A rare case of IgM Multiple Myeloma presenting with hyperviscosity syndrome

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BACKGROUND

- Multiple Myeloma (MM) is characterized by the neoplastic proliferation of immunoglobulin-producing plasma cells and has various subtypes, the rarest of which is IgM.
- Common presentations of MM are related to the plasma cell infiltration of the bone marrow or to kidney damage from excess light chains.
- These include anemia, bone pain, elevated creatinine, fatigue, and hypercalcemia.
- Hyperviscosity syndrome is a much more common presentation in Waldenstrom macroglobulinemia (WM) but can very rarely occur in patients with IgM MM.

CASE PRESENTATION

- History:

A 74-year-old female with no significant past medical history presented to the hospital with 2 weeks of blurry vision and lightheadedness.

- PE:

Afebrile, hemodynamically stable.
Ophthalmic evaluation noted bilateral central retinal vein occlusions.

- Labs:

CBC - anemia with hemoglobin of 8.0 g/dL.
SPEP/IFE – elevated IgM kappa monoclonal protein 5.02 g/dL.
UPEP/IFE – 335 mg/d paraprotein excretion.
IgM > 5,000 mg/dL, Kappa FLC 147.5 mg/L, K/L ratio 22.5.
Beta-2 macroglobulin 6.7 mg/L
Serum viscosity elevated 5.22 centipoise
MYD88 L265P mutation was not detected.

- Pathology

Bone marrow biopsy: significant for IgM multiple myeloma. CD138 stain showed plasma cells to account for approximately 30% of cellularity. No B-cell proliferation identified by morphology or by flow cytometry.

DIAGNOSIS AND TREATMENT

- Patient met criteria for IgM kappa MM with 30% bone marrow plasma cells and anemia with hemoglobin 8 g/dL.
- FISH panel did not demonstrate any high-risk features such as del(17p), t(4;14), t(14;16), t(14;20), or gain(1q).
- Patient received daily therapeutic plasma exchange until the IgM level improved to 2,618 mg/dL.
- When blurry vision and hyperviscosity syndrome improved, she was started on myeloma-directed therapy with bortezomib, lenalidomide, and dexamethasone.

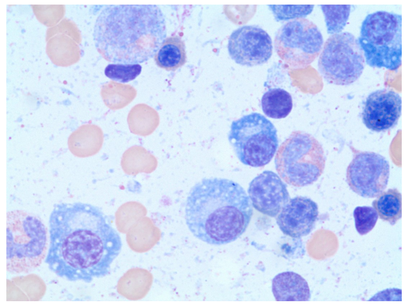

Figure 1:
Touch preparation from core bone marrow biopsy at 100x magnification showing the plasma cells with eccentric nuclei and perinuclear hof.

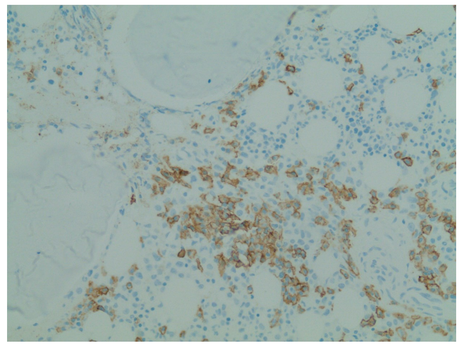

Figure 2:
CD138 immunohistochemical stain at 20x magnification.

DISCUSSION

- WM is a distinct clinicopathologic entity demonstrating lymphoplasmacytic lymphoma (LPL) in the bone marrow with an IgM monoclonal gammopathy in the blood.
- While the presence of an IgM monoclonal protein is more typical of lymphoplasmacytic lymphoma, the absence of MYD88 L265P mutation and presence of bone marrow infiltration with clonal plasma cells are suggestive of Multiple Myeloma.
- IgM MM is an extremely rare subtype, comprising only 0.5% of patients with MM.
- When there is hyperviscosity syndrome due to high serum IgM levels, it is important to consider IgM MM as the etiology.
- Therapeutic plasma exchange relieves the symptoms of hyperviscosity syndrome and should be performed regardless of the serum viscosity level.
- Myeloma-directed therapy can be commenced when IgM levels improve to 4000 mg/dL.

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