

Congenital Agenesis of the Inferior Vena Cava: A Diagnosis to Mention in Front of a Deep Vein Thrombosis

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Authors' contributions

This work was carried out in collaboration among all authors. Author RD designed the study, performed the statistical analysis, wrote the protocol, and wrote the first draft of the manuscript. Authors MAS and KB managed the analyses of the study. Author RA managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Congenital agenesis of the inferior vena cava (IVC) is an extremely rare morphological anomaly in the general population; found in approximately 0.0005% to 1% [1]. It is a rare cause of deep vein thrombosis (DVT) and could be responsible for less than 5% of thromboses without a contributory factor in subjects under 30 years of age [2].

We present a case of DVT agenesis, demonstrated by a CT angiography, performed as part of the etiologic assessment of extensive, bilateral and recurrent deep venous thrombosis (DVT), in a young 24-year-old man.

We discuss the mechanisms of occurrence of thrombosis associated with IVC agenesis and their possible clinical and therapeutic features.

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1. INTRODUCTION

The embryological development of IVC is a complex process. Malformations of the inferior vena cava result from the development or abnormal regression of certain segments of the venous system, during embryonic life. Almost 60 different anomalies of the IVC have been described since 1793 [3]. The estimated incidence of these anomalies is 0.05 to 8.7% depending on the series [1]. Agnesis of the inferior vena cava (AIVC) is one of the rarest congenital anomalies of IVC. Two forms of agnesis can be found; complete or segmented. Prevalence was estimated from 0.0005% to 1% [1]. It mainly affects young subjects with a male predominance in more than 70% of cases [4]. We report a new observation of AIVC in a young

man with recurrent bilateral DVT of the lower limbs.

2. CASE PRESENTATION

A 24 year old man, with no medical histories, having presented a low back pain a month before his admission, radiating to the two lower limbs without systematized territory. The lumbar spine scanner showed an L4-L5 discal hernia, justifying the prescription of anti-inflammatory drugs and analgesics. At the onset of symptoms of oedema and pain in the two lower limbs, a venous Doppler showed bilateral deep vein thrombosis of the two lower limbs, affecting the leg veins, popliteal, femoral and iliac veins, right and left, as well as beginning of the IVC (Fig. 1a and 1b).

