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CASE REPORT

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

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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).

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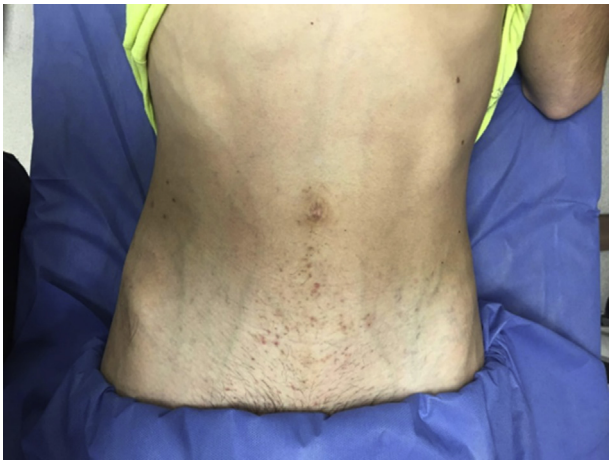

Figure 1. Acute prominent engorged abdominal collateral veins.

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.

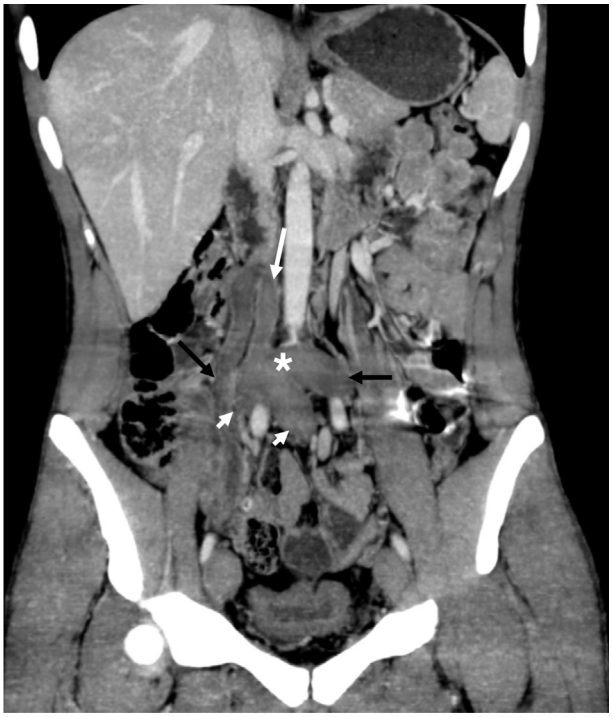

Figure 2. Computed tomography angiography, coronal reconstruction, shows a confluent inferior vena cava stump (asterisk), dilated gonadal vein (right black arrow), dilated lumbar vein (left black arrow) and iliac veins (white arrowheads), and thrombosed right gonadal vein (long white arrow).

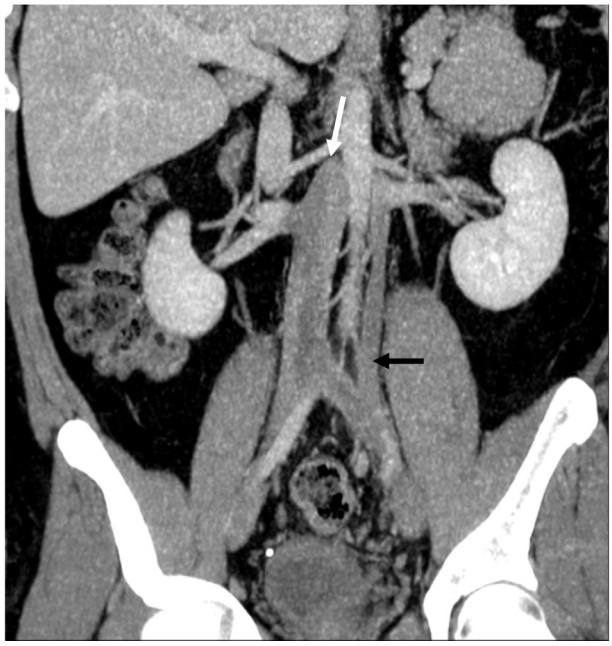

Figure 3. Computed tomography angiography, coronal reconstruction shows discontinuity of the inferior vena cava at suprarenal level (white arrow), duplicated inferior vena cava draining in to the left renal vein (black arrow).

