Hyperammonemic Encephalopathy: A Tale of an Innocent Liver

Shivani Sharma
*Henry Ford Health System*

Yaser Alkhatib
*Henry Ford Health System*

Thushara Paul
*Henry Ford Health System*

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Introduction

- Hyperammonemia is a common cause of hepatic encephalopathy in patients with severe liver cirrhosis. In Multiple Myeloma, it has been seldom reported.
- The main manifestations are confusion, lethargy, hallucinations, seizures, coma and death.
- We describe a rare case of Multiple Myeloma presenting with hyperammonemic encephalopathy.

Case Presentation

- A 76 year-old Caucasian lady was admitted with sudden onset altered mental status.
- Initial work up included normal Computed Tomography of the head and diffuse background slowing on EEG consistent with metabolic etiology. Her CSF analysis was normal.
- Ammonia level was elevated at 180 mcg/dL despite the lack of underlying liver disease as evidenced by normal liver enzymes and imaging.
- She had normocytic anemia with hemoglobin level of 9 g/dL and thrombocytopenia at 90 K/mcL. Kidney function and calcium levels were normal.
- Further work up for normocytic anemia revealed a large IgG lambda monoclonal protein at 1.8 g/dL.
- Due to suspicion for Multiple Myeloma, patient was started on pulse steroids. Bone marrow biopsy confirmed Multiple Myeloma with 61% plasma cells.
- Patient’s clinical situation continued to deteriorate, and she had cardiopulmonary arrest, resulting in anoxic brain injury.
- Eventually, she passed away before initiating treatment for Multiple Myeloma.

Discussion

- Hyperammonemic encephalopathy is most frequently seen in patients with advanced stage disease. In very few occasions, it can be the presenting symptom of Multiple Myeloma.
- The pathophysiology of its occurrence is poorly understood.
- It is associated with high hospitalization mortality. Treatment of Multiple Myeloma has been shown to improve the encephalopathy related symptoms.

Conclusion

- Diagnosis of hyperammonemic encephalopathy due to MM requires the extensive diagnostic workup applied to acute confusional states in the elderly.
- In elderly patients with unexplained confusion, blood ammonia level should be measured. Serum protein electrophoresis is warranted to detect underlying Multiple myeloma as encephalopathy improves dramatically with targeted therapy.

References


Presenting author: Shivani Sharma, MD (ssharma9@hfhs.org)

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