## **CE - MEDICAL ILLUSTRATION**



## Acute deep vein thrombosis (DVT) of the lower limbs in a 32-year-old man with chronic hypoplasia of the inferior vena cava (HIVC) without risk factors

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The anomalies of the inferior vena cava (IVC) have an incidence between 0.2 and 1 % in the general population [1]. The normal average diameter of the IVC is 17.5 mm (up to 25 mm maximum) and the spectrum of anatomical abnormalities of the IVC (e.g., absence or interruption of the vessel or variation in diameter) can be responsible for serious hemodynamic effects to the venous circulation of the lower limbs (i.e., venous stasis), and be a risk factor for the development of deep vein thrombosis (DVT). The association between hypoplasia of the IVC (HIVC) and DVT has been previously reported, even though the association of absence of the IVC and DVT is more frequently recorded [2]. Hypoplasia of the inferior vena cava should be taken into account when an unexplained DVT occurs in young patients, as DVT can occur in up to 5 % of young adults with IVC anomalies [3].

A 32-year-old man was referred to our Unit of Internal Medicine because of acute and intense back pain. History was unrevealing for risk factors related to venous disease, but the man had remained immobilised 3–4 days because of the intense pain. After admission, he underwent full (abdominal and pelvic) ultrasound examination, which revealed bilateral proximal (iliac–femoral veins) DVT (Fig. 1a, b; see Fig. 1c for comparison) and severe dilatation of the superficial venous circle of the lower limbs. Laboratory investigations for thrombophilic defects (including factor V Leiden, prothrombin G20210A, mutations

A1298C and C8677T, proteins C and S and antithrombin III) were all negative.

A computed tomography (CT scan) study of the superior abdomen showed hypoplasia of inferior vena cava (diameter  $11 \times 5$  mm) along with bilateral thrombosis of the iliac and hypogastric veins; the right iliac vein was dilated and the paralumbar veins were hypertrophic; dilatation of the right hypogastric venous circle was present (Fig. 2a; see Fig. 2b for comparison).

Treatment with subcutaneous low molecular weight heparin was initiated, and shortly followed by oral anticoagulant therapy (INR range 3.0) with application of stocking bandage compression to the lower limbs. Currently, the man is still under follow-up at our unit: his general condition is good with no further recorded episodes of thrombosis; he is still taking oral anticoagulant therapy.

Anomalies of the IVC are a rare finding in clinical practice, but should be suspected when an unexplained DVT presents in a young adult, particularly in the absence of risk factors such as inherited thrombophilia. The main findings in patients with DVT and HICV are summarised in Table 1. Notably, dilatation of the ascending lumbar veins, paravertebral venous plexus, azygous and hemiazygous veins are typical imaging findings of these patients. The hemodynamic data on venous return from the lower part of the body are crucial for differentiating acute and chronic anomalies of the IVC.

Although ultrasonography is useful to demonstrate anomalies of the IVC, by revealing the lack of continuity of the IVC segment and the presence of collateral veins in the abdomen or in the retroperitoneal region, the gold standard of imaging is a CT scan (Fig. 2) or magnetic resonance imaging studies, which are more sensitive and specific in showing the anomalies of the IVC, and to explain the



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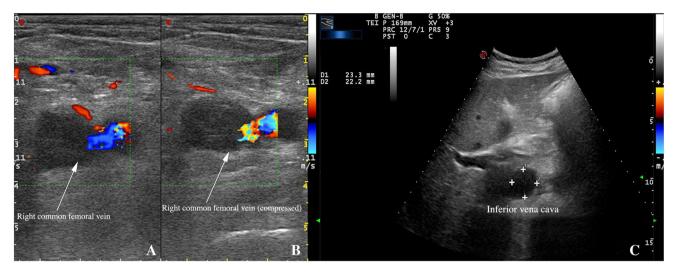


Fig. 1 Colour assisted (a) and colour assisted compression (b) ultrasound examination of the lower limbs in the reported patient showing deep venous thrombi in the right common femoral vein; normal deep venous vessels in an otherwise healthy 35-year-old patient (c)

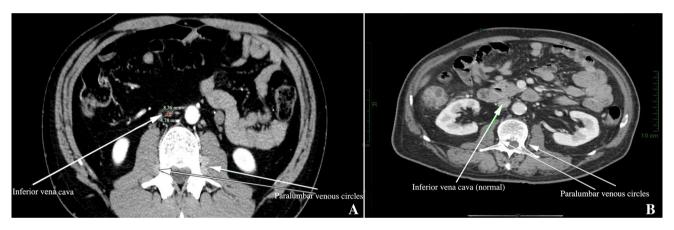


Fig. 2 Axial computerised tomography (CT) scans of the abdomen: **a** in the reported patient shows hypoplasia of the inferior vena cava and marked dilatation of the paralumbar veins (*white arrows*), **b** in a normal vena cava of an otherwise healthy 35 year-old man

Table 1 Common clinical, haemodynamic and imaging findings in patients with DVT and HICV

Age at presentation

Symptoms that should raise suspicion of DVT + HIVC

DVT presentation

Accompanying imaging findings

Hemodynamic tools

Young adulthood

Back pain and/or abdominal pain or flat before the DVT

Bilateral vein thrombosis (elevated frequency)

Dilatation of the ascending lumbar veins, of the paravertebral venous plexus, and of the azygous and hemiazygous veins

Venous return from the lower regions of the body (crucial)

pathophysiology and pathogenic causes underlying the DVT in lower limbs [4, 5].

Despite the increasing availability of such imaging techniques, the recorded cases of DVT and HICV are still limited. The most recent epidemiological data show that the general incidence of DVT (with or without pulmonary embolism) is around 160–182 cases/1,00,000/year [4].

Physicians need to be aware that, especially in a young patient, that if a venous thrombotic disease manifests without an overt cause (i.e. vascular anomalies, hypercoagulability, prolonged stasis) the occurrence of an anomaly of the IVC (e.g., agenesia, atresia, hypoplasia) must be taken into consideration, and that the current imaging techniques can easily detect such anomalies [5].



In young patients, back pain and a history of prolonged immobilisation represents crucial features that suggest the presence of a DVT and IVC. The current imaging tools (e.g., US, CT scan, MRI) are highly sensitive and specific in the diagnostic work-up. Early treatment avoids complications.

## Compliance with ethical standards

**Conflict of interest** None of the authors has any potential financial conflict of interest related to this manuscript.

**Statement of human and animal rights** All procedures performed in this study involving human participants were in accordance with the ethical standards of the institutional committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

**Informed consent** Informed consent was obtained from all individual participants included in the study.

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