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CASE IMAGE

Progress of pulmonary hypertension and high manganese levels through congenital portosystemic shunt closure

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Abstract

A 6-year-old girl with congenital portosystemic shunt presented with abnormal manganese levels and improving pulmonary hypertension even 1 year after shunt vascular ligation. As the progress after portal vein blood flow recovery varies among individuals, long-term follow-up of patients with congenital portosystemic shunt is needed.

KEYWORDS

arterial pressure, mesenteric artery, mesenteric veins, ventricular septum

1 | CASE DESCRIPTION

A 6-year-old girl who underwent surgery for congenital heart disease (ventricular septum defect patch closure and patent ductus arteriosus ligation) at the age of 1 year showed electrocardiogram and echocardiography findings of pulmonary hypertension (PH) during routine checkup. Elevated manganese (Mn) levels were detected, and abdominal enhanced computed tomography (CT) revealed that the superior mesenteric and splenic veins were inverted and connected to the left renal vein (Figure 1A). These tests were performed for the first time in this patient. Extrahepatic and intrahepatic portal veins were detected in the shunt vessel occlusion test (Figure 1B). Under the diagnosis of congenital portosystemic shunt (CPSS), shunt vascular ligation was performed. Finally, abdominal enhanced CT and superior mesenteric artery angiography explicitly revealed the portal vein structure (Figure 1C,D). 6 months after the surgery, the mean pulmonary arterial pressure as well as ammonia and total bile acid levels returned to normal immediately; however, Mn level did not normalize even after a year despite consuming a Mn-restricted diet.

Although improvement in PH after CPSS ligation is controversial,[1] her PH improved immediately. Unlike in this case, high manganese levels typically normalize immediately after surgery.[2] As the progress after portal vein blood flow recovery varies among individuals, long-term follow-up as well as observation for any complications of CPSS are required.

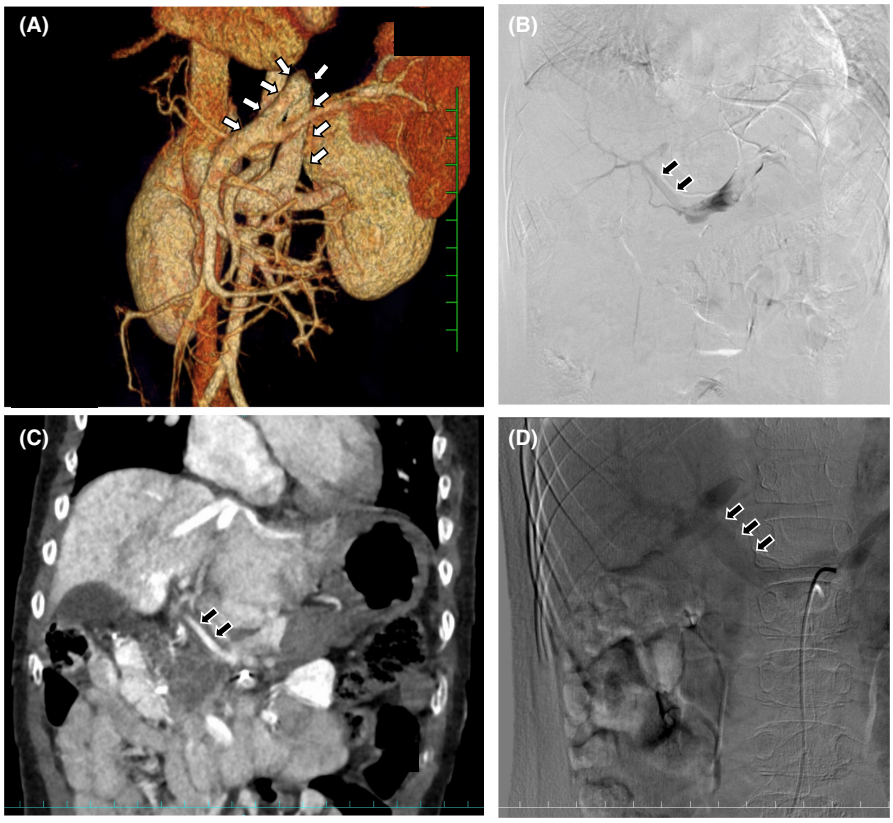

FIGURE 1 (A) Enhanced computed tomography (CT) revealed that the superior mesenteric and splenic veins were inverted and connected to the left renal vein. (B) Extrahepatic and intrahepatic portal veins were detected in the shunt vessel occlusion test. (C and D) Abdominal enhanced CT and superior mesenteric artery angiography explicitly revealed the portal vein structure after shunt vascular ligation. White arrows, shunt blood vessels; black arrows, portal vein.

AUTHOR CONTRIBUTIONS

Toshihiko Kakiuchi was involved in all stages of managing the patient and wrote the manuscript. Yuta Baba, Nobuya Minematsu, Hirohito Doi, and Takashi Kumamoto were involved in all stages of management. Takashi Kumamoto analyzed the data and collaborated as a reviewer. All authors read and approved the final manuscript.

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We would like to thank the patient's parents for providing consent for publication.

CONFLICT OF INTEREST

None.

DATA AVAILABILITY STATEMENT

All data generated and analyzed during this study are included in the published article.

ETHICAL APPROVAL AND PATIENT CONSENT

Written informed consent was obtained from the patient's parents. This report is exempt from ethical approval because it is an observational report after the current care.

CONSENT

Consent was obtained from parents/guardians.

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