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Rare disease

An extraordinary cause for deep venous thrombosis

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Summary

The authors present a case of a congenital absence of the infrarenal inferior vena cava in an 18-year-old man showing symptoms of deep venous thrombosis of the left leg. The congenital absence of the inferior vena cava is typically asymptomatic and is commonly reported as a fortuitous finding. Abnormalities of the inferior vena cava are risk factors contributing to the development of deep venous thrombosis. The absence of vena cava is underestimated in patients with deep venous thrombosis because in some cases compression B-mode ultrasonography will not reveal the condition. CT should be made available for all young patients with idiopathic deep venous thrombosis.

BACKGROUND

The absence of the inferior vena cava is an uncommon congenital anomaly. However, it is an important risk factor contributing to the development of deep venous thrombosis especially in young adults. In our case, ultrasound, the procedure of choice for diagnosing deep venous thrombosis, did not reveal the extent of thrombosis and the abnormal anatomy. CT provided us this information. Therefore, in all young patients presenting with deep venous thrombosis a CT scan should be made.

CASE PRESENTATION

An 18-year-old man was admitted to our hospital with complaints of a painful, red and warm left leg. He had difficulty walking. The patient had been taking non-steroidal anti-inflammatory drug medication for 10 days for complaints of pain in the lower back and lower abdomen for several weeks. He had no complaints of dyspnoea or chest pain. There was no history of immobilisation, recent flying, trauma, surgery or smoking. The medical history did reveal a preterm birth and a thrombosis of the renal vein in the neonatal phase of which no origin was found. There was no significant family history on thromboembolism or clotting disorders.

Physical examination revealed a painless mass in the right lower quadrant of the abdomen. Liver and spleen could not be palpated. The left leg was swollen, red and warm and the calf was on tension. Arterial pulsation was not compromised.

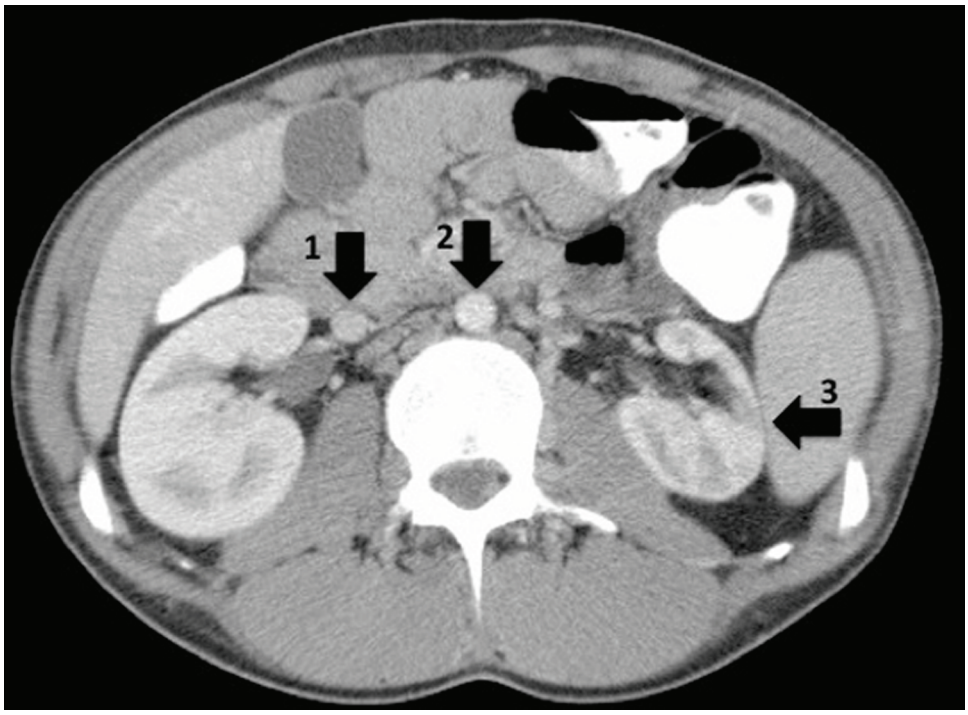

Figure 1 CT scan: Enlarged right subcardinal vein (1), Aorta (2), Agenesis of the left kidney (3).

