Large abdominal cutaneous bypass that developed naturally in Klippel-Trénaunay syndrome

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DESCRIPTION

A 73-year-old woman with liver cirrhosis and end-stage renal failure receiving haemodialysis presented to our hospital with fever. No signs of hepatitis B, hepatitis C, alcohol consumption, non-alcoholic steatohepatitis or autoimmune disease were observed. She was diagnosed with congestive hepatopathy based on liver biopsy and hepatorenal syndrome at 63 and 65 years of age, respectively, and haemodialysis was initiated. Her body temperature was 38.3°C, but her other vital signs were normal. On physical examination, a large cutaneous vein crossed her lower abdomen (**figure 1A**, arrowhead). The left leg showed pitting oedema with erythema, warmth and tenderness. Her left fingers exhibited oedema (**figure 1B**). Port-wine stains were observed on the extremities and lower back, which were present since birth (**figure 1C**). Laboratory findings showed the following: white cell count, $6.1 \times 10^9/L$ (56.3% neutrophils, 2.8% eosinophils, 0.3% basophils, 22.5% lymphocytes, 18.1% monocytes); haemoglobin level, 140 g/L; platelet count, $56 \times 10^9/L$; prothrombin time, 67.5%; albumin level, 1.8 mg/dL; total bilirubin level, 1.53 mg/dL; blood urea nitrogen level, 56 mg/dL; creatinine level, 6.43 mg/dL and C reactive protein level, 7.59 mg/dL. Radiography showed bony hypertrophy at the level of the proximal and distal interphalangeal joints (**figure 1B**). Contrast-enhanced CT showed that the vena cava and left external iliac vein

were duplicated and occluded, respectively. Left leg veins flowed to the right external iliac vein through a large abdominal cutaneous vein. She was diagnosed with cellulitis in her left leg caused by phlebostasis of Klippel-Trénaunay syndrome (KTS). CT or MRI of the head showed no evidence of intracranial arteriovenous malformation. Although left leg inflammation was improved after cefazolin administration, left leg oedema persisted.

KTS is characterised by a triad of capillary malformations, venous malformations and limb overgrowth.¹ The estimated incidence is 2–5 in 100 000 individuals without a sex difference.² Capillary malformations are found in 98%, varicosities or venous malformations in 72%, and limb hypertrophy in 67% of patients with KTS.³ Other clinical symptoms, such as thrombosis, bleeding, superficial thrombophlebitis, ulceration and pain as a result of chronic venous insufficiency may also occur.⁴ The prognosis depends on gastrointestinal bleeding and pulmonary embolism caused by venous malformations. Recently, intracranial vascular malformations have been reported.⁵ Brain imaging tests should be included in KTS to prevent critical brain haemorrhage. Although CT and MRI are used as the imaging modalities, ultrasonography is recommended especially in infants, considering radiation exposure, cost and unnecessary sedation. The treatment should be conservative using analgesics and antibiotics; however, surgical management may be applied in cases of severe chronic venous insufficiency. Iliac vein occlusion is treated by a femoral cross-over bypass using the great saphenous vein (Palma procedure).⁶ In this case, a large abdominal cutaneous bypass developed naturally without surgery. Careful visual examination is needed because venous malformations may be detected on the abdominal surface in addition to capillary malformations and varicosities.

Figure 1 (A) A large abdominal cutaneous vein from the veins in the left leg to the right external iliac vein (arrowhead) (B): bony hypertrophy at the level of the left proximal and distal interphalangeal joints (C) port-wine stains on the trunk.

Learning points

- ▶ A large abdominal cutaneous bypass can develop naturally in Klippel-Trénaunay syndrome patients.
- ▶ Venous malformations may be observed on the abdominal surface by careful visual examination.

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Disclaimer Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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REFERENCES

- 1 Wassef M, Blei F, Adams D, *et al*. Vascular anomalies classification: recommendations from the International Society for the study of vascular anomalies. *Pediatrics* 2015;136:e203–14.
- 2 Alwalid O, Makamure J, Cheng Q-G, *et al*. Radiological aspect of Klippel-Trénaunay syndrome: a case series with review of literature. *Curr Med Sci* 2018;38:925–31.
- 3 Jacob AG, Driscoll DJ, Shaughnessy WJ, *et al*. Klippel-Trénaunay syndrome: spectrum and management. *Mayo Clin Proc* 1998;73:28–36.
- 4 Lee A, Driscoll D, Gloviczki P, *et al*. Evaluation and management of pain in patients with Klippel-Trenaunay syndrome: a review. *Pediatrics* 2005;115:744–9.
- 5 Pérez-Alfayate R, Martínez-Moreno N, Rosati SD, *et al*. Klippel-Trenaunay-Weber syndrome associated with multiple cerebral arteriovenous malformations: usefulness of gamma knife stereotactic radiosurgery in this syndrome. *World Neurosurg* 2020;141:425–9.
- 6 Oderich GS, Panneton JM, Noel AA, *et al*. Relief of iliofemoral vein occlusion with the Palma bypass in a patient with Klippel Trenaunay syndrome. *Ann Vasc Surg* 2003;17:449–55.

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