

Blastoid Variant of Mantle Cell Lymphoma- a Rare Case Report

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Abstract Mantle cell lymphoma is now recognised as a rare but distinct entity in the revised WHO classification. It is now well recognised that MCL represent a broad spectrum of different histopathological subtypes. The term blastic or blastoid variant is generally used to describe cases with a homogeneous population of cells displaying lymphoblastic morphology. The blastic form of MCL may be difficult to diagnose however immunophenotyping and molecular analysis show typical mantle cell lymphoma pattern. We present a case of 30 year old male presenting with inguinal mass which was diagnosed as blastic transformation of mantle cell lymphoma based on routine histopathology and immunohistochemistry.

Keywords: mantle cell lymphoma, blastoid variant, immunohistochemistry

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1. Introduction

Since its original description by Lennert in 1964, a variety of names were used for this neoplasm before the recent general acceptance of the term mantle cell lymphoma (MCL). [1] MCL is a neoplastic lymphoid proliferation related to the naive pregerminal center cells localized in the primary follicle and to the mature mantle zone of the secondary lymphoid follicles. Mantle cell lymphoma is a rare entity, representing from 3-10% of non-Hodgkin's lymphomas. It occurs more frequently in middle-aged to older males (male:female ratio = 2- 7:1). [2] It is listed among peripheral B-cell lymphomas with *CCND1* translocation.

It is now well recognised that MCL represent a broad spectrum of different histopathological subtypes. Blastoid variants of MCL have been described and represent approximately 10% to 15% of all cases in various studies. In the World Health Organization (WHO) classification, two blastoid variants of MCL are recognized: classic and pleomorphic. [3] Classic blastoid variant is composed of cells that can closely resemble lymphoblasts, starry-sky pattern can be present. In the pleomorphic blastoid variant, the neoplasm is composed of a heterogeneous population of cells including large cleaved or round cells that can have prominent nucleoli. Mitotic figures are also increased and this variant can resemble large-cell lymphoma. Classic blastoid variant MCL often presents de novo. In contrast, the pleomorphic blastoid variant MCL can occur in patients with a history of MCL or in patients with another simultaneously detected site involved by typical MCL.

2. Case Presentation

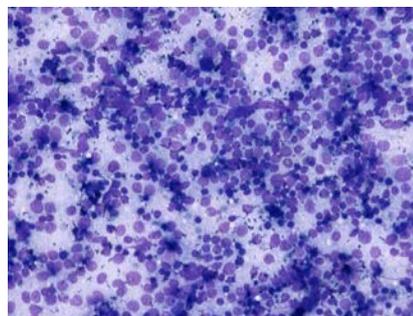


Figure 1. FNA suggestive of lymphoproliferative disorder (200x, Geimsa)

We present a case of 30 year old male patient who presented with 3 month history of bilateral inguinal lymphadenopathy which was more on the right side along with complaints of generalized weakness. On examination, bilateral inguinal lymph nodes ranged in size from 1.5-2.0 cm in size and were soft to rubbery, nontender and mobile on palpation. FNA done from the largest lymphnode suggested a lymphoproliferative disorder (Figure 1). Complete blood count, bone marrow aspiration and trephine biopsy showed normal studies. However flow cytometry and FISH studies were not performed. Excision biopsy of right inguinal lymph node revealed sheets of large lymphoid cells with moderately abundant eosinophilic cytoplasm, stippled nuclear chromatin and small membrane bound nucleoli (Figure 2). On immunohistochemistry, these cells were Cyclin D1, CD 10, CD 20, bcl-2, positive (Figure 3 A, Figure 3 B, Figure

3 C) and Tdt, CD 3, CD5 negative. A final diagnosis of non-Hodgkin lymphoma, blastic transformation of mantle cell lymphoma was made.

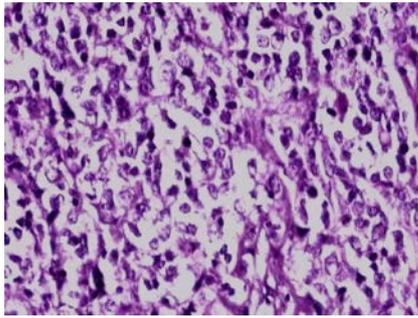


Figure 2. Medium to large lymphoid cells with moderately abundant eosinophilic cytoplasm, stippled nuclear chromatin and small membrane bound nucleoli (400x, H&E)

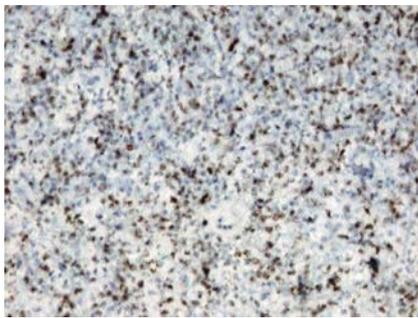


Figure 3 A. Blast like cells are cyclin D1 positive (200x, IHC)

