

ASYMPTOMATIC WALDENSTROM'S MACROGLOBULINEMIA IN A YOUNG ADULT

Abdul N. Memon, M.D.*

Summary

We report a case of WM which is unusual in two respects: the patient was 36-years-old and was asymptomatic at diagnosis. Because of the young age of our patient, WM was not an initial consideration and was a somewhat surprising finding. However, upon review of the literature on WM, it appears that approximately 5% of the patients with WM are between 30-40 years of age.^{4 5 6} The percentage of these young patients who were asymptomatic at diagnosis, such as our patient, is impossible to determine from the published data.

Waldenstrom's macroglobulinemia (WM) is a neoplastic disorder of B lymphocytes, which are intermediate in differentiation between mature lymphocytes and plasma cells and which synthesize and secrete monoclonal IgM¹. Most patients with WM seek medical care because of symptoms due to high levels of circulating paraprotein (i.e. hyperviscosity), lymphadenopathy, hepatosplenomegaly, or bone marrow hypofunction caused by infiltrating neoplastic cells.

Case Report

A 36-year-old white female was hospitalized for dehydration resulting from nausea and vomiting due to gastroenteritis. Her past medical history was unremarkable except for infectious mononucleosis seven years previously. Physical examination revealed no abnormality except signs of dehydration. The fundi were unremarkable. There was no lymphadenopathy or hepatosplenomegaly.

Laboratory data included a hemoglobin of 10 g%, WBC 5700/cumm with a normal differential count, platelets 288,000/cumm, and erythrocyte sedimentation rate (ESR) 66 mm/hr. Other chemistry tests were normal. Her dehydration was corrected with intravenous fluids and with oral feedings. Gastrointestinal evaluation and roentgenograms were negative for malabsorption or intestinal pathology. Before her anemia could be evaluated, she was discharged at her insistence.

During the nine subsequent months, the patient was asymptomatic. A hematological evaluation at the time showed a hemoglobin of 11.4 g%, WBC 6600/cumm, platelets 316,000/cumm, serum viscosity 2.6, and ESR 116 mm/hr. Coagulation and iron studies were normal and hemolysis work-up

(including Coomb's testing) was negative. A bone marrow aspirate and biopsy revealed a normal cellularity with increased plasma cells, lymphocytes, and plasmacytoid lymphocytes consistent with Waldenstrom's macroglobulinemia. Gallium scan and liver-spleen scan were normal. A serum protein electrophoresis showed an albumin of 4.1 g/dl and globulin of 2.7 g/dl with monoclonal spike (Fig. 1). Quantitative immunoglobulin analysis revealed an IgG of 430 mg/dl, IgA 62 mg/dl, and IgM 4200 mg/dl. An immunoelectrophoresis demonstrated a monoclonal IgM of the kappa type (Fig. 1). Additional chemistry studies were normal.

On follow-up, during the next 18 months, the patient's hematologic parameters have remained unchanged. She has received no therapy since she continues to be asymptomatic without organomegaly or signs of hyperviscosity.

Discussion

The diagnosis of WM in this 36-year-old patient was based upon the presence of anemia, a high ESR, a monoclonal IgM kappa protein, and a bone marrow infiltration with lymphocytes, plasma cells, and plasmacytoid lymphocytes. Although unusual for WM,^{2,3} she was asymptomatic and

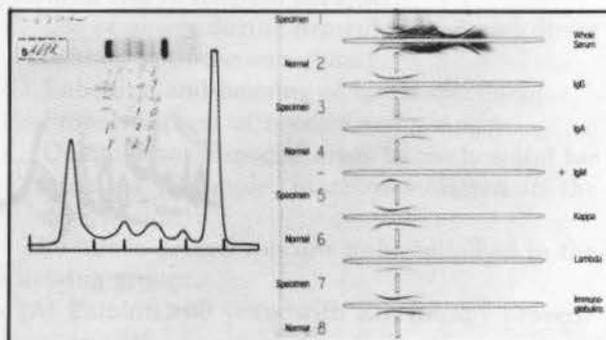


Fig.1 Serum protein electrophoresis (left) shows monoclonal protein peak and immunoelectrophoresis (right) demonstrates a monoclonal IgM kappa protein.

*Internist (Diplomate, ABIM) Oncologist (Diplomate, ABIM), The Memorial Hospital, Pawtucket, R.I.

without organomegaly at diagnosis and during the 1½ years of follow-up evaluation. Our patient showed no evidence of a malignant disorder, infectious disease, or connective tissue disorder to explain her IgM paraprotein. Since her episode of infectious mononucleosis was 7 years previously it is unlikely that it was in some way related to her IgM paraproteinemia.

A review of 196 cases of WM demonstrated fatigue, weakness, lymphadenopathy, hepatosplenomegaly, hemorrhage, visual disturbances, and/or ocular findings in over 95% of the cases.^{4 5} Less than 5% (8/196) of patients with WM were asymptomatic at the time of diagnosis.^{4 5 6} Among these asymptomatic cases, WM was found after evaluation of elevated ESR's, mild anemia, or hyperglobulinemia.⁵ Very rarely pruritus has been the only presenting symptom of WM.^{7 8} The five asymptomatic patients reported by McCallister et al were followed for 2-9 years without any apparent progression of their WM,⁵ currently similar to our patient.

An investigation of our patients was prompted only by her mild anemia and elevated ESR. Almost 90% of patients with WM have anemia.^{5 9 10 11} The etiology of anemia can be multifactorial and includes disturbed erythropoiesis,⁴ hemolysis with or without a positive Coomb's test^{12 13} and blood loss.^{4 13} An elevated ESR appears to be universal in WM.^{10 14 11}

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Request For Reprints: Abdul N. Memon, M.D.,
The Memorial Hospital, Pawtucket, RI 02860