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CASE REPORT

# Waldenstrom macroglobulinemia- Diagnostic dilemma in hemato-oncology

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**ABSTRACT**

Macroglobulinemia refers to the excess production of IgM monoclonal protein that occurs in certain lymphoproliferative disorders and plasma cell dyscrasias. This broad definition includes patients with monoclonal gammopathy of undetermined significance of the IgM type (IgM MGUS), smoldering Waldenström macroglobulinemia (SWM), Waldenström macroglobulinemia (WM), and a number of related disorders in which an IgM monoclonal protein is detected, such as chronic lymphocytic leukemia (CLL), a number of lymphoma variants, and primary (AL) amyloidosis.

WM is a diagnosed by demonstrating lymphoplasmacytic lymphoma (LPL) in the bone marrow with an IgM monoclonal gammopathy in the blood. Infiltration of the hematopoietic tissues or the effects of monoclonal IgM in the blood causes the symptoms associated with disorder.

**Case**: A 42-years-Male presented with complains of generalised weakness, dizziness, easy fatiguability and fever (mild, intermittent) for last 2 yrs. He denied any history of cough, abdominal pain, jaundice, bladder/ bowel disturbances, rashes, headache, blurred vision. He was addicted to tobacco in form of 'surti'. There was no evidence of chronic illness/comorbidity. On examination, patient was average built with moderate pallor and mild bilateral pitting pedal edema. Generalised lymphadenopathy (1-2 cm, firm, nontender, discrete bilateral cervical, axillary and inguinal region) was evident. Systemic examination was unremakable. On investivating, his haemogram showed a haemoglobin of 5.7 g% (dimorphic anaemia in GBP), white count of 4,900/cumm (N68% L25%), thrombocyte count of 168,000/cumm, ESR 36 mm/hr. His random blood sugar, renal parameters, liver function tests (except for total proteins 8.9 mg/dl and albumin 2.4 mg/dl) and serum lactate dehydrogenase levels were within normal limits. Serum for HIV and hepatitis B and C markers were negative. Electrocardiogram and X-ray Chest and skull were normal. USG abdomen revealed mild hepatosplenomegaly with prominent portal vein (14.8mm) and prominent splenic vein (12.1mm). Serum electrophoresis suggested features of monoclonal gammapathy with 'M' spike. Serum IgM levels were markedly raised (813 microgram/dl). Urinary bence jones proteins were weakly positive. Axillary lymph node biopsy suggested features of lymphoproliferative disorder (suspicious of malignancy), hence bone marrow biopsy was performed, which showed abnormal findings of prominent rouleax formation with depressed normoblastic

erythropoesis, M:E ratio 40:1 , with 5% lymphoplasmacytoid cells and 1% plasma cells in lymphoid series favouring the diagnosis of Waldenstrom's Macroglobulinemia.

## DISCUSSION

Among the lymphoproliferative disorders, Waldenström's macroglobulinemia (WM) is unique accounting for approximately 2% of haematologic malignancies[1]. It is characterised by increase production of a monoclonal immunoglobulin (IgM) protein and bone marrow shows lymphoplasmacytic infiltrate. The median age at presentation is 63 years[2]. Presence of pleomorphic B-lineage cells at different stages of maturation, such as small lymphocytes, lymphoplasmacytoid cells, and plasma cells is also characteristically seen in WM[3].

The common presenting features are weakness and fatigue, haemorrhagic manifestations, and weight loss because of lymphocytic infiltration of marrow leading to cytopenias, especially anemia with hepatosplenomegaly and lymphadnopathy. Further hyperviscosity causes neurologic symptoms in form of visual disturbances and Raynaud's phenomenon. The median age of survival ranges between 5 and 10 years.

Conventional treatment regimen includes alkylating agents such as chlorambucil or cyclophosphamide. The most common causes of death in these patients are progression of the malignant lymphoproliferative process, infection and cardiac failure.[4]

## REFERENCES

1. Dimopoulos MA, Alexanian R. Waldenstrom's macroglobulinemia. *Blood* 1994; **83**: 1452-9.
2. Fonseca R and Witzig TE. "Waldenström macroglobulinemia," in Wintrobe's Clinical Hematology, J.P. Greer, J. Forester, J.N. Lukens, G.M. Rodgers, F. Paraskevas, and B. Glader, Eds. Lippincott Williams & Wilkins, Phildelphia, Pa, USA, 11th edition, 2004, 2667–74.
3. Treon P, Morel P, Leblond V, and Fermand JP, "Report of the third International Workshop on Waldenström's Macroglobulinemia*," Clinical Lymphoma* 2005; **5(4),** 215–16.
4. Kyle RA, Garaton JP. The spectrum of IgM monoclonal gammopathy in 430 cases. *Mayo Clin Proc* 1987; **62**: 719-31.