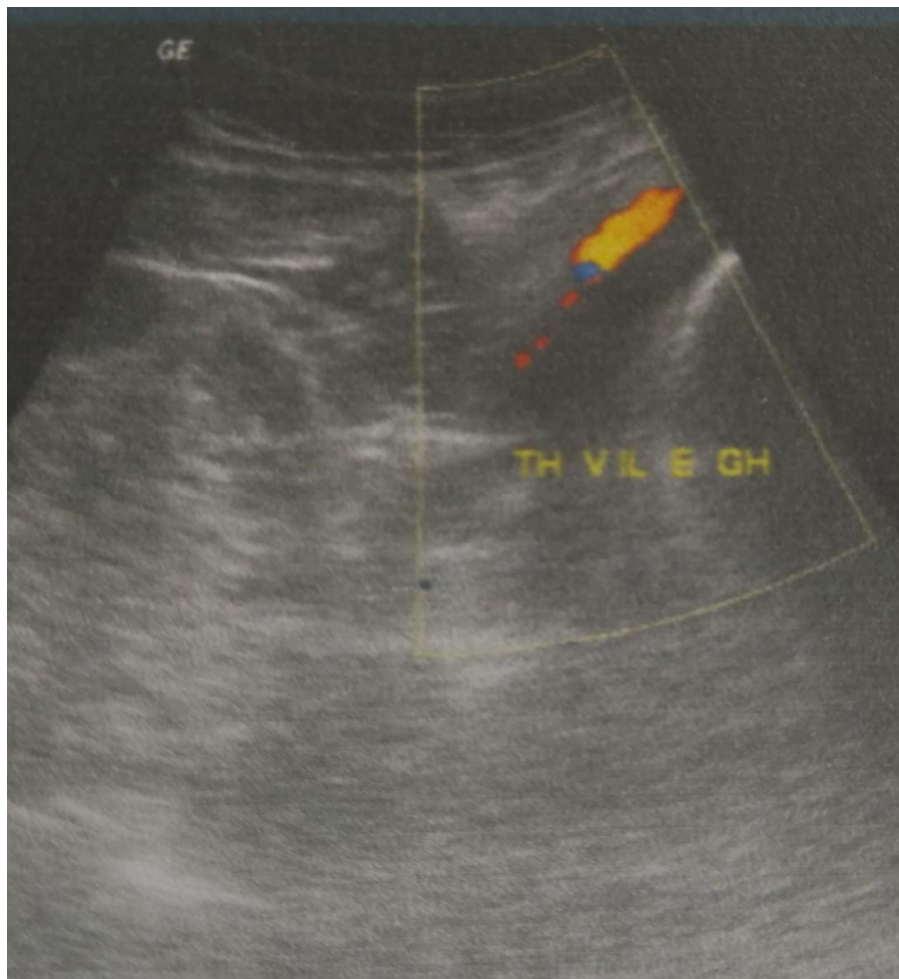


Fig. 1a. Doppler ultrasound showing thrombosis of the left external iliac vein

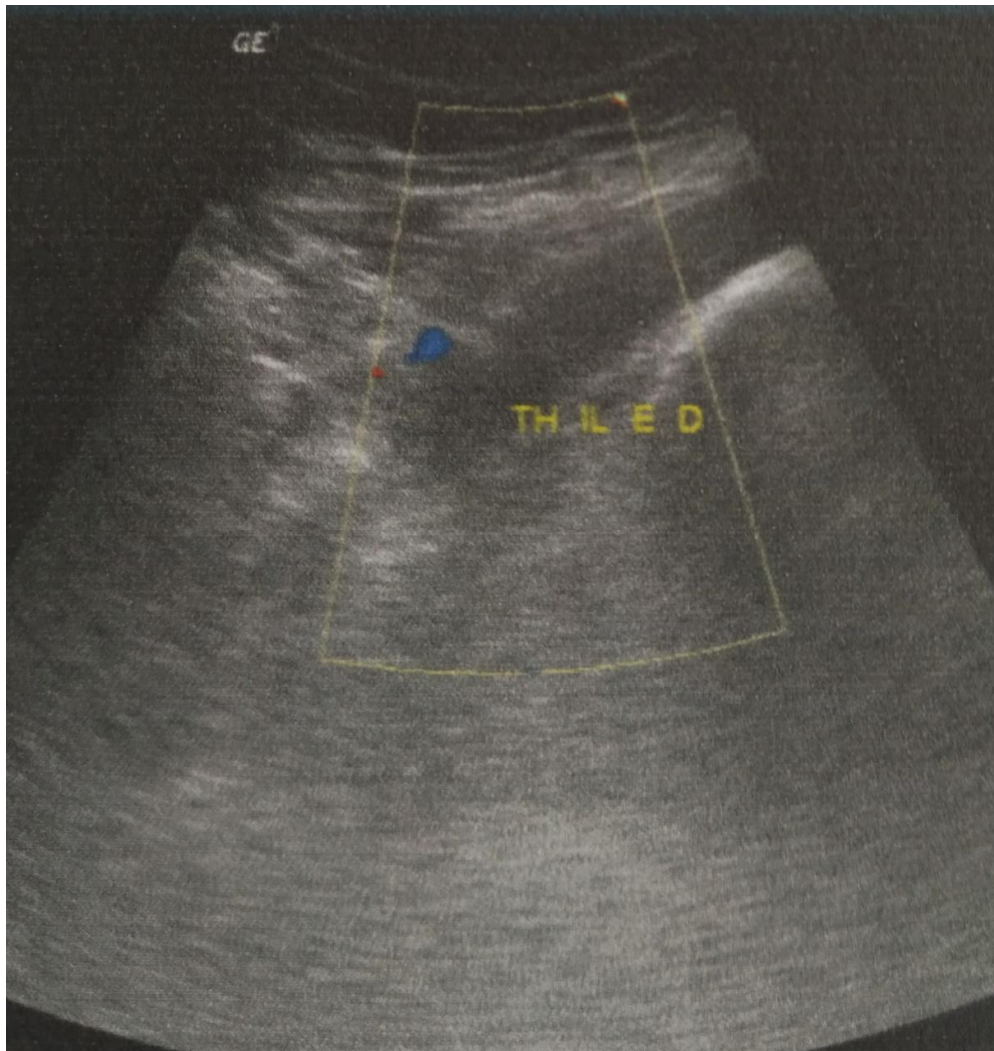


Fig. 1b. Doppler ultrasound showing thrombosis of the right external iliac vein

The patient had no risk factor for thrombophlebitis, the constitutional thrombophilia assessment revealed no abnormality (protein C and S, resistance to activated protein C, antithrombin III and homocysteinemia). He had no aphthosis and the pathergy test was negative. There were no neoplastic call signs (PSA, chest X-ray and abdominopelvic and vesico-prostatic sonography were normal). Long-term oral anticoagulation and elastic compression were prescribed. The patient was lost to follow-up before the end of the explorations and remained on anti-vitamin K (AVK) without controls.

