Altered Mental Status in a Teenager

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CASE

A 13-year-old Hispanic male presented to the emergency department with an altered mental status (AMS) after a 4-day history of nausea and vomiting. Values for electrolytes, glucose, blood urea nitrogen, creatinine, alanine aminotransferase, aspartate aminotransferase, alkaline phosphatase, calcium, and a complete blood count were all within their reference intervals. The patient's symptoms were treated with intravenous fluids and an antiemetic medication. He felt better and was discharged home. At home, the patient continued to vomit everything he ate or drank, even after receiving his antiemetic medication. Two hours after taking the antiemetic, he began saying things that did not make sense. He went to sleep, but he woke several hours later screaming, agitated, and in need of restraint in order not to hurt himself.

The patient was brought back to the emergency department 40 h after his initial presentation. He was obtunded, randomly reacting to touch, but not responsive to voice. He was admitted to the pediatric intensive care unit. He received both acyclovir for possible herpes encephalitis and cefotaxime until the cause of his symptoms was determined not to be sepsis. Tests of a sample from a lumbar puncture showed normal values for glucose, protein, and cell count, and antivirals and antibiotics were discontinued. The patient also received lorazepam for agitation and midazolam for sedation before undergoing a head computed tomography scan, the results of which were normal. A comprehensive urine drug screen showed only benzodiazepines from the midazolam.

The patient's medical history indicated presentation at age 11 with a 3-day history of emesis and changes in mental status. At that presentation, he was sleepy, was difficult to arouse, and showed decreased muscle tone. The results of a head computed tomography scan at that time were essentially normal. The results of laboratory tests, including those for electrolytes, glucose, calcium, magnesium, phosphorus, blood gases, and a complete blood count with differential were within their respective reference intervals. A urine drug screen detected promethazine, which had been prescribed for nausea and vomiting. At the time of this first visit, the change in the patient's mental status was attributed to an adverse reaction to promethazine. A test for ammonia was not ordered. The family history was notable in that his mother's first child had died on day of life 8 from unknown causes.

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Upon arrival in the pediatric intensive care unit for the current visit, the patient underwent testing for electrolytes and blood gases with a point-of-care instrument, which showed increases just above the reference intervals for sodium, pH, and bicarbonate, with a borderline-low value for PCO_2 (CO_2 partial pressure). The ammonia concentration was 308 μmol/L (reference interval, <50 μmol/L). We consulted Genetics, and they interviewed his parents. The interview revealed that although the patient was not strictly a vegetarian, he avoided animal protein in his diet. He ate no meat whatsoever and avoided milk and cheese. Analyses for urine organic acids and plasma amino acids were ordered.

Questions to Consider
• What is the differential diagnosis for a teenager who presents with AMS?
• What is in the differential diagnosis of hyperammonemia?
• What is the relevance of the family history in this case?

Final Publication and Comments

The final published version with discussion and comments from the experts will appear in the October 2013 issue of *Clinical Chemistry*. To view the case and comments online, go to http://www.clinchem.org/content/vol59/issue10 and follow the link to the Clinical Case Study and Commentaries.

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