

Case Report

MANTLE CELL LYMPHOMA PRESENTING AS LEUKEMIA- A CASE REPORT AND REVIEW OF LITERATURE

Gajendra Singh Rajput¹, *Pooja Gupta¹ and Alpesh Goyal²

¹Department of Pathology, Government Medical College, Kota

²Department of Medicine, AIIMS, New Delhi

*Author for Correspondence

ABSTRACT

Mantle cell lymphoma (MCL) is a type of B-cell lymphoma accounting for six percent of all Non Hodgkin Lymphoma (NHL). It commonly presents with lymphadenopathy and hepatosplenomegaly; a leukemic presentation is however rare. Its clinical course is progressive with poor response to chemotherapy. We describe a case of elderly male who presented with acute leukemia, and on further characterization with immunophenotyping, diagnosed as blastoid MCL.

Keywords: Mantle Cell Lymphoma, Leukemia, Flow Cytometry

INTRODUCTION

Mantle cell lymphoma (MCL) is a rare B cell lymphoma characterized by CD5+ CD23-,cyclin D1+ and t(11;14)(q13;q32). They arise from pre germinal center B lymphocyte present in primary lymphoid follicles and mantle zones of secondary follicles. Most patients present with advanced disease, and with frequent extranodal involvement. It was previously called centrocytic lymphoma or intermediately differentiated lymphoma. Cytologically, there are two main variants-classic and blastoid, of which blastoid form is highly aggressive. Bone marrow involvement is not infrequent and may be nodular, diffuse and paratrabecular. Early recognition of blastoid MCL is required using immunophenotyping and bcl-1 molecular study. Standard chemotherapy gives poor results and an alternative treatment should be proposed.

CASES

A 65 year old male presented to the outpatient department with complaints of fullness and dragging sensation in left upper abdomen associated with fatigue and prostration for four months. There was no history of fever, weight loss, palpable neck masses, jaundice, or altered bowel habits. Examination revealed massive splenomegaly, reaching upto the umbilicus. For further evaluation, patient underwent a number of investigations, the findings of which have been summarised below.

Peripheral Blood Smear (PBS)

PBS revealed bicytopenia with leukocytosis and increased blasts (Table 1). Blasts were 2–3 times the size of small mature lymphocytes with scant agranular cytoplasm, large nucleus with irregular contour, fine chromatin and 1–2 prominent nucleoli.

Bone Marrow Aspiration and Biopsy (Microphotograph 1 and 2)

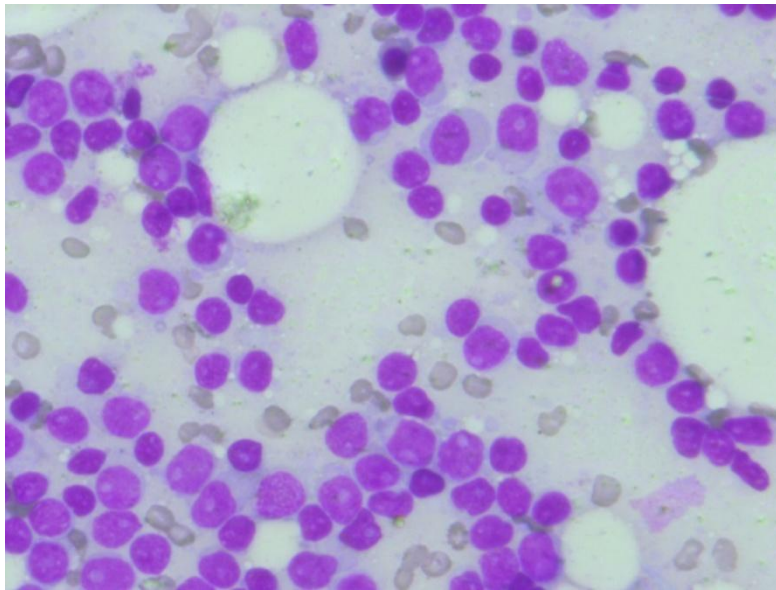
Bone marrow aspirate revealed markedly hypercellular marrow. Myeloid: erythroid ratio was highly raised with almost complete replacement of normal hematopoietic cell by malignant lymphoma cells. Morphologically they were atypical lymphoid cells with predominant population of lymphoid cells with prominent nucleolus. Biopsy showed diffuse effacement of marrow architecture with infiltration by atypical lymphoid cells.

