RESEARCH ARTICLE

Infrarenal inferior vena cava agenesis.

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Abstract

Congenital anomalies of the inferior vena cava are a rare malformation, and most of the time they are found incidentally. Compared to the general population, the incidence of deep vein thrombosis in young people between the age of 20 and 40 is 10 times lower, and it is in this age range where we should carry out a comprehensive study to search for probable anomalies, especially if there are no thrombotic risk factors, as in the case in point. As for treatment, there is no consensus on the appropriate treatment, although it seems that, after reviewing the evidence, the best option would be factor Xa inhibitors. Further studies are therefore necessary to confirm this assertion and, above all, to know the best option in asymptomatic cases, where findings have been incidental.

Introduction

The incidence of deep vein thrombosis (DVT) in the general population is 1 case per 1,000 inhabitants a year, whereas in young people between the age of 20 and 40 the incidence is 10 times lower.

Congenital anomalies of the inferior vena cava are a rare vascular malformation, with a prevalence of around 0.2 – 0.5% of the general population, and involve a failure between the 6th and 8th week of embryogenesis, when 3 pairs of embryonic veins (posterior cardinal, subcardinal and supracardinal)1,2 are formed.

As to its manifestation, the most common symptoms are chronic venous insufficiency in LL, PTE, DVT3,4 or even mimicking lumbar paravertebral masses due to the intense collateral circulation.5

We introduce the case of a 28-year-old male patient, who was admitted to the Vascular Surgery Department with a diagnosis of infrarenal inferior vena cava agenesis and secondary deep vein thrombosis (DVT) in the lower limbs (LL).

Case report

A 28-year-old male patient with no personal history of concern, except smoking, and no family history of thrombotic disease came to the hospital's Emergency Department due to non-specific abdominal discomfort described as "bloated feeling", dull pain in the mesogastrium and oedema in the LL for a month, for which he had consulted his primary care facility on several occasions.

Upon arrival at the Emergency Department, the physical examination revealed mild bilateral bimalleolar oedema, with no other significant findings. A complete blood test was performed, showing a D-dimer of 5,600 IU/l. In view of the suspicion of DVT in the LL, a Doppler echocardiography was performed, which reported filling defects inside the common femoral vein at the origin of the saphenous arch and, bilaterally, in both externaliliac veins and left varicocele. Due to these findings, he was admitted to the Vascular Surgery Department to complete the study.

During admission, a thorax-abdomen-pelvis CT scan was performed, with the following results: enlarged azygous vein (Figure 1), absence of inferior vena cava at infrarenal level, with a large amount of collateral circulation instead. Prominent collateral circulation in the lumbar paravertebral region. Bilateral common external and internal iliac veins with filling defects in their interior, acquiring a maximum calibre of 3 cm in the left common iliac vein and multiple pelvic varices (Figure 2).

Given the aforementioned findings, treatment with low-molecular-weight heparins and subsequent anticoagulation therapy with vitamin K antagonists was started after hospital discharge.