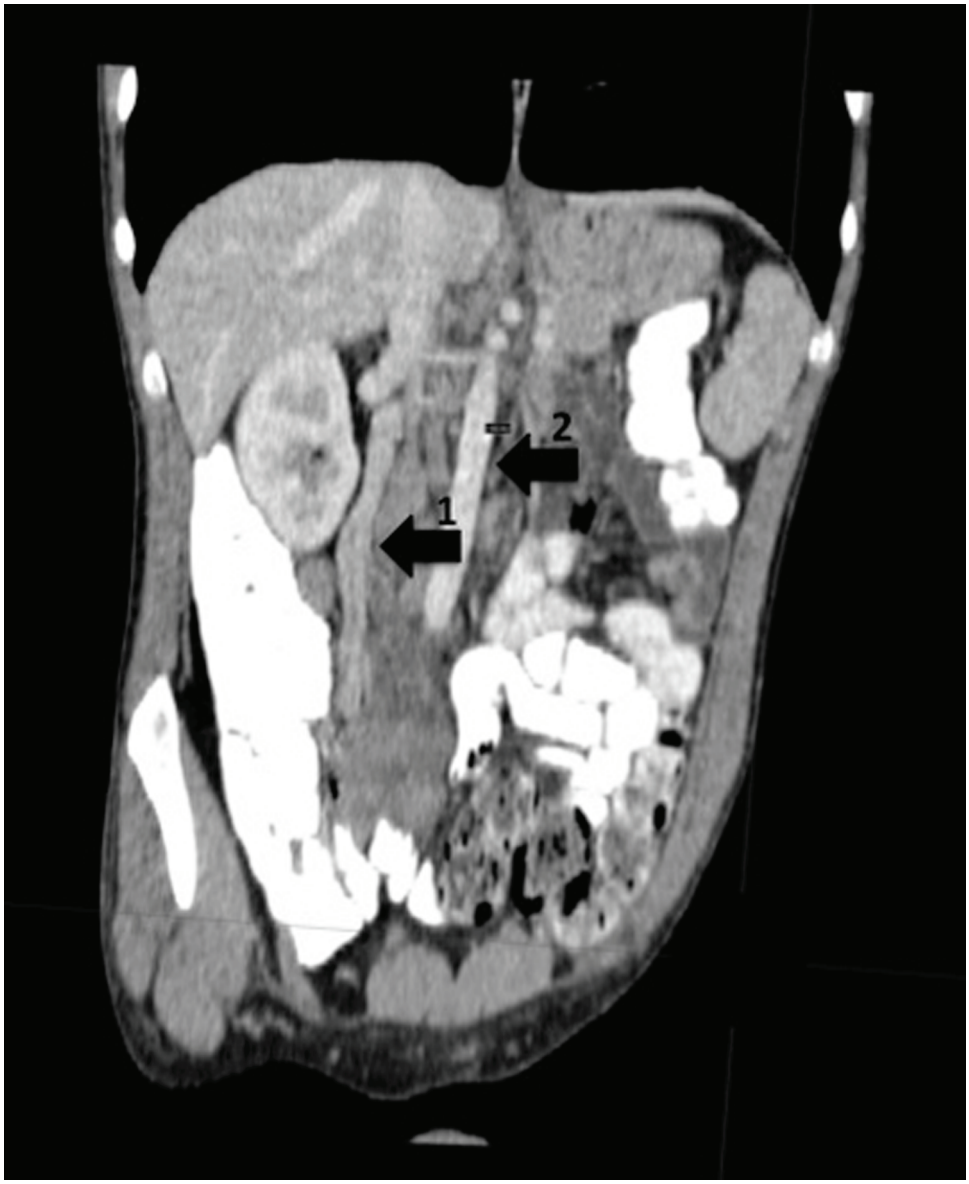

Figure 2 CT scan: Enlarged right subcardinal vein (1), Aorta (2).

INVESTIGATIONS

Routine laboratory tests were normal. Ultrasound showed a venous thrombosis of the left leg up to the level of the common iliac vein. In the lower abdomen the inferior vena cava could not be identified and the mass in the right lower abdomen was identified to be a thrombosed varicose vein. Ultrasound of the right leg revealed no thrombosis.

A contrast-enhanced CT was performed and demonstrated the absence of the infrarenal inferior vena cava. Both common iliac veins reach the suprarenal vena cava through a collateral network of vessels consisting of dilated azygos vein and persisted subcardinal veins. On the right the subcardinal vein was thrombosed, enlarged and tortuous (figure 1 and 2). The right internal iliac as well as the left common iliac vein were thrombosed. Agenesis of the left kidney was noted. No abnormalities were seen on the organs in the upper abdomen.

TREATMENT

The patient was treated with compression bandage application and subcutaneous low weight heparin followed by coumarin treatment.

OUTCOME AND FOLLOW-UP

The following day clinical improvement was seen and our patient was discharged. He was recommended lifelong anticoagulation treatment.

DISCUSSION

Congenital interruption of the inferior vena cava is an extremely rare condition and is present in 0.3–0.6% of the general population.[1 2] During the 6th to 8th week of embryologic development, the infrarenal inferior vena cava derives from fusion of three sets of veins (eg, the posterior cardinal, subcardinal and supracardinal veins). If

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these veins fail to fuse either an anomalous inferior vena cava emerges or the inferior vena cava remains absent.[3] In the absence of the infrarenal vena cava the blood from the lower extremities drains to the heart via a deep venous collateral system. This system consists of azygos and hemiazygos veins with compensatory enlargement.

Absence of the vena cava is generally asymptomatic and most cases are reported as a fortuitous finding during routine radiological screening, abdominal surgery or cardiac catheterisation.[1] If the collateral system fails despite compensatory enlargement, venous stasis will emerge and, subsequently, deep venous thrombosis will form. In around 5.3% of cases of deep venous thrombosis in young adults (eg, below the age of 30 y) absence of the inferior vena cava is present.[2]

The procedure of choice for the diagnosis of deep venous thrombosis is compression B-mode ultrasonography. However, in some cases ultrasound will not reveal the absence of the vena cava. CT (with intravenous contrast) and a MRI scan are more reliable techniques for diagnosing anomalies of the vena cava.[4]

Treatment of deep venous thrombosis due to an anomalous inferior vena cava is standard anticoagulation. Due to its rareness the optimal duration of anticoagulation treatment has not been determined yet.[1] In some cases, surgical reconstruction of the inferior vena cava is necessary.[3]

In conclusion, whenever confronted with a case of a deep venous thrombosis in young adults, especially in the absence of precipitating factors or clotting disorders, absence or anomalies of the inferior vena cava should be considered Therefore, in addition to ultrasonography a CT with intravenous contrast should be made.

Learning points

- The absence of the inferior vena cava is an uncommon congenital anomaly.
- Abnormalities of the inferior vena cava are important risk factors contributing to the development of deep venous thrombosis especially in young adults.
- The absence of vena cava is underestimated in patients with deep venous thrombosis because in some cases compression B-mode ultrasonography will not reveal the condition and a CT scan should be undertaken.

Competing interests None.

Patient consent Obtained.

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