RARE CASE OF MORTALITY FROM NON-CIRRHOTIC HYPERAMMONEMIA

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INTRODUCTION: Acute hyperammonemia is life threatening and can lead to encephalopathy, cerebral edema, and herniation if not treated aggressively. [1] Hyperammonemia is a known sequela of chronic liver disease, and is associated with significant morbidity in this population. [2] Less commonly, patients without liver disease will present with elevated ammonia levels. In these rare cases of acute non-cirrhotic hyperammonemia the differential diagnosis shifts to infectious or drug-induced.

CASE PRESENTATION: We report a case of a 58 year old female presenting with altered mental status in the setting of B-cell lymphoma, common variable immune deficiency (CVID), and hemorrhagic cystitis from cyclophosphamide. 2 weeks prior she had presented to the emergency department for shortness of breath, where workup showed coronavirus infection and asymptomatic pyuria, which was treated with 3 doses of fosfomycin due to her history of hemorrhagic cystitis. Workup for altered mental status revealed an elevated ammonia level of 306 umol/L without evidence of liver dysfunction. Ultrasound imaging confirmed a normal appearing liver. She was started on broad-spectrum antimicrobials and lactulose, but quickly required intubation for airway protection and was transferred to the ICU. Despite an aggressive regimen of lactulose every hour, her ammonia level increased to 642 umol/L. There was no underlying heavy metal exposure, and drug screens (including valproic acid) were unrevealing. Her urine culture and bronchoalveolar lavage grew Klebsiella, suspected to be the cause of such refractory and severe hyperammonemia. Dialysis was initiated in an attempt to clear the ammonia, which decreased to as low as 206 umol/L but did not normalize. Intravenous carnitine was tried without any improvement. Brain imaging demonstrated progressive cerebral edema eventually leading to uncal herniation and brain death. Neurosurgical intervention was not attempted due to refusal from family.

DISCUSSION: Hyperammonemia without liver dysfunction or an underlying urea cycle disorder is suspicious for an acute ingestion or infection. Therefore, prompt initiation of antibiotics, removal of ammonia, and testing for toxic substances are critical steps to initiate concurrently. Patients with urinary stasis and/or urinary tract anomalies are at increased risk for developing a complicated UTI. [3] We suspect that this patient’s immunocompromised state led to a severe infection with Klebsiella, and subsequently to the catastrophic elevation of ammonia.

CONCLUSIONS: Acute hyperammonemia without underlying liver dysfunction can be overlooked, but it carries significant morbidity and mortality. Therefore prompt intervention with lactulose, emergent dialysis, ICP monitoring and CNS decompression (if indicated) should be pursued.


DISCLOSURES: No relevant relationships by Kyle Chapman, source=Web Response
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