A rare case of Waldenstrom Macroglobulinemia

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Introduction

Waldenström macroglobulinemia (WM) is a rare lymphoplasmacytic disorder with an incidence of 3 per million per year in the US, twice as common in the white population [1,2]. It involves medullary and extramedullary infiltration by B-type lymphocytes and plasma cells. Median survival is negatively correlated to age at diagnosis, with estimated survival of 10 years in patients under 70 and a median of 4 years in patients over 80 years of age [1,3]. Symptoms include pancytopenia, hepato-splenomegaly, hyperviscosity and rarely extramedullary disease, with pulmonary involvement present in 3-5% of cases [3]. Our case presented with symptoms mimicking COPD exacerbation.

Take-Home Points

- ❖ WM can masquerade as nonspecific respiratory or cardiac symptoms or mimick specific clinical entities such as Pulmonary embolism, COPD or CHF exacerbations.
- ❖ Serum viscosity is an inexpensive and highly specific test for Waldentrom macroglobulinemia
- ❖ Plasmapheresis is a symptomatic treatment while cytotoxic chemotherapy remains the only curative therapy

Conclusion

We need to maintain high clinical suspicion for hyperviscosity syndrome in patients with cytopenia and pulmonary or cardiac complaints. Early diagnosis and plasmapheresis initiation prevent complications. Cytotoxic chemotherapy is the only curative treatment, requiring compliance and rigorous follow-up.

Case Study

In 2022, a 78-year-old African American female with history of COPD, CAD, PVD, HTN, HLD, T2 DM, and hypothyroidism presented with progressive shortness of breath, chest pain, dizziness and cold-like symptoms. On arrival, she was saturating well on RA with increased respiratory rate and unremarkable findings on chest x-ray. CT chest was also negative for PE. She had a history of pancytopenia with megaloblastic anemia previously attributed to her underlying cirrhosis. The clinical presentation was suggestive of acute on chronic bronchitis. SPEP and IFE were ordered and results showed elevated serum proteins with gamma gap and IgM kappa monoclonal gammopathy 4.0.

Based on bone marrow biopsy results she was diagnosed with Waldenstrom macroglobulinemia.

She was readmitted the same year for hyperviscosity syndrome that required emergent plasmapheresis. Therapy with Bendamustine and Rituxan was initiated, with Rituximab as maintenance treatment. The patient was further seen in outpatient Hematology-Oncology Clinic and started on Xarelto for gastrocnemius DVT, possibly secondary to hyperviscosity, but patient was lost to follow-up. When she reestablished care 6 months later she was having severe symptoms with a serum viscosity was 11.8 cP (NV 1.4-1.8cP), she underwent emergent plasmapheresis a second time and chemotherapy was restarted, with good clinical outcome.

Discussion

Lung involvement is rare but could be the primary manifestation in patients with WM. Pulmonary symptoms include chest pain, cough, shortness of breath and rarely malignant pulmonary effusions [3]. These symptoms are due to protein deposition (amyloid), hyperviscosity or direct plasma cell infiltration [3]. While only 13% of WMG cases present with hyperviscosity syndrome, roughly 80 to 90% of symptomatic hyperviscosity cases are attributed to the disorder [4], thus serum viscosity remains an invaluable test that is rapid, inexpensive and readily available in most centers.

Plasmapheresis is the first-line therapy for symptomatic hyperviscosity syndrome. Definitive treatment is addressing the underlying disease with first line chemoimmunotherapy: alkylating drugs and proteasome inhibitors, or Ibrutinib, in combination with rituximab [3,5].

References

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