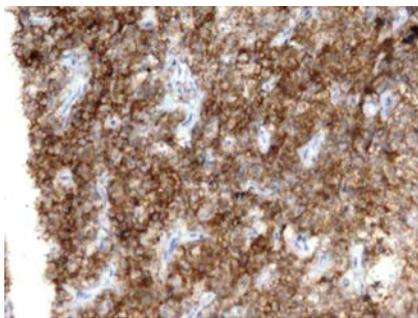


Figure 3 B. Blast like cells are CD10 positive (400x, IHC)

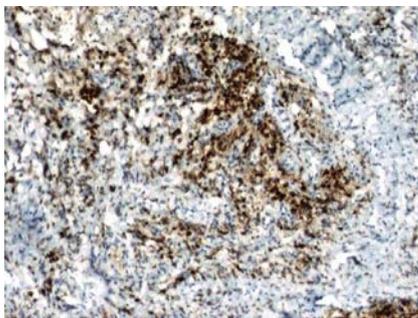


Figure 3 C. Blast like cells are Bcl-2 positive (100x, IHC)

3. Discussion

The median age of MCL patients is the late sixth or seventh decades in many studies. [4] In the Non-Hodgkin's Lymphoma Classification Project study, the median age was 63 years, with an age range of 37 to 82 years. Mantle cell lymphoma is uncommon under the age

of 40 years and rare in patients under the age of 30 years. At the M.D. Anderson Cancer Center, the youngest patient with MCL to date was 24 years old. There is a case report of an 18-year-old girl with blastoid variant MCL.

Systemic (B-type) symptoms occur in 30% to 50% of patients. Weight loss is most common, whereas night sweats and fever are less frequent and patients often have good performance status. Most patients with MCL present with advanced clinical-stage disease (Ann Arbor III–IV). Lymphadenopathy is very common, often generalized, and lymph nodes are typically not bulky and range in size from 2 to 5 cm. [5] Extranodal sites of disease are common in MCL patients, with the gastrointestinal (GI) tract being one of the most common.

Three histological variants are observed: mantle zone, nodular and diffuse. Cells of a typical infiltrate in MCL are small to intermediate-sized with angulated nuclear contours, inconspicuous nucleoli and scant cytoplasm. Epithelioid histiocytes and hyalinized blood vessels are commonly seen.

Blastoid variants of MCL represent approximately 10% to 15% of all cases in various studies. However, the frequency of blastoid variants may be much higher in patients who are followed for long periods of time. Classic blastoid variant MCL is composed of medium-sized cells with evenly dispersed nuclear chromatin and a very high mitotic rate that can mimic pre-B lymphoblastic leukemia/lymphoma (pre-B ALL/LBL). Immunophenotypic studies are very helpful as pre-B ALL/LBL expresses Tdt and is negative for surface Ig and cyclin D1, unlike MCL.

Cytogenetically, nearly all mantle cell lymphomas harbor the t(11;14) translocation. Immunophenotypically, cells are usually CD5 positive and nearly all cases are positive for cyclin D1 overexpression; however, immunophenotypic variants are not uncommon in mantle cell lymphomas. [6] Occasional atypical cases of mantle cell lymphoma may be CD10 positive (8%), CD5 negative (12%) as depicted in our case, CD23 positive (21%), and/or rarely negative for cyclin D1 expression but there appears to be no definite correlation between immunophenotypic variation and mantle cell lymphoma morphologic variants. BCL-6 is almost always negative, although rare cases of blastoid variant MCL can be Bcl-6 positive. In addition, rare cases of MCL, usually blastoid variants, can be CD10+ which was true in the case presented.

MCLs are frequently misdiagnosed, the pleomorphic variant has to be distinguished from diffuse large B cell lymphoma. Clinically, approximately half of diffuse large B-cell lymphoma (DLBCL) patients present with localized disease that can be bulky, unlike MCL. Usually, immunophenotypic and molecular studies are needed to distinguish DLBCL from the pleomorphic blastoid variant MCL. Diffuse large B-cell lymphoma cases are cyclin D1–, usually CD5–, and do not carry the t(11;14). The karyotypes of blastoid variants of MCL tend to be complex. In one study using conventional cytogenetic analysis, Khoury and colleagues (77) showed that 24 of 27 (89%) blastoid MCLs had a complex karyotype (>3 abnormalities).

Pathologic parameters that have been correlated with poorer prognosis include high leukocyte count, high mitotic or proliferation (Ki-67) rates and blastoid variants.

In contrast, a pathologic parameter that has been associated with a better prognosis is a mantle zone pattern.

Patients with MCL have a poor prognosis and are incurable using traditional cyclophosphamide, doxorubin, vincristine, and prednisone (CHOP) chemotherapy; their median overall survival is 2 to 5 years. Patients with blastoid variant MCL are associated with particularly short durations of response after chemotherapy and poorer overall survival [7].

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