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Clinicopathological definition of Waldenstrom's macroglobulinemia: consensus panel recommendations from the Second International Workshop on Waldenstrom's Macroglobulinemia.

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This presentation represents consensus recommendations for the clinicopathological definition of Waldenstrom's macroglobulinemia (WM), which were prepared in conjunction with the Second International Workshop held in Athens, Greece during September 2002. WM is an uncommon lymphoproliferative disorder characterized primarily by bone marrow infiltration and IgM monoclonal gammopathy. It should be considered a distinct clinicopathological entity rather than a clinical syndrome secondary to IgM secretion. The underlying pathological diagnosis in WM is lymphoplasmacytic lymphoma as defined by the World Health Organization (WHO) and Revised European-American Lymphoma (REAL) classification criteria. The concentration of monoclonal IgM can vary widely in WM and it is not possible to define a concentration that reliably distinguishes WM from monoclonal gammopathy of undetermined significance (MGUS) and other lymphoproliferative disorders. A diagnosis of WM can therefore be made irrespective of IgM concentration if there is evidence on a bone marrow trephine biopsy of bone marrow infiltration by lymphoplasmacytic lymphoma with predominantly an intertrabecular pattern, supported by appropriate immunophenotypic studies. Simple criteria to distinguish patients with symptomatic WM who require therapy from those with asymptomatic WM and MGUS were also proposed. Patients with clinical features attributable to IgM monoclonal gammopathy but no overt evidence of lymphoma are considered to constitute a distinct clinical group and the term "IgM-related disorders" is proposed. Copyright 2003 Elsevier Inc. All rights reserved.