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Acute hyperammonemic encephalopathy due to a portosystemic shunt in a non-cirrhotic adult patient

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ABSTRACT

Objective: To report a successfully treated hyperammonemia due to a portosystemic shunt in adult patient.
Data source: A patient with an altered mental status due to severe elevated ammonia level because of a portosystemic shunt.
Conclusions: Hyperammonemia is not always related to liver failure in critically ill patients, but should be considered in all unknown origins of an altered mental status. A portosystemic shunt can be the responsible for this phenomenon, and it has a newly treatment technique named plug-assisted retrograde transvenous obliteration (PARTO), which can be quickly performed with high technical success rate and clinical efficacy for the treatment of the splenorenal and/or gastrorenal shunt.

1. Case report

A 66 year-old male presented in the emergency room due to abrupt apraxia, bilateral amaurosis, visual confabulations, disorientation and confusion.

Medical history included arterial hypertension, type 2 diabetes mellitus, infectious endocarditis with residual mitral lesion surgically repaired and coronary artery bypass. The patient had no history of alcohol consumption. Home medications were insulin, double antiplatelet therapy and antihypertensive drugs.

A month ago the patient was admitted to the hospital due to a decrease in visual acuity, especially in the right eye, with an increase in erythrocyte sedimentation rate and weight loss. He was diagnosed with suspicion of temporal arteritis and treated with glucocorticoids. At the time of consultation in corticoid-descending regimen.

In the clinical examination upon arrival at the emergency room the patient was confused and disoriented, with bilateral amaurosis, visual confabulations but fluent and coherent speech with conserved strength and sensitivity symmetrically. An emergency cranial computed tomography (CT) was performed and no acute intracranial alterations were observed. Blood testing revealed a serum ammonia level of 289 μmol/L (normal range, 9 to 32 μmol/L), with normal hepatic enzymes and normal coagulation study. Electroencephalogram showed generalized slowdown.

The patient was admitted to the intensive care unit because the mental state gradually changed to a deep coma (GCS = 5) and mydriatic pupils slowly reactive. His vital signs were stable. Endotracheal intubation was performed and the brain scan was repeated 24 h after the first one because of the progressive coma and mydriatic pupils, not present at the first evaluation in the emergency room. The brain CT scan showed left frontal cortical hematoma (36 × 26 × 33 mm) with mild perilesional edema and minimal mass effect with no midline shift. The patient had no history of traumatic brain injury and no signs of cerebral edema nor underlying vascular malformations or aneurysms were observed. The intracranial hematoma was managed conservatively.

Due to persistently elevated level of ammonia continuous renal replacement therapy was initiated. The next few days, after forcing extrarenal purification therapy, the ammonium values progressively decreased but never until the normal values, and when the extrarenal therapy subsided, the ammonium increased again. A CT angiography found a portosystemic shunt due to gastrorenal shunt that communicates the short gastric vessels with the left renal vein, without signs of liver cirrhosis. Other causes of hyperammonemia were ruled out, such as pharmacological, liver cirrhosis, gastrointestinal bleeding or infections (Table 1).

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Table 1
Differential diagnosis of hyperammonemia in adults

Liver disease/portal hypertension: cirrhosis, schistosomiasis…
Gastrointestinal bleeding
Renal disease: distal renal tubular acidosis, hemodialysis (fluid overload)
Portosystemic shunt
Renal disease
Urinary tract infections (*Proteus mirabilis*)
Septic shock
Ureterosigmoidostomy
Parenteral nutrition
Reye's syndrome
Chemotherapy
Organ or bone marrow transplantation
Drugs: valproic acid, barbiturates, alcohol, diuretics…

Following bilateral femoral vein access, two 6.0-French curved tip sheaths were inserted in the gastrorenal shunt, one proximal and the other one distally (Fig. 1). Through the distal sheath, a 14 mm Amplatzer vascular plug (Abbott Vascular International, Diegem, Belgium) was deployed and through the proximal one, the gastrorenal shunt was embolized with 0.035 in. Interlock coils (Boston Scientific, Massachusetts). No immediate complications were observed.

