Digestive and Liver Disease 57 (2025) 1504–1505

Contents lists available at ScienceDirect

Digestive and Liver Disease

journal homepage: www.elsevier.com/locate/dld

Image of the Month

Severe abdominal wall varices and acute-on-chronic liver failure caused by the familial clustering of Budd–Chiari syndrome

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A 54-year-old man presented with jaundice and abdominal pain two months ago. He was initially diagnosed with obstructive jaundice by the local hospital. However, endoscopic retrograde cholangiopancreatography combined with endoscopic nasobiliary drainage proved ineffective. Physical examination revealed prominent abdominal wall varices with blood flowing upward from below (Fig. 1), concurrent with severe jaundice and hepatic encephalopathy. Laboratory tests indicated total bilirubin levels of 576.8 μmol/L (normal range: 0–26 μmol/L), direct bilirubin levels of 410.4 μmol/L (normal range: 0–4 μmol/L), international normalized ratio of 1.66 (normal range: 0.8–1.2), blood ammonia levels of 88.4 umol/L (normal range: 0.0–30.0 umol/L), and antithrombin III levels of 47.0 % (normal range: 75.0–125.0 %). Magnetic resonance imaging showed obstruction in the inferior vena cava (Fig. 2A); cirrhosis; and abdominal wall varices (Fig. 2B). Bone marrow examination ruled out myeloproliferative disorder. The patient has two siblings who also suffer from Budd–Chiari syndrome (with one being deceased). Their clinical and imaging manifestations are similar (Fig. 2C-D). Genetic testing conducted on the patient, his surviving sibling and his parents excluded hereditary thrombophilia. The patient underwent artificial liver support system and anticoagulation therapy, opting against interventional procedures and liver transplantation, leading to only limited improvement in liver function. Acute-on-chronic liver failure caused by the familial clustering of Budd–Chiari syndrome is rare and necessitates prompt recognition and intervention [1].

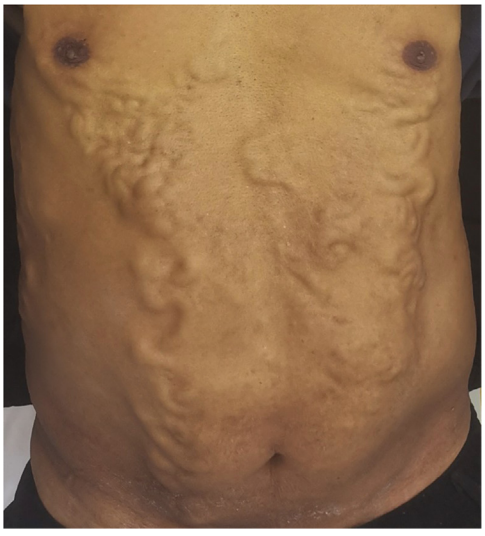

Fig. 1. Physical examination revealed prominent abdominal wall varices and severe jaundice in this patient.

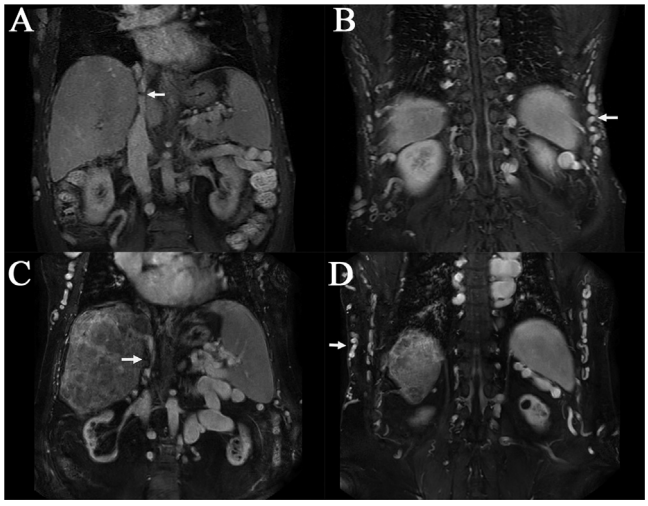

Fig. 2. Magnetic resonance imaging findings of this patient and his deceased brother. A. This patient has Budd–Chiari syndrome. B. This patient has prominent abdominal wall varices. C. The deceased brother of this patient had Budd-Chiari syndrome. D. The deceased brother of this patient had prominent abdominal wall varices.

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https://doi.org/10.1016/j.dld.2025.03.030
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Informed consent

The informed consent was obtained from the patients and their relatives for the publication of their information and imaging.

Conflict of interest

None.

Author contribution

Chen Li: Conceptualization, Data curation, Formal analysis, Methodology, Writing – original draft. **Hong Guo:** Conceptualization, Data curation, Formal analysis, Methodology, Writing – original draft. **Yuhui Peng:** Conceptualization, Investigation, Methodology. **Haibin Su:** Conceptualization, Methodology, Project administration, Writing – review & editing.

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