



“I Have Macroglobulinemia, and Now What?” —Case Report

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Abstract

Waldenström macroglobulinemia (WM) is a chronic, indolent, lymphoproliferative disease of B lymphocytes, characterized by a lymphoplasmocytic lymphoma in the bone marrow and by Immunoglobulin (Ig) M monoclonal hypergammaglobulinemia. WM is considered incurable and the treatment of asymptomatic patients doesn't show benefit in terms of mortality. In this article, the authors present a case report of a 64-year-old woman, without relevant pathological history and asymptomatic, which resorted to a consultation on her Family Health Unit to show the results of a blood analytical study that was requested in a private medical routine consultation. In this study, erythrocyte sedimentation rate (ESR) had a value of 89 mm/h. One month later, ESR remained elevated, 94 mm/h. The further study revealed IgM monoclonal hypergammaglobulinemia of 26 g/L. The patient was sent for additional evaluation in the Haematology Service of the Hospital from the home area. In the course of the study, the patient showed increased anxiety and depressed mood due to the absence of a specific diagnosis. Finally, the diagnosis of WM was established, but, in the absence of symptoms related to the disease, no treatment was offered. The patient remained anxious, depressed and reluctant in accepting the diagnosis of a chronic disease without proper treatment. Therefore, the Family Physician has an essential role not only in choosing the most appropriately complementary diagnostic exams for their patients, but also in dealing with abnormal results of these exams. It's crucial to protect patients from unnecessary exams or other health interventions. Sometimes, some diseases don't have a proper treatment, requiring more effort in clarifying and monitoring these cases.

Subject Areas

Hematology, Psychiatry & Psychology

Keywords

Complementary Diagnostic Exams, Erythrocyte Sedimentation Rate, Quaternary Prevention

1. Introduction

The erythrocyte sedimentation rate (ESR), also called sedimentation rate or Westergren ESR, is the rate at which red blood cells precipitate in a period of one hour. The ESR is a nonspecific test that is often used as a screening test for patients with unexplained fevers, certain types of arthritis, muscle symptoms, or other vague symptoms of unknown origin [1].

Waldenström macroglobulinemia (WM) is a chronic, indolent, lymphoproliferative disease of B lymphocytes, characterized by a lymphoplasmocytic lymphoma in the bone marrow and by Immunoglobulin (Ig) M monoclonal hypergammaglobulinemia [2]. The WM is a rare disease, with an estimated incidence of approximate three per million people per year, accounting for about 2% of all hematological cancers [3]. There is a higher incidence in individuals aged between 63 and 68 years. Approximate 60% of patients are men, and it is more common in Caucasian individuals [4]. The average survival is 5 years, however, approximate 10% of patients survive up to 15 years [5]. No definite etiology exists for WM. Environmental, familial, genetic, and viral factors have been reported. IgM monoclonal gammopathies of undetermined significance are considered as a precursor of WM [6].

Most patients have clinical signs and symptoms related to hyperviscosity resulting from IgM monoclonal gammopathy, and/or cytopenias resulting from bone marrow infiltration by lymphoma. The differential diagnosis with other lymphomas is essential for the assessment of prognosis and therapeutic approach [7].

The International Prognostic Staging System for WM (IPSSWM) adopts five variables that correlate with poor survival of patients under treatment (Figure 1). The absence or presence of one or more prognostic factors categorizes the patient into 3 risk levels: low, intermediate or high. In patients at low risk, the average survival time is 12 years, and treatment should involve low toxicity, preserving quality of life [7]. Patients who meet the criteria for WM (IgM monoclonal hypergammaglobulinemia, bone marrow lymphoplasmacytic infiltration, or both) without end-organ damage are considered to have indolent disease or smoldering WM. However, there is a 10-fold higher risk of transformation to an active WM and such patients should be monitored closely (*i.e.*, every 6 months) [6].

Treatment of patients with asymptomatic WM does not improve the quality of life of patients, or increase their survival, being recommended, therefore, their follow-up [2]. Patients can be observed carefully with periodic measurement of the M component, immunoglobulin, and serum viscosity [7].