Immunophenotyping

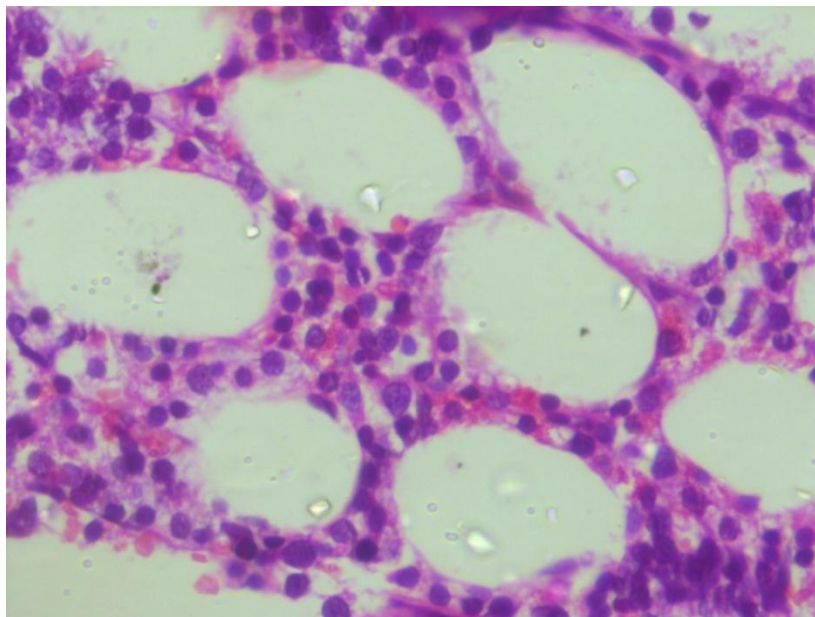
Flow cytometry studies of bone marrow aspirate revealed large population of cells showing expression of pan leukocyte marker CD45, CD19, CD20, cyclinD1 and CD5. It was however, negative for CD23, CD10 and terminal deoxy nucleotidyl transferase (TdT).

Based on cytomorphology and flow cytometric analysis, a final diagnosis of mantle cell lymphoma, manifesting as acute leukemia was given.

Case Report



Microphotograph 1: Mantle Cell Lymphoma- Bone Marrow Aspirate Showing Atypical Lymphoid Cells with Predominant Population of Lymphoid Cells with Prominent Nucleolus in a Case of Mantle Cell Lymphoma (400x)



Microphotograph 2: Mantle Cell Lymphoma- Bone Marrow Biopsy Showing Infiltration by Atypical Lymphoid Cells in a Case of Mantle Cell Lymphoma (400x, H & E)

DISCUSSION

MCL is a clinically aggressive B-cell neoplasm. A chromosomal translocation t (11;14) (q13;q32) is the molecular hallmark of MCL, resulting in the over expression of cyclin D1. Age >65-70 years, peripheral blood involvement at diagnosis, splenomegaly, and high lactate dehydrogenase (LDH) level are associated with a worse prognosis (Vose, 2012). The differential diagnosis of MCL includes small lymphocytic lymphoma, marginal zone lymphoma, and follicular lymphoma (Francesc *et al.*, 1998). Immunophenotypically, the neoplastic cells express CD5 antigen along with a number of pan B-cell markers (CD19, CD20) but not CD23 (Bernard *et al.*, 2001). The absence of SOX-11 or a low Ki-67 may

Case Report

correlate with a more indolent form of MCL (Francesc *et al.*, 1998). MCL is usually diagnosed once it has spread throughout the body. Patients with blastoid variant MCL are associated with particularly short durations of response after chemotherapy and poorer overall survival (Lee *et al.*, 2002).

Leukemic presentation of MCL is a rare phenomenon with only few such cases reported in the literature. The clinical presentation, blood and bone marrow picture may prompt the initial consideration of acute leukemia in these cases. The blastic form of MCL may be difficult to diagnose and is akin to centroblastic large cell or lymphoblastic lymphoma (Lee *et al.*, 2002). However, flow cytometric studies help in reaching the right diagnosis. CD45+, CD19+, CD20+, CD5 +, cyclinD1+, CD23-, CD10- is the characteristic flow cytometric presentation of MCL. Similar to the index patient, leukemic presentation of MCL have been reported by Lee *et al.*, (2002), Shukla *et al.*, (2011) and Sathyanarayan *et al.*, (2014) have previously reported leukemic presentation of MCL.

To conclude, we describe an elderly male with clinically aggressive MCL, masquerading as acute lymphoblastic leukemia. Accurate diagnosis in such case requires immunophenotyping and cytogenetic studies.

Table 1: Peripheral Blood Picture

Hemoglobin	8.9g/dl
WBC	57000/ μ L
Platelets	80,000/ μ L
Blasts	56%

REFERENCES

- Bernard M *et al.*, (2001).** Blastic variant of mantle cell lymphoma: a rare but highly aggressive subtype. *Leukemia* **15** 1785-1791.
- Francesc *et al.*, (1998).** Mantle Cell Lymphoma Presenting Features, Response to Therapy, and Prognostic Factors. *Cancer* **82** 567-575.
- Lee MJ *et al.*, (2002).** Mantle Cell Lymphoma, Blastoid Variant, Diagnosed on the Basis of Cytomorphology and Flow Cytometric Immunophenotyping of the Lymph Node Aspirate and Peripheral Blood. *Journal of Korean Medical Science* **17** 173-178.
- Sathyanarayan V *et al.*, (2014).** Mantle cell lymphoma (MCL): A single-center experience from a tertiary care institute in south India. *Journal of Clinical Oncology* **32**.
- Shukla S *et al.*, (2011).** Mantle cell lymphoma, blastoid variant presenting as acute leukemia: A rare case report. *Indian Journal of Cancer* **48** 370-37.
- Vose JM (2012).** Mantle cell lymphoma: 2012 update on diagnosis, risk-stratification, and clinical management. *American Journal of Hematology* **87**(6) 604-609.