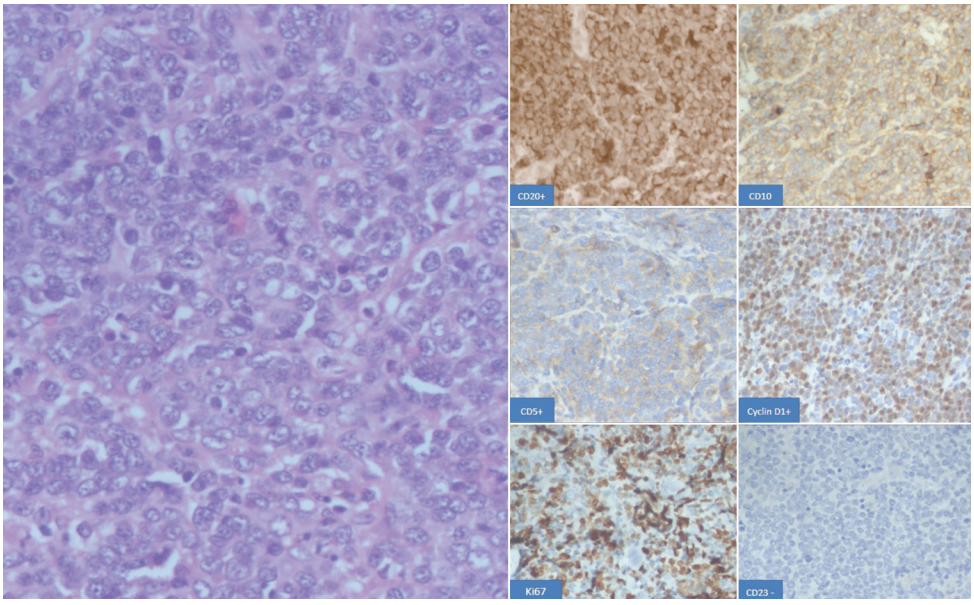

Figure 2: Biopsy from chest wall swelling shows diffuse sheets of large atypical lymphoid cells with conspicuous nucleoli (H and E, ×400), IHC: The neoplastic cells are strongly positive for CD20, CD5, Cyclin D1, CD10 with high Ki 67, and CD23 negative (IHC, ×400)

retroperitoneal, abdominal, and bilateral lungs along with the subcutaneous and muscular location. The patient expired after two cycles of bendamustine + rituximab chemotherapy.

Discussion

The blastoid variant of MCL constitutes about 10%–20% of all MCL cases and typically presents as the advanced stage with the worst prognosis.[2] This lymphoma not only primarily affects lymph nodes but is known for its common extranodal involvement, which commonly includes the spleen, bone marrow, liver, gastrointestinal tract (GIT), and Waldeyer ring.[3]

MCL usually presents with B symptoms, associated with lymphoma. Our case primarily presented as multiple soft-tissue/subcutaneous masses is extremely uncommon; however, they do form part of the clinical presentation of MCL in old age with B symptoms. This patient did not show any skin involvement grossly. Among high-grade B lymphomas, 40% of DLBCL can present as extranodal and those are termed as primary extranodal DLBCL, which includes GIT, central nervous system, testis, and breast.[4,5] Followed by this is MCL, which can present as skin, scalp, earlobes, extensive cutaneous, soft tissue, and ocular extranodal involvement.[2,4,6] Furthermore, there are reports showing MCL cases with skin involvement as nodules, erythematous (infiltrated) plaques, a macular rash, papules, and petechiae, ecchymosis.[5]

Most of the MCL cases are positive for pan-B-cell antigens, CD5, BCL2, and Cyclin D1, and negative for CD23 and CD10. Most of the MCL shows over-expression in Cyclin D1, which is a surrogate marker for *t* (11;14)(q13;q32)/*IG::CCND1* detected by conventional cytogenetic or FISH studies which is currently considered as gold standard method in diagnosing MCL cases. However, there are 5% Cyclin D1-negative MCL cases, a small subset which shows *t*(11;14)(q13;q32)/*IG::CCND2* translocation.[7] SOX11, a neuronal transcription factor, was identified as a very specific and useful diagnostic marker for MCL, with high specificity, including cyclin D1-negative cases.[8,9]

However, in our case, CD10 and BCL6 are positive, which is a follicular center cell-associated antigen. Often, CD10 is useful in distinguishing MCL from other CD10+ B cell lymphomas.[1] Diagnosis of MCL requires both morphological and immunophenotypic analysis.

The CD10 (110-kd transmembrane glycoprotein) is expressed in normal germinal center B-cells and early lymphoid progenitors. It is often expressed in follicular lymphoma, Burkitt lymphoma, lymphoblastic lymphoma/leukemia, and a subset of DLBCL, because of its follicular center cell origin. However, CD10 expression has been seen rarely in other nonfollicular center B cells, such as extranodal marginal zone lymphoma and chronic lymphocytic leukemia/small lymphocytic lymphoma.[10,11]

Dong *et al*. also mentioned that co-expression of CD5 and CD10 was observed in lymphomas with large cell morphology, and the differential diagnosis considered in such scenarios were acute leukemia, large cell lymphoma, Burkitt lymphoma, and blastoid MCL.[10]

The literature on MCL with aberrant CD10 positivity is limited. Ziemba *et al*. retrospectively studied 37 MCL cases for 7 years, in which two patients showed CD10 positivity at recurrence in a previously diagnosed CD10-negative MCL along with increased Ki67 >90% and no bone marrow or nodal involvement.[12] This study suggests that recurrent MCL with CD10 positivity and a high Ki-67 index indicate more aggressive disease progression.

Conclusion

The blastoid variant of MCL is rare to present as a disseminated disease involving extranodal soft tissue/subcutaneous tissues. The Blastoid variant of MCL with CD10 aberrancy and high Ki67 labeling index carries the worst prognosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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