IPSS WM risk factors (sum the nº of factors)	Determine risk category and prognosis			
• Age > 65 years (advanced age alone = intermediate risk)	nº of risk factors	Risk category	% of patients	Median survival (months)
• Hemoglobin ≤ 11.5 g/dL	0 or 1 (except age)	Low	27	142
• Platelet count ≤ 100 × 10 ⁹ /L	Any 2, or age alone	Intermediate	38	99
• Serum B ₂ -microglobulin > 3 mg/L	≥3	High	35	43
• Serum monoclonal protein > 70 g/L				

Figure 1. International prognostic scoring system for WM.

Sometimes, health activities not only produce benefits, but also harm. This case report aims to highlight the holistic consequences for the patient after conducting an unnecessary diagnostic test (ESR).

2. Case Report

A 64-year-old woman, retired, without relevant pathological history and asymptomatic, recurred to her Family Health Unit to show the results of a blood analytical study that was requested in a private medical routine consultation. In this study, ESR had a value of 89 mm/h. One month later, ESR remained elevated, 94 mm/h. The further study revealed an IgM monoclonal hypergammaglobulinemia of 26 g/L.

The patient was sent for additional evaluation in the Haematology Service of the Hospital from the home area, where she performed bone marrow biopsy, immunophenotyping of B-lymphocytes and cytogenetic study. Cytogenetic study was negative. Bone marrow biopsy showed small lymphoid cells, some with plasma cell differentiation, positive for CD38 and CD138 antibodies. Immunophenotyping of B-lymphocytes showed bone marrow lymphoproliferative disease of B lymphocytes with inconclusive phenotypic characteristics (CD9+; CD20+; CD10-; CD5-; CD23-; FMC7-; Surface Ig M+; kappa light chain+). The imaging study of the skeleton didn't reveal injuries. The analytical study revealed anemia (hemoglobin 11.5 g/dl), IgM 30.2 g/L, kappa light chain 539 mg/dL and protein electrophoresis with *gamma* fraction of 29.9%.

In the course of the study, the patient showed increased anxiety and depressed mood due to the absence of a specific diagnosis. She expressed frustration with the medical assistance and intended to be evaluated in a specialized oncology institution. She was transferred to the Oncology Institute from the home area. Here, it was explained to the patient that clinical and analytical performed evaluation was compatible with WM IPSS 1 (Low Risk-Hemoglobin \leq 11.5 g/dL). It was also explained that, being a low risk patient without symptoms, treatment doesn't improve life's quality, or increases survival, being recommended, therefore, only follow-up. The patient remained anxious, depressed and reluctant in accepting the diagnosis of a chronic disease without proper treatment.

The longitudinal and holistic care provided by the Family Physician helped the patient to understand and accept her illness, resuming normal activities (walking and swimming).

The hemato-oncology doctor considered the disease to be indolent since the patient did not present end-organ damage. Currently the disease still remains indolent. She has medical oncology consultations every 6 months.

3. Discussion

In this case report, WM was diagnosed in an asymptomatic patient due to the requirement of unnecessary routine tests. Despite the importance of this diagnosis, in order to ensure proper monitoring, this situation caused anxiety and suffering to the patient that initially didn't accept the fact of having a chronic illness without appropriate treatment

and with uncertain prognosis.

Therefore, the Family Physician has an essential role not only in choosing the most appropriate complementary diagnostic exams for their patients, but also in dealing with abnormal results of these exams. It's crucial to protect patients from unnecessary exams or other health interventions. Sometimes, some diseases don't have a proper treatment, requiring more effort in clarifying and monitoring these cases, and affecting significantly life's quality of the patient that may not accept the diagnosis of a chronic disease without available treatment. Besides, the Family Physician, through holistic and longitudinal care, plays an essential role in helping patients to accept and adapt to their illness, relieving the suffering.

4. Conclusion

This case report highlights two aspects that are essential to any doctor, however particularly important to general practitioners: quaternary prevention ("primum non nocere") and longitudinal and holistic care provided by Family Physician, which are essential to help the patient to accept and adapt to their illness.

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