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Waldenstrom Macroglobulinemia

Waldenstrom Macroglobulinemia is a non-Hodgkin lymphoma of plasma cells. It can present with peripheral neuropathy and hyperviscosity syndrome. Due to hyperviscosity, there can be headaches, blurred vision, Raynaud phenomenon, and thrombosis. Petechiae can be seen as well. On bone marrow biopsy, abnormal plasma cells with Dutcher bodies are typically evident. In the blood, monoclonal IgM antibodies are elevated. Management includes observation, chemotherapy, or plasmapheresis.



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Characteristics

Non-Hodgkin Lymphoma

Nun-hog-king with Lime-foam

Waldenstrom macroglobulinemia is a Non-Hodgkin lymphoma. Non-Hodgkin lymphomas are defined as a solid malignancy of lymphocytes without Reed Sternberg cells.

Clinical Features

Peripheral Neuropathy

Purple-wavy Neuron-extremities

In peripheral neuropathy, mixed peripheral nerve damage is present. It starts with proximal muscle weakness and overlying paresthesias.

Hyperviscosity Syndrome

Hiker-viscous-honey

When circulating antibodies increase in blood, it becomes viscous. Blood flow to organs becomes sluggish and microvasculature is affected causing a myriad of symptoms deemed hyperviscosity syndrome.

Headache, Blurred Vision

Headache-lump Blurry-eye

Due to hyperviscosity, impaired blood flow to the retina and brain causes blurred vision and headaches.

Raynaud Phenomenon

Blue Rain-cloud

Due to hyperviscosity, cold agglutinins are formed leading to impaired acral blood flow and causes Raynaud phenomenon. Raynaud phenomenon is characterized by intermittent interruptions of blood flow to the fingers, causing them to become pale and painful.

Thrombosis

Trombones

Sluggish blood flow causes venous stasis. Venous stasis is a component of of Virchow's triad and can lead to thrombosis.

Petechiae

Tiki-mask

Consumption of platelets due to coagulopathy causes an increased risk of bleeding. Petechiae are defined as non-palpable, non-blanchable lesions less than 3mm.
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Diagnosis

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Abnormal Plasma Cells

Abnormal Plasma Cell

The bone marrow is a primary lymphoid organ where plasma cells haven't undergone class switching. A bone marrow lymphoma of plasma cells on bone marrow biopsy would show >10% of abnormal plasma cells which have not undergone class switching.

Dutcher Bodies

Dutch-boy with Dutcher Bodies

The abnormal plasma cells in Waldenstrom macroglobulinemia contain intranuclear, Periodic Schiff acid positive inclusion bodies called Dutcher bodies.

Monoclonal IgM Antibodies

Monocle Mountain-goblin Ant-tie-body

Plasma cells before isotype switching produce monoclonal IgM. The plasma cells seen in Waldenstrom Macroglobulinemia produce monoclonal IgM.

Management

Observation

Observatory

Waldenstrom Macroglobulinemia is an indolent disease of old age. So, to avoid over-treatment asymptomatic patients are kept under observation.

Chemotherapy

Chemo-head-wrap In symptomatic patients, Rituximab, Bruton tyrosine kinase inhibitors, or alkylating agents are used.

Plasmapheresis

Plasma-fairy

To relieve the patient of hyperviscosity syndrome, IgM antibodies are filtered out of plasma using plasmapheresis.