Hyper-viscosity: Rare complication of Multiple Myeloma

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Introduction

- ❏Hyper-viscosity syndrome (HVS) is a rare complication seen in certain clonal lymphoproliferative disorders and plasma cell dyscrasias.
- ❏HVS is a group of clinical signs and symptoms triggered by abnormal increases in blood viscosity.
- ❏The three classic symptoms of HVS are neurological symptoms, visual disturbances and mucosal bleeding.
- ❏Clinicians should be aware of suggestive clinical findings, while maintaining a high degree of suspicion, preventing a delay in diagnosis and increased morbidity and mortality.

Case

- ❏A 72-year-old white male with a recent past medical history of Smoldering IgA Multiple Myeloma presented to the Hematology clinic with complaints of headaches, dizziness, confusion, memory loss, poor concentration, fatigue and worsening shortness of breath on exertion. Due to concerns for new onset neurologic symptoms, patient was immediately sent to the hospital for evaluation.
- ❏Imaging and labs were done as follows: Computer Tomography of the brain and x-ray of the chest showed no acute findings.
- ❏Labs were significant for Creatinine 1.21 mg/dL (baseline 0.8mg/dL), albumin 9.2g/dL, IgA > 6400mg/dL, IgM and IgG <150mg/dL and Hemoglobin 7g/dL. Plasma viscosity levels (PV) were 6.2cp. After 2 cycles of apheresis, patient's PV level was 2.6cp. Patient endorsed improvement in his symptoms after treatment.

Discussion

- ❏HVS is a rare and lesser known complication of MM. 2-6% of HVS are reported with MM patients and among those 25% account of IgA type paraproteinemia. IgA exists in dimeric/trimeric form in the plasma in contrast to most plasma proteins that exist in spherical form. These large shaped proteins travel through the serum, undergo high degree of polymerization causing aggregation of plasma components with increase in serum viscous drag.
- ❏Congestion and engorgement of blood vessels causing sluggish blood flow causing reduced oxygen and nutrient delivery to the tissues resulting in significant end organ damage if left untreated. If clinical symptoms raise suspicion, prompt action is a must, even without laboratory evidence.
- ❏Normal viscosity levels lie between 14-1.8 cp relative to water. Viscosity levels exceeding 4-5cp is when symptoms occur. The threshold for onset of symptoms for IgA is >6 and clinically with IgA MM in the range of 7000mg/dL Current guidelines recommend initiating treatment of symptomatic HVS rather than specified immunoglobulin levels. In such cases, symptoms take precedence over measured values.
- ❏Temporizing measures in acute situations are TPE and intravenous hydration; One cycle of Plasmapheresis can decrease serum viscosity by 30-50% and immunoglobulin levels by 60%. When plasma exchange is not available, phlebotomy can be performed.

Conclusion

- ❏ HVS should be considered in those with characteristic symptoms with an underlying paraproteinemia.
- ❏ Early recognition of symptoms with good history taking and timely intervention can potentially prevent catastrophic multi organ failure and save lives.

References:

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