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Case Report

Deep Venous Thrombosis Revealing Agenesis of Inferior Vena Cava

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Abstract

We report the case of a previously well 35-year-old female who presented to the Emergency Department with bilateral lower limb swelling and low back pain. An ultrasound demonstrated bilateral iliofemoral deep vein thrombosis (DVT) and computed tomography demonstrated inferior vena cava agenesis.

Agenesis of the IVC has an incidence of < 1% in the general population. It is a rare congenital malformation that predisposes to DVT, by favouring venous stasis. This diagnosis should be considered in young patients with spontaneous and bilateral DVT.

Keywords: *Agenesis; Inferior Vena Cava; Deep Vein Thrombosis*

Introduction

Agenesis of the inferior vena cava (IVC) is a rare vascular malformation. It has an incidence of < 1% in the general population [1]. IVC agenesis remain asymptomatic as long as venous flow from the lower part of the body is compensated by collateral flow. The diagnosis may be established after an episode of deep venous thrombosis or incidentally during surgery, angiography, computed tomography scans.

We describe the case of female patient with congenital absence of IVC presented with bilateral acute deep venous thrombosis.

Case Report

A 35-year-old female presented with low back pain radiating to both thighs and bilateral lower limb swelling. The pain increased in severity over the preceding 24-hours and resulted in the patient having difficulty mobilising. There were no precipitating factors, and no comorbidities or significant family history. Her only relevant medical history was some lower extremity varicose veins that were treated conservatively. On examination, she was haemodynamically stable but had a temperature of 38oC. Cardio- respiratory examination was unremarkable and her abdomen was soft with mild lower abdominal tenderness.

Haematological analysis confirmed a mild normocytic anaemia with haemoglobin 10 g/dl. The C-reactive protein was elevated at 150 mg/L. All other haematological, coagulation, and biochemical analyses were normal. Plain X-rays of the chest, abdomen and lumbar spine were normal. Doppler ultrasound confirmed bilateral iliofemoral deep vein thrombosis. Abdominal and pelvic CT scan was performed and showed the absence of the entire posthepatic IVC. An enlarged collateral network of lumbar veins, abdominal wall veins, umbilical vein and veins in the splenic region was visible. The network connected to the azygos and hemi-azygos systems. No abnormalities such as polysplenia were detected. The diagnosis of congenital absence of the IVC was made.

The patient was treated conservatively with subcutaneous low molecular weight heparin followed by life- long vitamin K antagonist (acenocoumarol) therapy.

The lower limb swelling resolved weeks later, aided by compression hosiery. The patient remains well 12- months later.

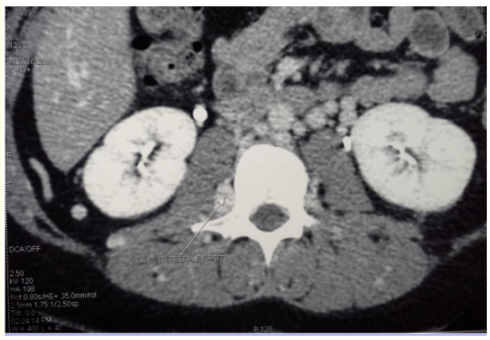

Figure 1: *Computed tomography axial scan of the abdomen showing a large lumbar vein and collateral vessel.*

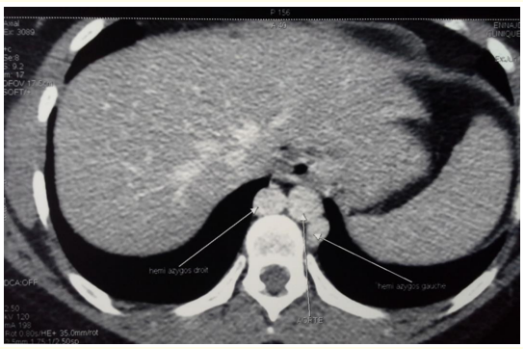

Figure 2: *Abdominal CT scan demonstrating absence of the IVC and showing large azygos and hemiazygos veins.*

Discussion

A review of the embryogenesis of the IVC is essential in understanding developmental anomalies. IVC developmental abnormalities occur at 6-10 weeks of gestation when the infrahepatic IVC develops as a composite structure from three pairs of embryonic veins: the postcardinal, subcardinal, and supracardinal veins. Persistence or regression of these embryonic veins can lead to numerous rare congenital anomalies such as IVC hypoplasia, left IVC, double IVC, and agenesis of infrarenal IVC [2,3]. Absence of the entire posthepatic IVC implies that all three-paired venous systems failed to develop properly. But some authors suggest that it is the result of an intrauterine or perinatal thrombosis [4].

In patients with agenesis of the infrarenal IVC collateral circulation development is common, usually through the four large ways: the gonadal venous system, which drains to the suprarenal cava; the paravertebral venous plexus, which drains to the superior vena cava through the azygous-hemiazygous system; the hemorrhoidal plexus, draining to the portal vein; and the superficial pathway, which drains

to the subclavian veins and superior cava through superficial abdominal veins [3,5,6]. The azygous veins dilate together with lumbar veins and compensate the venous capacity of the lower extremities.

IVC agenesis remain asymptomatic as long as venous flow from the lower part of the body is compensated by collateral flow, which is the case in 25% of the patients [7]. When thrombogenic risk factors accompany the IVC anomaly, DVT often affects the iliofemoral veins on both sides. Patients may present with symptoms of back and abdominal pain, bilateral lower limb swelling.

Duplex ultrasound confirms deep iliofemoral vein thrombosis, usually bilateral. However, in the absence of orientation, venous Doppler ultrasonography does not diagnose vena cava agenesis, but finds abnormalities that encourage additional morphological examinations. The CT scan does not always allow the detection of vena cava agenesis and may leave a doubt. The Magnetic resonance imaging confirms the diagnosis.

Despite the incidence of proximal venous thrombosis, the probability of PE is very low because the migration of thrombi is prevented by the extensive network of compensatory collateral circulation.

The search for thrombophilia is most often negative for the antithrombin III, protein C, protein S and antiphospholipid antibody assays. The search for a factor V Leiden mutation is found in 15% of cases, an abnormality of the transition of the prothrombin gene (G20210A) is found in 13% of cases.

The best treatment strategy of DVT associated with agenesis of inferior vena cava (AIVC) is unclear. An anticoagulation treatment is proposed and prolonged with compression stocking. However, there is no evidence in the literature to define the exact duration of anticoagulation.

Other therapeutic approaches include deep venous thrombectomy and prosthetic surgical replacement of the IVC in combination with temporary arteriovenous fistula [8], but their use is not common.

Conclusion

Inferior vena cava agenesis is a rare situation that must be suspected in the context of bilateral deep venous thrombosis. Morphological investigations of the inferior vena cava allow the diagnosis. Conservative therapy with anticoagulation and compression stockings is sufficient in many cases.

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