He consulted after 22 months, for bilateral leg ulcers (Fig. 2), a recurrence of thrombosis

in the right lower limb was diagnosed. Considering the extent of the thrombosis, its initial bilaterality, as well as the recurrence under AVK, a CT angiography was carried out and showed a congenital agenesis of the IVC under hepatic and suprarenal, relayed by a very dilated and tortuous azygos system (Figs. 3, 4 and 5). The etiological balance being negative, the AIVC was retained as the etiology for this DVT. Lifetime anticoagulation and elastic stocking were prescribed. The indication for surgical treatment of AIVC is asked. The patient is currently suffering aftermath type of post-thrombotic syndrome.



Fig. 2. Leg ulcers

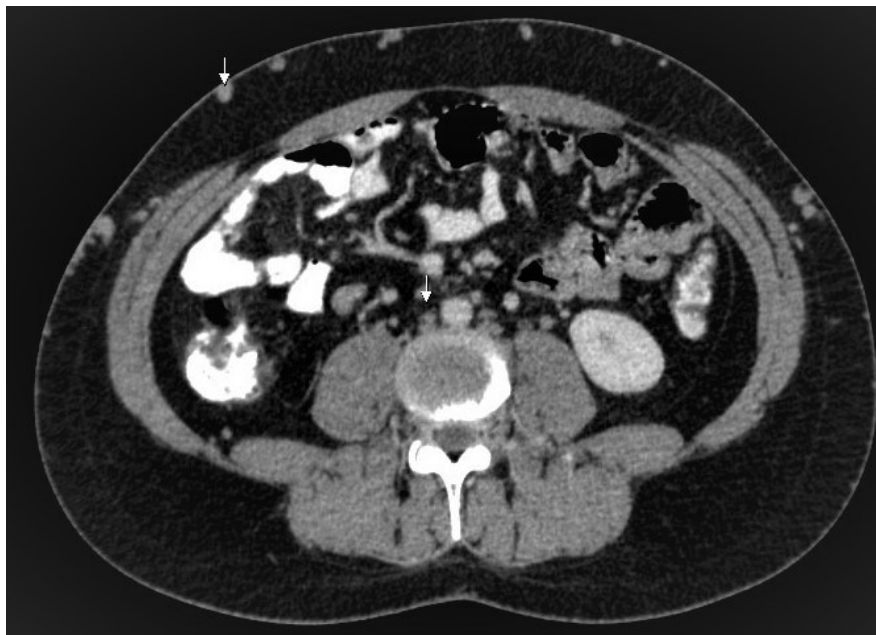


Fig. 3. Axial section showing the agenesis of the IVC with development of a subcutaneous collateral venous circulation of the abdominal wall