Figure 1

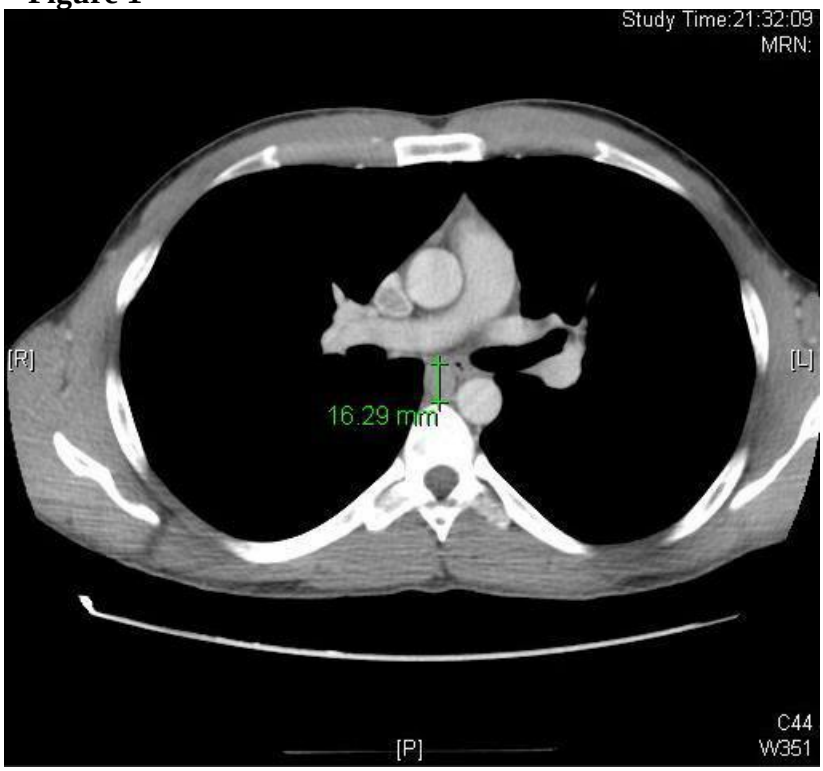

Figure 2

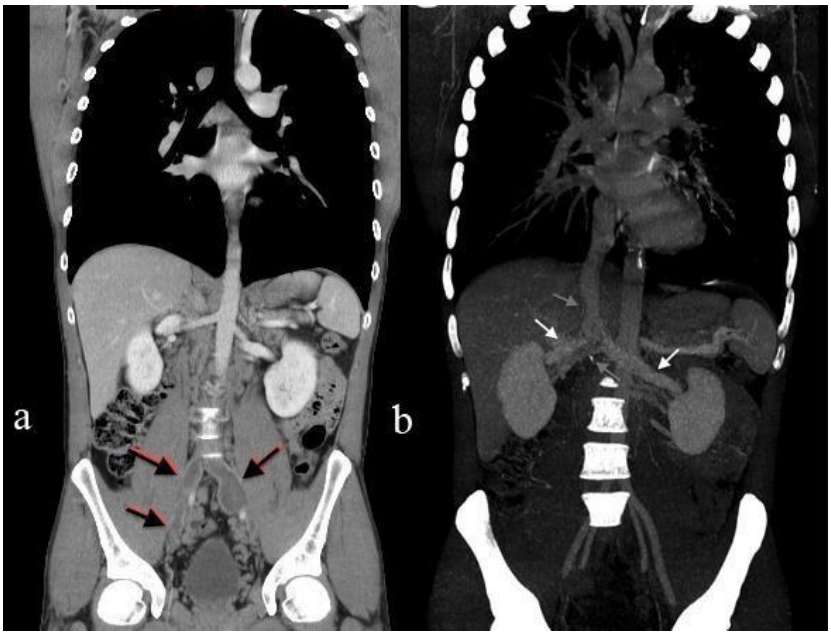

Figure caption: CT image *(sagittal section)* Inferior vena cava agenesis (Figure 1b grey arrow), increased lumbar paravertebral collateral circulation and pelvic varices (Figure 1a black arrow) and unaltered renal veins (Figure 1b white arrow).

Diagnosis

1. *Infrarenal inferior vena cava agenesis*
2. *Secondary bilateral deep vein thrombosis*

Discussion

Most patients with inferior vena cava vascular anomalies are asymptomatic, and findings are usually incidental. The most common manifestation is DVT; other less frequent forms are sciatic neuropathy due to venous compression caused by dilation of the epidural veins, obstructive pyelonephritis and especially pelvic congestion syndrome in women and varicocele in men.

It is therefore in young patients, especially males, where there are no identifiable risk factors for developing DVT that congenital inferior vena cava anomalies should be suspected and further diagnostic studies carried out.

Given the rare condition of this anomaly, there are currently no clinical guidelines for the treatment or for recommending prophylaxis for thrombotic events, when the findings are incidental. But the most appropriate in symptomatic patients would be to keep the patient on anticoagulation for more than 6 months, and although most cases go unnoticed and asymptomatic, relapses appear in patients when anticoagulation is discontinued.

We therefore believe that it would be reasonable and advisable to include diagnostic imaging tests when screening for vascular anomalies in young patients with DVT without identifiable risk factors, as in the case described above, since inferior vena cava agenesis is a risk factor,[6] especially when the iliac veins are involved[7].

Further studies will be necessary in asymptomatic patients and in those whose findings of congenital anomalies have been incidental, especially so as to agree on whether they are candidates to initiate and maintain anticoagulation or antiplatelet therapy for life[8] or simply for progress monitoring. The EINSTEIN CHOICE[9] showed that anticoagulation with rivaroxaban at a dose of 10 mg or 20 mg in patients with DVT prevents thrombotic events, compared to antiplatelet therapy with acetylsalicylic acid at a dose of 100 mg per day, without evidence of major bleeding episodes in the anticoagulant group. Based on the currently published evidence, it seems reasonable that in young patients with no identifiable risk factors and the only procoagulant factor being congenital vascular anomalies, the safer and more efficient treatment would be factor Xa inhibitors compared to vitamin K antagonists[10]. Although more studies are clearly needed to affirm that this would be the safest alternative in this type of patient.

Literature

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