Treatment Advances in Waldenstrom's Macroglobulinemia.

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Waldenström's macroglobulinemia (WM) is a distinct B-cell disorder resulting from the accumulation, predominantly in the bone marrow, of clonally related IgM secreting lymphoplasmacytic cells. Asymptomatic patients should be observed. Patients with a disease related hemoglobin level <10g/L, platelet count<$100x10^9$/L, bulky adenopathy or organomegaly, symptomatic hyperviscosity, peripheral neuropathy, amyloidosis, cryoglobulinemia, cold-agglutinin disease or evidence of disease transformation should be considered for therapy. Nucleoside analogues and oral alkylators should be avoided in younger patients due to increased risks of secondary malignancies. Plasmapheresis should be considered for symptomatic hyperviscosity, and as a prophylactic measure prior to rituximab administration in patients with high IgM levels. Cyclophosphamide based therapy in combination with steroids and rituximab can be considered as an appropriate first line treatment. The use of vincristine should be avoided in such combinations due to the risk of neuropathy. Bortezomib in combination with steroids and rituximab may be particularly beneficial to those patients with symptomatic hyperviscosity and those needing more immediate disease control. Thalidomide in combination with rituximab is active and can produce prolonged remissions. Newer agents such as bendamustine, everolimus and pomalidomide are under investigation for the treatment of WM, and updates of ongoing WMCTG efforts examining these agents will be presented.