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CASE REPORT

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Non-cirrhotic Hyperammonemia causing altered sensorium in a patient who underwent Urinary diversion 43 years ago.

Sirotik olmayan hiperamoniemi, 43 yıl önce üriner derivasyon yapılan bir hastada, değişmiş duyarlığa neden oldu.

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ABSTRACT

Ureterosigmoidostomy, a urinary diversion procedure performed for treatment of congenital urologic defects or bladder cancer, can rarely presents with hyperammonemia even in the absence of coexistent liver disease. Here, we report the case of a 47-year-old man who developed hyperammonic encephalopathy 43 years after ureterosigmoidostomy. Therefore, hyperammonemic encephalopathy after ureterosigmoidostomy is an iatrogenic, but treatable problem which must be considered in the differential diagnosis of altered consciousness in the critical care settings.

Key words: Hyperammonemia, non-cirrhotic, urinary diversion

ÖZ

Doğuştan ürolojik defektlerin veya mesane kanserinin tedavisi için yapılan bir üriner diversiyon prosedürü olan Ureterosigmoidostomi, eşlik eden karaciğer hastalığının yokluğunda bile nadiren hiperammonemi ile seyredebilir. Burada, üreterosigmoidostomi sonrası 43 yıl hiperammonik ensefalopati gelişen 47 yaşında bir erkek hastayı sunuyoruz. Bu nedenle, üreteroopigmoidostomi sonrası hiperammonemik ensefalopati, kritik bakım ortamlarında değişmiş bilinçliliğin ayırıcı tanısında dikkate alınması gereken iyatrojenik, ancak tedavi edilebilir bir problemdir.

Anahtar kelimeler: Hiperamonyemi, sirotik olmayan, idrar yolu sapması

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INTRODUCTION

Hyperammonemic encephalopathy without coexistent liver disease has been rarely reported in patients with urinary stasis secondary to ureterosigmoidostomy, thought to be resulting from the diffusion of ammonia into the systemic circulation by the exposure of the colon to urine[1]. After passage of the blood brain barrier, ammonia accumulates in the astrocytes where it disturbs neuronal function, clinically presents as somnolence, seizures and behavioural alterations [2]. Here, we report the case of a 47-year-old man who developed hyperammonic encephalopathy 43 years after ureterosigmoidostomy. To the best of our knowledge, this is the third case of hyperammonic encephalopathy after ureterosigmoidostomy in the absence of additional pre-existing metabolic pre-disposition in the literatüre [3,4].

CASE REPORT

A 47 year old manual labourer was brought to our hospital with complaints of blurred vision for 3 days and disoriented behavior for the past 1 day. He also gave a history of fever and cough 1 week ago, that subsided with self-medication. He had no known co-morbidities and his medical history was unremarkable, except for a urological procedure performed in childhood. There were no reported addictions.

On examination, he was conscious, responding to call, but agitated and disoriented to time, place and person. Asterixis was present.There were no obvious signs of meningismus. He exhibited no focal neurologic deficits and his pupillary reflexes were normal. Deep tendon reflexes were normal too. He was afebrile, with normal breathing and hemodynamics. His abdomen was soft, non-tender and showed a transverse scar over the hypogastrium. His penis also showed evidence of corrective surgery in the past

In view of a recent (1 week) history of a fall from height, a CT Scan of the brain was performed which revealed no significant abnormalities. An MRI of the brain showed bilateral hyperintense areas in the Centrum semiovale, posterior limbs of internal capsule on the FLAIR and Diffusion weighted sequences.Though a meningo-encepahlitis seemed clinically discordant, we started him on Ceftriaxone and Acyclovir.

The blood workup revealed no dyselectrolytemias, however his Serum Ammonia level was 364, and his arterial blood gas analysis showed a Normal Anion Gap Metabolic acidosis . There were no clinical, radiologic or biochemical evidence in favour of liver cirrhosis. A detailed enquiry into the surgery performed in childhood revealed that the patient had a Bladder exstrophy and Epispadiasis, for which he had multiple corrective surgeries, including bladder excision and bilateral ureterosigmoidostomy at the age of 4 years. He was started on Lactulose syrup, Rifaximin and L-Ornithine L-Aspartate and subsequently the Ammonia levels decreased to 138 and sensorium improved. He was transferred to the ward after 2 days and thereafter discharged from the hospital in a week's time.

DISCUSSION

Ammonia is a highly potent neurotoxin well known for its implication in encephalopathy in severe, often cirrhotic liver disease. However, increased ammonia production or decreased ammonia elimination is one of the alternative etiologies of hyperammonemia. Ureterosigmoidostomy can lead to hyperammonemia, due to increased ammonia formation by bacterial degradation of the urine excreted directly into the sigmoid colon [1].

Ammonia directly affects neuronal electric activity by inhibiting the generation of both excitatory and inhibitory postsynaptic potentials. Furthermore, increased ammonia metabolism in astrocytes and elevated extracellular glutamate levels leads to an increase in the production of reactive nitrogen or oxygen species, resulting in increased intracellular osmolarity and vasodilatation, leading to increase in intracranial pressure and brain edema [5].

The exact reason for the 43 year lag after urinary diversion until presentation of hyperammonemia remains unclear. Our case is unusual in comparison with previous case reports of hyperammonemia associated with a ureterosigmoidostomy because of the 43 year long delay in the onset and the presence of normal renal and hepatic function. In conclusion, hyperammonemic encephalopathy after ureterosigmoidostomy is an iatrogenic, but treatable problem which must be considered in

the differential diagnosis of altered consciousness in the critical care settings.

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