After the embolization, continuous renal replacement therapy was discontinued. Within the next 24 h the serum ammonia levels decreased to normal values (23 μmol/L) and maintained (Fig. 2). The patient's mental state began to improve gradually and he achieves complete consciousness in the next few days. His major neurologic sequelae

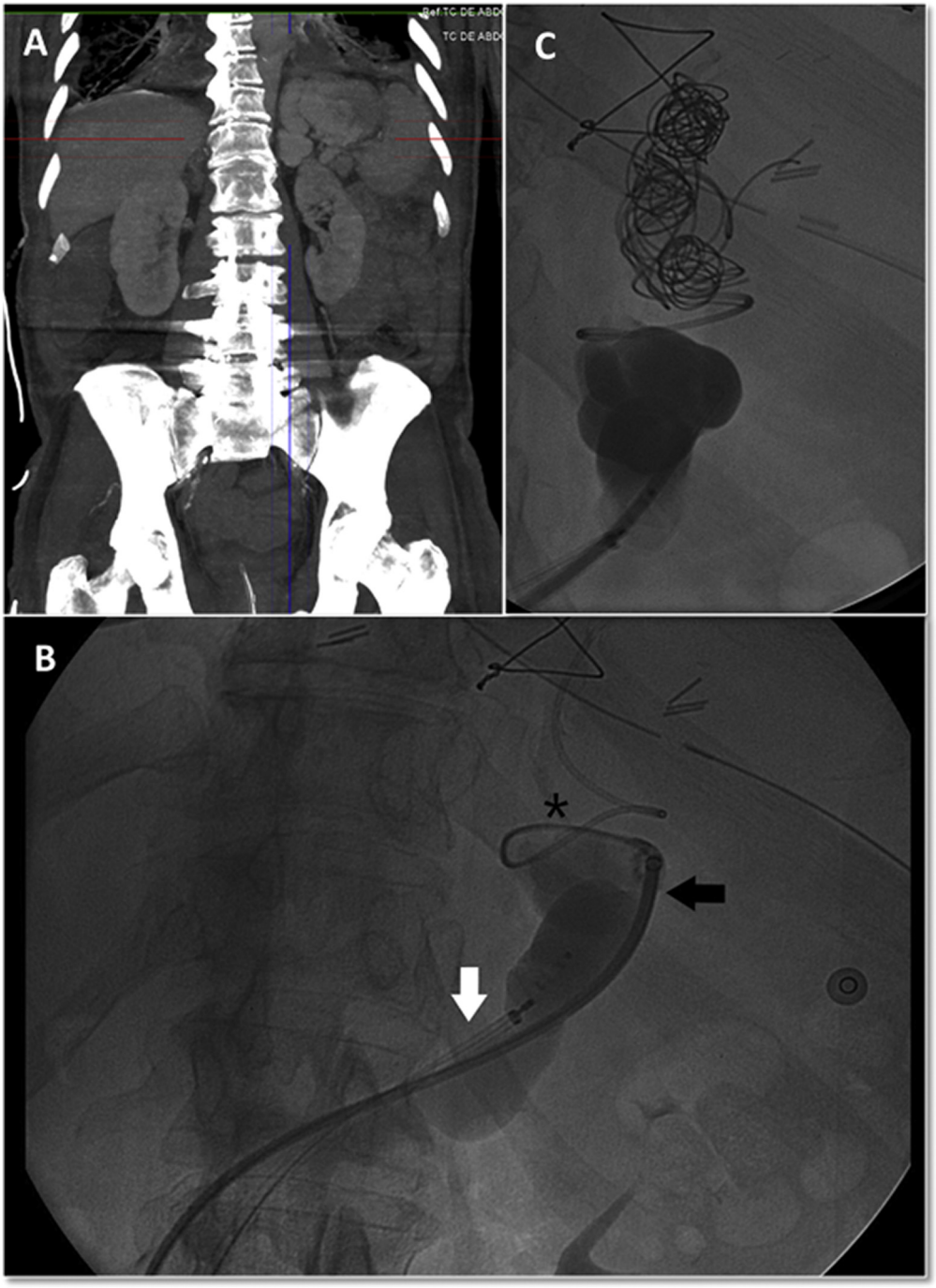

Fig. 1. A. Coronal reformatted abdomen computed tomography before the intervention. Perisplenic collateral vessels are prominent above the left kidney. B. Fluoroscopic image of the BRTO intervention. A double sheath in the gastrorenal shunt is inserted through the femoral veins. One sheath is in proximal position (black arrow) and a microcatheter is used for coil deployment (asterisk). The other sheath is in distal position (white arrow); a vascular plug is inserted for embolization. C. Fluoroscopic image. After proximal embolization with coils and distal embolization with vascular plug a venography is performed to confirm complete occlusion of the efferent shunt.

