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CASE IMAGE

Adult-onset noncirrhotic hyperammonemic encephalopathy probably caused by splenorenal shunts

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Abstract

This case illustrates adult-onset noncirrhotic hyperammonemic encephalopathy, which is most likely caused by splenorenal shunts and is a rare but potentially fatal cause of altered mentation in the critical care setting. Splenorenal shunts should be considered as a differential in cases of hyperammonemic encephalopathy without liver cirrhosis.

KEYWORDS

brain diseases, congenital abnormalities, emergency medicine, hyperammonemia, splenorenal shunt

A 66-year-old man presented to an emergency department due to altered mentation that progressed in 2 days. The patient was lethargic and did not respond to commands. The abdomen was soft and mildly distended. The serum ammonia level was 196 μg/dl (normal range, 12–66 μg/dl). An abdominopelvic computed tomography scan revealed intestinal obstruction due to a right diaphragmatic hernia, and splenorenal shunts discovered incidentally without signs of liver cirrhosis. Figure 1 reveals a splenorenal shunt between the left renal vein and splenic vein via three-dimensional computed tomography scan. The patient underwent exploratory laparotomy, which included a 25 cm transverse colectomy. On day 2, his ammonia levels decreased to 99 μg/dl. He became alert and could follow commands.

Upper endoscopy revealed no gastroesophageal varices. Amino acid analysis for urea cycle revealed no abnormalities; thus, he was diagnosed with noncirrhotic hyperammonemic encephalopathy, most likely due to splenorenal shunts,[1,2] which were treated with branched-chain amino acids, lactulose, and kanamycin sulfate without any endovascular or surgical intervention. He did not, however, have congenital heart disease, polysplenia,

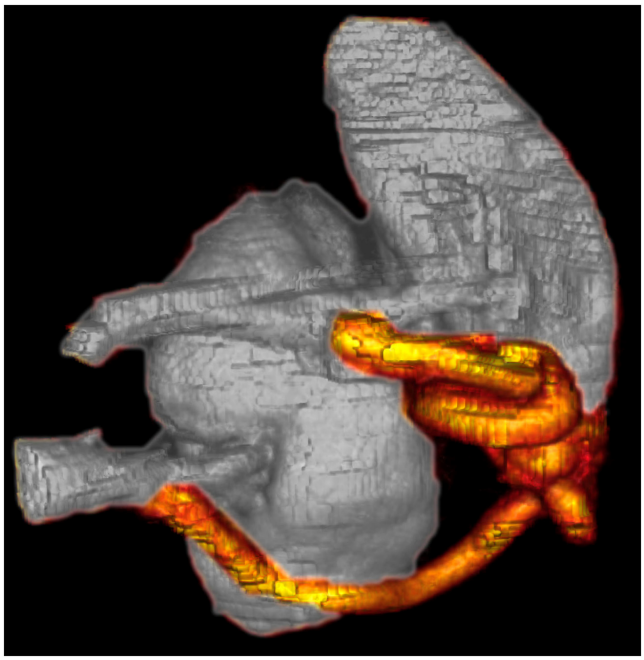

FIGURE 1 Three-dimensional reconstruction of the abdominal computed tomography on admission, revealing a splenorenal shunt between the left renal vein and splenic vein

or skeletal anomalies. He was discharged on day 33 with mild cognitive dysfunction and normal ammonia level, indicating persistent cognitive impairment associated with hyperammonemic encephalopathy.[1,2]

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None.

CONFLICTS OF INTEREST

The authors declare that they have no conflict of interest.

AUTHOR CONTRIBUTION

KY cared for the patient and initiated and wrote the manuscript. NY collected, analyzed, and interpreted the clinical data, and revised the manuscript. TT interpreted the clinical data and critically reviewed the manuscript. KS cared for the patient, interpreted the clinical data, and critically reviewed the manuscript. All authors of this paper have read and approved the final version submitted.

CONSENT

Published with the written consent of the patient.

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REFERENCES

1. Baiges A, Turon F, Simón-Talero M, et al. Congenital extrahepatic portosystemic shunts (Abernethy malformation): an international observational study. *Hepatology*. 2020;71:658-669.
2. Tang H, Song P, Wang Z, et al. A basic understanding of congenital extrahepatic portosystemic shunt: incidence, mechanism, complications, diagnosis, and treatment. *Intractable Rare Dis Res*. 2020;9:64-70.

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