Hyperammonemic Encephalopathy due to Intravenous Essential Amino Acid Administration in a Patient with Renal Insufficiency

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Dear Sir,

Essential amino acids are widely used in the management of patients with renal failure [1].

Hyperammonemic encephalopathy (HE) is a rare complication of intravenous essential aminoacid (IV EAA) administration [2–4]. HE due to IV EAA administration in a patient with renal insufficiency is presented.

A 20-year-old woman with a history of familial Mediterranean fever was admitted to Hacettepe University Hospital for a renal biopsy. Her problems included diffuse edema, renal insufficiency and nephrotic syndrome. Her renal biopsy showed amyloidosis, and therapy with oral colchisin and IV EAA (Ne-phramine) was initiated. Her mental status deteriorated after 8 h of 1,000 cm³ Ne-phramine infusion. Her blood pressure was 150/90 mm Hg, heart rate 110 beats/min, ventilatory rate 32 breaths/min and temperature 37°C. Neurologic examination revealed no focal abnormalities. She was not responsive to deep pain, and Babinski's reflex was extensor bilaterally. Laboratory studies disclosed the following results; hemoglobin 10.2 g/dl; leukocytes 7,800/mm³; blood urea nitrogen 71 mg/dl; serum creatinine 3.8 mg/dl; total protein 6.2 g/dl; albumin 1.9 g/dl; glucose 84 mg/dl; sodium 138 mEq/l; potassium 3.9 mEq/l; calcium 8.1 mg/dl; phosphorus 3.2 mg/dl; ALT 29 IU/l; AST 24 IU/l; Mg 2.1 mg/dl; prothrombine time 15 s. Arteriel blood test results were; P_{O2} 100.9 mm Hg; p_H 7.40; PCO₂ 31.4 mm Hg; HCO₃ 26 mEq/l. Abdominal ultrasonography and cranial CT scan were normal. A lumbar puncture showed normal results of cerebrospinal fluid (CSF), glucose, chloride, protein and cytology. Her blood, urine and CSF cultures were negative. Her serum ammonia was 300 µg/dl (normal 40–100 µg/dl). IV EAA administration was stopped, and the encephalopathy completely resolved after 12 h. Her laboratory tests were repeated when her mental status resolved, and the only significant difference was the decrease in serum ammonia from 300 to 83 µg/dl.

Hepatic pathologies are the most common cause of HE. It may also occur as a complication of urinary tract infection, as-paraginase therapy, valproic acid therapy, systemic carnitine deficiency and chemotherapy of acute leukemia [5]. Hyperammonemia due to IV EAA administration is rare in adults, and to the best of our knowledge, this patient is the second in the literature who also had renal insufficiency [2[^]1].

This patient's neurological examination was consistent with metabolic encephalopathy, and no structural central nervous system lesion was identified. We did not detect any medications or infections to explain the encephalopathy, and hepatic functions were normal. Clinical findings

did not suggest any enzyme deficiency either. Elevated serum ammonia level, absence of any other cause of encephalopathy and resolution of her encephalopathy and abnormal ammonia levels after discontinuation of IV Nephramin therapy were the evidence which showed that the patient's HE was due to IV EAA administration. Thus, metabolic encephalopathy due to IV EAA administration should be considered in the differential diagnosis of encephalopathy in patients with renal failure.

References

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