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Clinical Diversity and Treatment Outcomes in IgM MGUS/Waldenstrom disease Associated with Cold Agglutinin Disease and Cryoglobulinemia: A Single-Center Study

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Introduction: In primary cold agglutinin disease (CAD), autoimmune hemolytic anemia is caused by monoclonal IgMk cold agglutinins that bind erythrocyte antigens optimally at 4°C, leading to agglutination and complement activation. Pathogenic antibodies active above 28°C result in mostly extravascular hemolysis in the liver and, in severe cases, intravascular hemolysis.

Cryoglobulinemia involves immunoglobulins that precipitate below 37°C and redissolve on warming. Monoclonal IgM can be associated with type I cryoglobulinemia (monoclonal immunoglobulins only) and type II cryoglobulinemia (monoclonal IgM with rheumatoid factor activity binding to polyclonal IgG), often linked to hepatitis C.

Aim: This single-center descriptive study aims to highlight the diversity of patients with CAD and cryoglobulinemia associated with IgM monoclonal gammopathy/Waldenstrom's macroglobulinemia.

Methods: Between 2018 and 2024, 159 cases of IgM monoclonal gammopathy were diagnosed at the Fundeni Hematology Clinic in Bucarest, Romania. Among these, 68 patients (42.8%) had Waldenstrom's macroglobulinemia, 13 patients (8.2%) had smoldering Waldenstrom's macroglobulinemia, 36 patients (22.6%) had IgM MGUS, 18 patients (11.3%) had IgM MGCS, 13 patients (8.2%) had IgM AL amyloidosis, and 11 patients (6.9%) had CAD and cryoglobulinemia associated with IgM monoclonal gammopathy/Waldenstrom disease.

Results: The median age at diagnosis is 45 years, with 45% of the patients being diagnosed at this age. Among the patients, 54.5% are women (6 patients), and the time to diagnosis is 12 months. Most patients (7 patients, 63.6%) presented with CAD, 2 patients (18.2%) had both CAD and cryoglobulinemia, and 2 patients (18.2%) had only cryoglobulinemia associated with the IgM hematological disease. Five patients (45.5%) had IgM MGUS, and six patients (54.5%) had Waldenstrom's disease.

All patients were IgM Kappa, with a median light chain value of 371 mg/l. The median monoclonal component value was 1.8 g/dl, the median Hb was 8.5 g/dl. Five patients with CAD (45.5%) had cold antibody titers >1/1,000,000, and all patients with cryoglobulinemia (18.2%) had renal involvement of the GNMP I type, with one case (9,09%) showing coexistent thrombotic microangiopathy. Five patients (45.5%) had associated symptoms such as Raynaud's syndrome and 3 patients (27.3%) had peripheral neuropathy. Treatment included Rituximab in 10 patients (90.9%), R-Bendamustine in 4 patients (36.4%), R-CVP in 5 patients (45.5%), R-Cyclophosphamide in 1 patient (9.1%), and CVP in 1 patient (9.1%). Hematologic overall response rate (ORR) ≥VGPR was achieved in 2 patients (18.2%), PR in 2 patients (18.2%), SD in 2 patients (18.2%), and in 5 patients (45.5%), it could not be evaluated. Anemia improved by at least 2 g/dl in 7 patients (63.6%) after 2 cycles, while the remaining patients could not be evaluated. Seven patients (63.6%) died, with 3 patients (27.3%) dying from infections, 3 patients (27,3%) from cardiovascular disease, and 1 patients (9,09%) from disease progression.

Conclusions: This study highlights the clinical complexity and diverse manifestations of CAD and cryoglobulinemia in patients with IgM monoclonal gammopathy/Waldenstrom's disease. Treatment outcomes varied, with a notable improvement in anemia observed in most cases. Despite advancements in treatment modalities, mortality rates remain high, primarily due to infections and cardiovascular disease, emphasizing ongoing challenges in managing these conditions effectively.