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WALDENSTROM MACROGLOBULINEMIA: CAUSES, PATHOGENESIS AND SYMPTOMS

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Waldenström macroglobulinemia is a rare type of malignant lymphoma that is one of the malignant monoclonal gammopathies. A high level of a macroglobulin (immunoglobulin M [IgM]), higher serum viscosity, and a lymphoplasmacytic infiltration in the bone marrow are all symptoms of this condition. The overproduction of an antibody termed immunoglobulin M, or IgM, in the blood is a symptom. IgM is generated by a subset of white blood cells termed B lymphocytes, or B-cells for short, as are all antibodies. B-cells begin as stem cells in the bone marrow, where they go through several stages of development. When they reach adulthood, they leave the bone marrow and enter the bloodstream, where they are transported to secondary lymph organs including the spleen and lymph nodes. These B-cells can later develop into plasma cells, which give birth to immunoglobulins, within the secondary lymph organs. As a result, Waldenstrom's macroglobulinemia is now known as a lymphoplasmacytic lymphoma. These macroglobulins thicken blood and aggregate with one another, causing hyperviscosity syndrome, which causes blood vessels to become engorged, hypercoagulability, and a reduction in overall blood flow rate.

The presence of the IgM paraprotein and malignant lymphoplasmacytic cell infiltration of the bone marrow and other tissue locations cause the clinical symptoms of Waldenström macroglobulinemia. The clinical presentation is similar to that of multiple myeloma, with the exception that organomegaly is common in Waldenström macroglobulinemia but uncommon in multiple myeloma, and lytic bony disease and renal disease are uncommon in Waldenström macroglobulinemia but common in multiple myeloma.

The specific source of this overproduction like with many tumors is unknown but it is hypothesized to be linked to mutations in the MYD88 and CXCR4 genes. Two key factors contribute to the clinical manifestations of this condition.

First, IgM paraprotein secretion causes hyperviscosity and vascular problems due to the paraprotein's physical, chemical, and immunologic characteristics. Hyperviscosity syndrome, cryoglobulinemia types 1 and 2, coagulation abnormalities, sensorimotor peripheral neuropathy, cold agglutinin illness and anemia, primary amyloidosis, and tissue deposition of amorphous IgM in the skin, GI tract, kidneys, and other organs are among the consequences.

Second, malignant lymphoplasmacytic cells invade the bone marrow, spleen, and lymph nodes. Less typically, these cells can invade the liver, lungs, GI tract, kidneys, skin, eyes, and central nervous system. Infiltration of these organs results in a slew of clinical symptoms and indications.

Although Waldenström macroglobulinemia is incurable, a variety of medicines have showed promise in treating the condition. Careful supervision is required for asymptomatic individuals who do not have end-organ impairment. Monotherapy with rituximab is typically used in symptomatic patients, particularly in nonbulky illness. Bulky symptomatic illness may need the use of a combined regimen, such as chemotherapy. Ibrutinib has shown effectiveness as monotherapy for rituximab-resistant patients, particularly those with MYD88 gene mutations. Hyperviscosity syndrome may necessitate urgent plasmapheresis. In certain circumstances, autologous stem cell transplantation is an option.