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CASE REPORT

Inferior Vena Cava Agenesis: An Unusual Cause of Deep Vein Thrombosis and Pulmonary Embolism in Young Adult Patients

J. Ramos Aranda [a,*], C. Ramírez Cerda [b], S. Cohen Mussali [b], J. Valdés Flores [b]

[a] General Surgery Department Resident, American British Cowdray Hospital, Mexico City, Mexico
[b] Vascular Surgery Department, American British Cowdray Hospital, Mexico City, Mexico

Introduction: Inferior vena cava agenesis (IVCA) is one of the many anomalies of this vessel. It is one of the most uncommon anomalies, with an estimated prevalence of 0.0005–1% in the general population. Around 5% of the patients younger than 30 years with a diagnosis of deep vein thrombosis (DVT) have a total or segmental IVCA.
Report: Here two unique cases of young and previously healthy male patients are reported: one with bilateral lower extremity DVT, the second with lower extremity DVT and pulmonary embolism. Both patients were found to have segmental agenesis of the inferior vena cava on computed tomography angiography (CTA). Treatment consisted of ultrasound enhanced thrombolysis (EKOS + alteplase) and venous angioplasty. Both patients were discharged with long-term (up to 24 months) oral anticoagulation and compression stockings. Follow up at 3 and 12 months revealed no new thrombotic episode.
Discussion: IVCA can be asymptomatic but the majority of the symptomatic patients present with DVT. IVCA confers a risk factor for DVT. IVCA should be considered and ruled out as a rare but important risk factor and cause of DVT in previously young healthy patients. Once diagnosed, aggressive treatment must be started because of the high risk of post-thrombotic syndrome.

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INTRODUCTION

Inferior vena cava agenesis (IVCA) constitutes one of many inferior vena cava (IVC) congenital anomalies. Around 15–60 different IVC anomalies have been described since 1793, many of which do not have any clinical significance.[1] IVCA can be asymptomatic but the majority of the symptomatic patients present with deep vein thrombosis (DVT). IVCA is a risk factor for DVT.

Two cases are presented of young male patients diagnosed with IVCA after the onset of lower extremity (LE) DVT.

CASE PRESENTATION

Case 1

A 23 year old man was admitted with a 1 week history of bilateral LE swelling, pain, and erythema, gradually progressing to the inability to walk. One month prior to admission because of post-traumatic pain, the patient decided to mobilise as little as possible and remained in bed for a week.

On physical examination, swelling and hyperthermia involving both legs and thighs were found. Acute prominent engorged abdominal collateral veins were seen (Fig. 1).

Venous Doppler ultrasound (VDU) showed bilateral DVT extending to both common iliac veins. A computed tomography angiogram (CTA) showed agenesis of the infrarenal segment of the IVC, DVT of both common iliac, lumbar and gonadal veins (Fig. 2), and patent renal veins draining in to the azygous system on the right and hemiazygous on the left.

Low molecular weight heparin (LMWH) was started, and ultrasound enhanced thrombolysis (UET) was performed. The EKOS System (BTG International Ltd, West Conshohocken, PA, USA) was used bilaterally via popliteal access, with alteplase administration for 24 hours.

After the venography, a partial response was found so treatment was continued for another 24 hours.

The next venogram showed excellent response at the level of the popliteal and femoral veins bilaterally and an 80% response in both common iliac veins. A stenosis was found in the left common iliac vein; thus, a self-expanding stent was placed across it (Zilver Vena, Cook Medical LLC, Bloomington, IN, USA).

* Corresponding author. Calle Rio de la Plata #17-503, Cuauhtémoc, 06500, Mexico City, Mexico.
E-mail address: j.ramos.aranda@gmail.com (J. Ramos Aranda).

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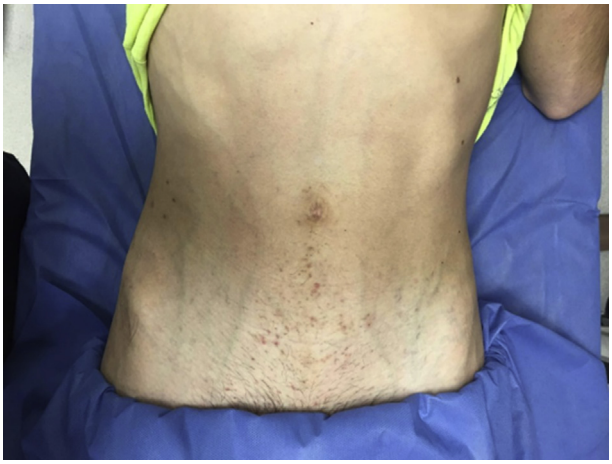

Figure 1. Acute prominent engorged abdominal collateral veins.

He was discharged with rivaroxaban and compression stockings. At 12 month follow up the patient was still anticoagulated and asymptomatic.

Case 2

A 30 year old male was diagnosed with a left LE DVT and treated with dabigatran 2 weeks prior to admission to the hospital because of shortness of breath. Bilateral pulmonary embolism (PE) was confirmed on CTA.

