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Hyperammonemic Encephalopathy: A Tale of an Innocent Liver

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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.

Radiographic and Clinical Images

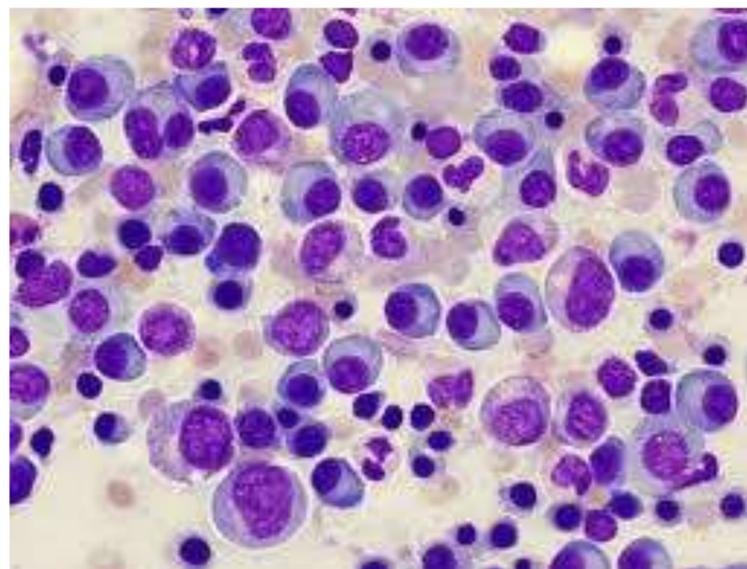


Figure 1. Patient's bone marrow biopsy showing Plasma cells.

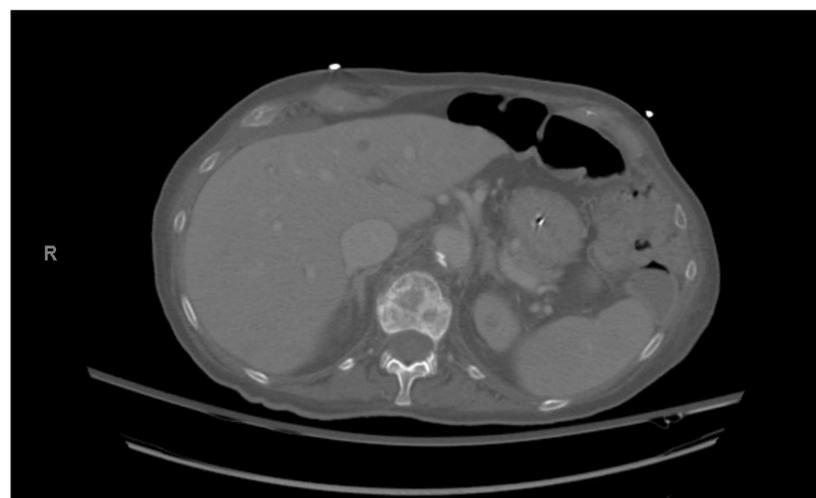


Figure 2. Patient's CT Abdomen Pelvis showing a normal liver morphology.

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.

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