

Title: *Waldenstrom's Macroglobulinemia: Clinical presentation & treatment outcome from a tertiary care centre from India*

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Introduction: Waldenstrom's macroglobulinemia (WM) is a low grade, B-cell lymphoma characterized by the infiltration of the bone marrow by clonal lymphoplasmacytic cells that secrete monoclonal IgM immunoglobulin. This is a rare B NHL, which has an indolent course but remains incurable. It is characterized by MYD88 mutation, which is seen in 90% of patients. CXCR4 is the second most common mutation seen in 40% of patients. Since this is a rare malignancy, there is not much published literature in Indian settings. We herein present baseline characteristics and treatment outcomes of patients with Waldenstrom macroglobulinemia from a tertiary care centre in north India.

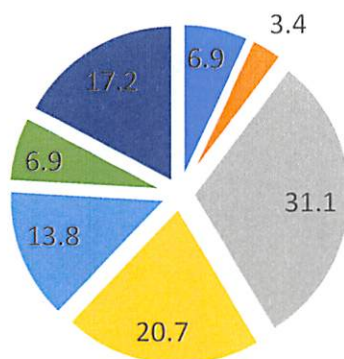
Material & Methods: We retrospectively collected the data from Hospital information system (HIS) & Radiology department for patients of Waldenstrom macroglobulinemia from Jan 2016 to January 2024.

Results: We had a total of 46 patients diagnosed with WM. The median age was 65 years with male preponderance (82.6%). Lymphadenopathy was seen in 18 patients (39.1%), splenomegaly in 11 patients (23.9%) and hepatomegaly in 7 patients (15.2%). One patient had presented with features of hyperviscosity. Mean Hb was 7.6g/dl and mean platelet count was 1.31L/dl. Mean M band on SPEP was 2.7. On IFE, majority were IgM kappa (89.1%). MYD88 mutation status was seen in 25 patients. 23 out of these 25 patients (92%) had positive MYD88. The patients were further stratified according to R IPSSWM. 2(4.3%) patients belonged to low risk, 8 (17.4%) had intermediate risk, 21 (45.7%) had high risk and 15 (32.6%) had very high risk. 29 out of the 46 patients underwent therapy at our institute. The commonly used regimens were Bortezomib, Dexamethasone and Rituximab (BDR), Rituximab Bendamustine and Rituximab Cyclophosphamide Dexamethasone (RCD). 5 patients were lost to follow up after initiation of therapy and hence their response could not be assessed. Out of the remaining patients, CR was observed in 2 patients, VGPR in 1 patient, PR in 9 patients and MR in 6 patients. ORR was 75%. 4 patients had stable disease and 2 patients had progressive disease. 7 patients had relapsed after frontline therapy. 3 patients received R Benda and 2 patients received BDR as 2nd line therapy. 1 patient expired and 1 patient was lost to follow up after relapse. 1 patient, who had received R Benda as 2nd line relapsed and was started on Acalabrutinib. A total of 4 patients died. 2 patients died due to disease complications in the form of hyperviscosity and sepsis with DIC. 1 patient died at relapse due to sepsis. 1 patient, in 2nd remission, died to road traffic accident.

Conclusions: In this study, we report the presenting features as well as the treatment outcomes of Waldenstrom macroglobulinemia. The response rates were lower when compared to Western literature. However, this is one of the few Indian studies, till date, to report on the characteristics of the disease.

Baseline characteristics	N=46
Median age	65 years (47-85 years)
Sex	Male- 82.6% Female- 17.4%
Lymphadenopathy	18 (39.1%)
Hepatomegaly	7 (15.2%)
Splenomegaly	11 (23.9%)
Lung lesion	1 (2.1%)
Hyperviscosity	1 (2.1%)
Mean Hb (g/dl)	7.6 (3-13.7)
Mean platelet count($\times 10^9/l$)	131 (8-331)
Mean M band (mg/dl)	2.7 (0.3-5.4)
IFE	IgM kappa- 41 (89.1%) IgM lambda- 5 (10.9%)
MYD88	Positive- 23/25(92%) Not done- 21
R IPSSWM	Very low- 0 Low- 2 (4.3%) Intermediate- 8 (17.4%) High- 21 (45.7%) Very high- 15 (32.6%)

Response



■ CR ■ VGPR ■ PR ■ MR ■ SD ■ PD ■ LTFU