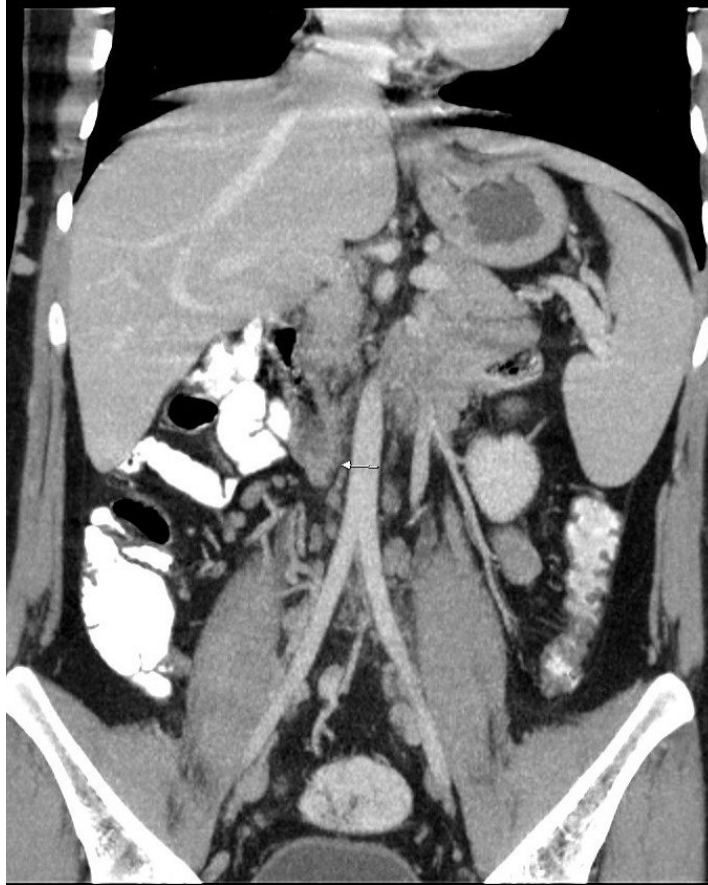


Fig. 4. Coronal section showing the absence of visualization of the IVC

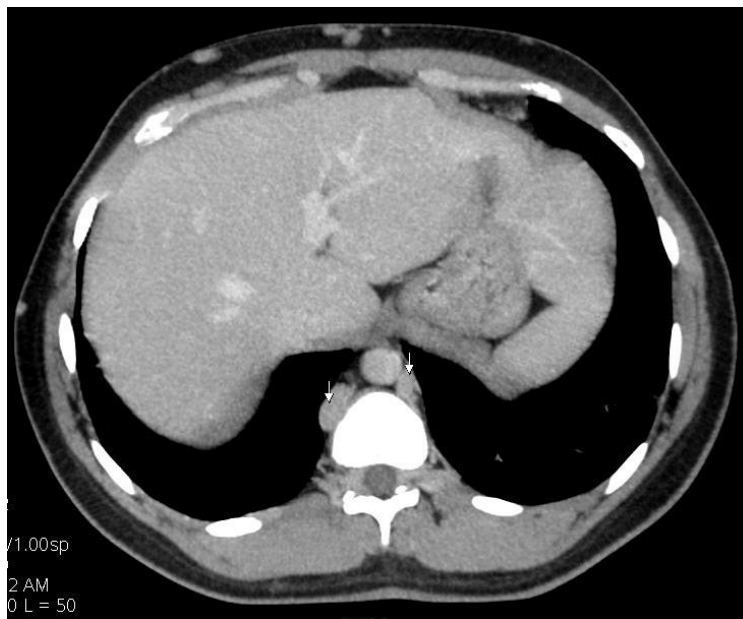


Fig. 5. Axial section showing the dilation of the azygos systems

3. DISCUSSION

The majority of IVC abnormalities, including AIVC, have no clinical significance due to the extent of the collateral venous network of the abdomen and lower limbs. In symptomatic cases, venous thromboses of the lower limbs represent the most frequent clinical case [5]. They mainly affect the proximal network and are often bilateral in around 50% of cases [6]. Some studies estimate that 5% of patients under the age of 30 who have DVT of the lower limbs have a congenital abnormality of the IVC [2].

In 10% of cases, other atypical clinical presentations can reveal these agenesis [5]. In fact, when the thromboses undertake the iliac or para-lumbar circulations, these vascular abnormalities can appear in the form of abdominal or lumbar pain [5,7–11]. The initial clinical presentation, of lumbar pain in our observation joins the description below.

Furthermore, AIVC are very rarely complicated by pulmonary embolism, because this implies that the blood clot must be propelled and not trapped through the small azygos network [12–14]. A case of haemoptysis by spontaneous rupture of a bronchial vein [15] and one case of haematuria were also reported [16].

AIVC has been shown to cause venous stasis and endothelial damage. Therefore, any additional risk factor, which causes hypercoagulability, could lead to venous thrombosis [17]. Venous stasis is due to insufficient venous drainage or inability to support an increase in blood flow, due to the small collateral venous networks of the lower limbs, particularly the azygos and semi azygos networks [18]. This hypothesis could explain that 20% of patients with AVCI, declared intense physical activity, preceding DVT [2,19].

The correlation between IVC anomalies and thrombophilia has not yet been established. [5] In addition, the classic risk factors for venous thrombosis (combined hormonal contraceptives, confinement to bed, travel, etc.), which are less frequent during these vascular anomalies, lead to the conclusion that AIVC could be a sufficient cause for the development of thromboses [4,19].

Venous ultrasound is the examination of choice for the diagnosis of venous thrombosis, but its sensitivity for the exploration of retroperitoneal veins is low. A CT angiography appears useful in

the search for these vascular anomalies, which are often overlooked. In our observation, AIVC was unrecognized during the first episode of DVT.

Long-term anticoagulation and elastic compression are required, even if there is no consensus [6]. Other therapeutic options were discussed; thrombolysis was described as a possible option reserved for patients progressing unfavourably despite well-managed anticoagulation [20]. The surgical approach, whether or not associated with a thrombectomy, has also been described in the literature, but the re-intervention rate for thrombosis bypass surgery remains high, reaching 50% [21].

The long-term prognosis is unknown. It is difficult to estimate the thrombotic risk represented by AIVC, due to its underestimation and the limited number of cases reported. In the published AIVC case series, there was no DVT recurrence after 24 months of oral anticoagulation. Nevertheless, long-term monitoring seems necessary. [22] In our case, a recurrence of DVT was noted, despite curative anticoagulation.

4. CONCLUSION

AIVC is a rare situation of venous thrombosis, which should be suspected when bilateral, extensive and recurrent DVT of the lower limbs occurs in men under 30 years of age. To help the diagnosis strategy, multiplication of radiological explorations seems necessary. More large studies are required in order to better understand the therapeutic and evolutionary modalities of these vascular anomalies.

CONSENT

As per international standard or university standard, patient's written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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