Physical examination revealed swelling and hyperthermia on the left LE. VDU showed left LE DVT in the femoropopliteal segment. On questioning he admitted to the use of anabolic steroids and extreme physical activity over the previous two months. He had no family history of thrombosis. LMWH at therapeutic dose was started.

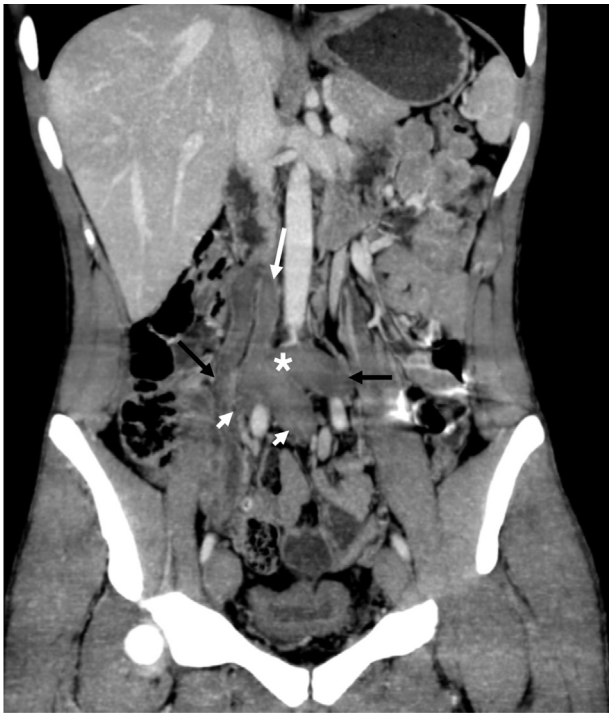

Figure 2. Computed tomography angiography, coronal reconstruction, shows a confluent inferior vena cava stump (asterisk), dilated gonadal vein (right black arrow), dilated lumbar vein (left black arrow) and iliac veins (white arrowheads), and thrombosed right gonadal vein (long white arrow).

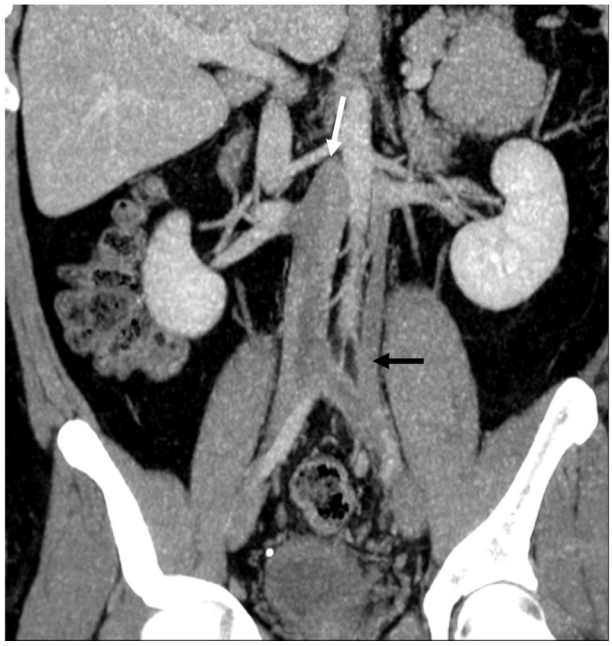

Figure 3. Computed tomography angiography, coronal reconstruction shows discontinuity of the inferior vena cava at suprarenal level (white arrow), duplicated inferior vena cava draining in to the left renal vein (black arrow).

CTA of the abdomen and pelvis showed extensive thrombosis of the femoropopliteal segment, suprarenal IVCA and duplicated IVC (Fig. 3) with azygous continuation (Fig. 4) of a left retro-aortic renal vein.

A diagnostic venogram showed DVT of the left common iliac vein, both renal veins, and the infrarenal segment of

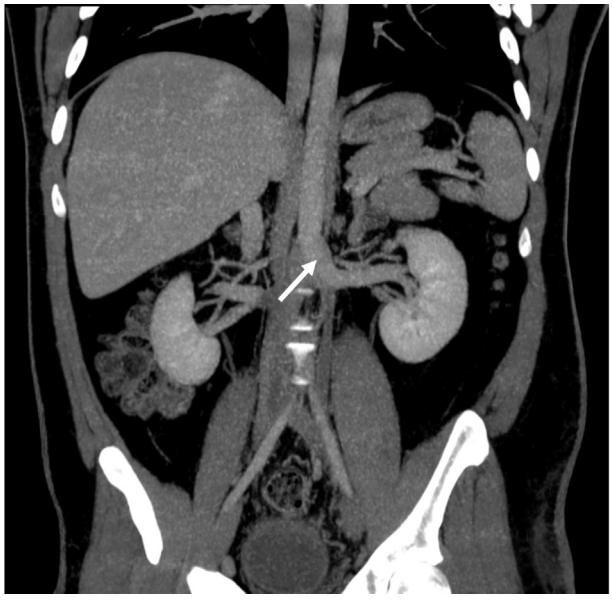

Figure 4. Computed tomography angiography, coronal reconstruction shows the drainage of the left renal vein (white arrowhead) into the azygous system.