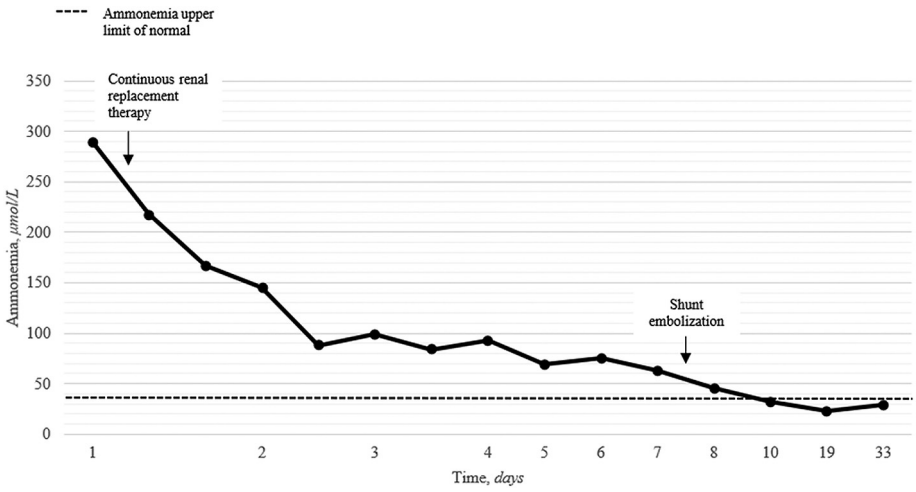

Fig. 2. Line-graph shows serum ammonia levels (in μmol/L) during the hospitalization.

were concordant with the left frontal hematoma and he was discharged from the intensive care unit.

2. Discussion

The most well-known etiology of hyperammonemic encephalopathy is liver failure and portal hypertension, but should also be considered even if there are no signs of cirrhosis [1]. A portosystemic shunt acts as a bypass in the ammonium circulation that must be eliminated by the liver through the portal vein, but ends in the inferior vena cava system and, therefore, in the general circulation and the brain, producing altered mental status.

Hyperammonemic encephalopathy should be considered in all unexplained altered mental status [2]. Severe increase ammonia values could be life threatening, especially in values above 200 μmol/L due to cerebral edema and alteration of cerebral self-regulation with high risk of ischemia or intracranial hemorrhage.

So, the shunt closure is an intervention to relief of encephalopathy symptoms. Balloon-occluded Retrograde Transvenous Obliteration (BRTO) was first described to close the splenorenal shunt, using different sclerosing agents under indwelling balloon occlusion catheter [3-5]. The vascular Plug-Assisted Retrograde Transvenous Obliteration (PARTO) is a newly modified BRTO, where a balloon occlusion catheter is replaced by vascular plug/coils to minimize some of the complications associated with the balloon catheter [6]. The proximal closure of the splenorenal shunt is performed with coils through a 5 french catheter and the distal closure is achieved with a vascular plug. With this technique a successfully thrombosis of the gastrorenal shunt and gastric varices is induced. The vascular PARTO can be quickly performed with high technical success rate and clinical efficacy for the treatment of the splenorenal and/or gastrorenal shunt [6]. The most frequent complications include bleeding, other venous thrombosis, secondary portal hypertension and persistent neurological symptoms.

In this case we report a severe serum ammonia elevation that induced an altered mental status and cerebral self-regulation with an intracranial hemorrhage as an outcome, due to gastrorenal shunt treated successfully by vascular PARTO.

Conflict of interests

The authors declare that they have no conflict of interest.

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