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

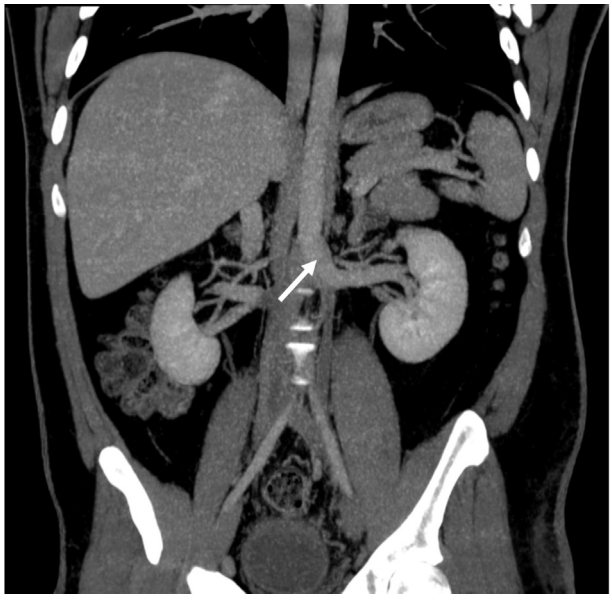

Figure 4. Computed tomography angiography, coronal reconstruction shows the drainage of the left renal vein (white arrowhead) into the azygous system.

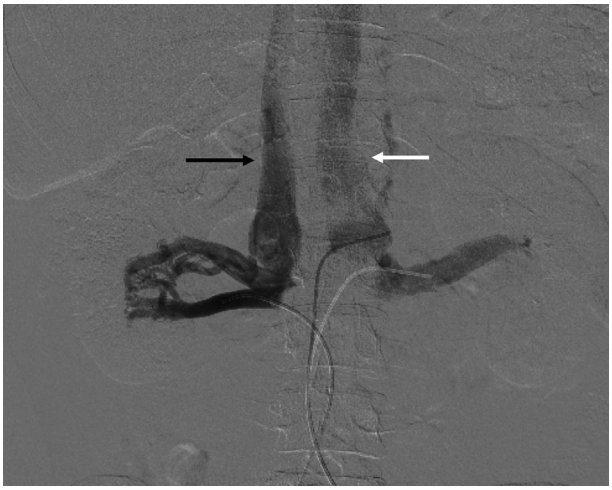

Figure 5. Renal venography shows both renal veins draining directly into the thrombosed renal inferior vena cava, the right renal vein (black arrow) drains through a collateral into the right ventricle and the left renal vein drains into the azygous system (white arrow), also through a collateral.

the IVC. Interruption of the suprarenal segment of the IVC was seen (Figs. 5 and 6) with drainage of the right renal vein directly into the right ventricle (Fig. 6).

UET was performed using the EKOS System (BTG International Ltd) with the administration of alteplase for 24 hours.

A 24 hour venogram showed residual thrombosis or stenosis of the left iliac vein, balloon angioplasty (Atlas, Bard PV Inc., Tempe, AZ, USA) was performed and a self-expanding stent (Zilver Vena, Cook Medical LLC) was placed across it, achieving 100% resolution.

The patient was discharged on oral anticoagulation rivaroxaban and compression stockings. At 6 month follow up, the patient is asymptomatic with no new DVT episode.

DISCUSSION

The embryological development of the IVC is a complex process consisting of multiple steps, all of which occur in a specific order. It evolves from three primitive veins: supracardinal veins, the posterior cardinal veins, and the subcardinal veins, which will develop, predominate temporarily, regress, then finally anastomose forming the IVC. Any event that alters these steps will cause a congenital anomaly. The incidence of such anomalies in the global population has been estimated to be 0.05—8.7%.[2]

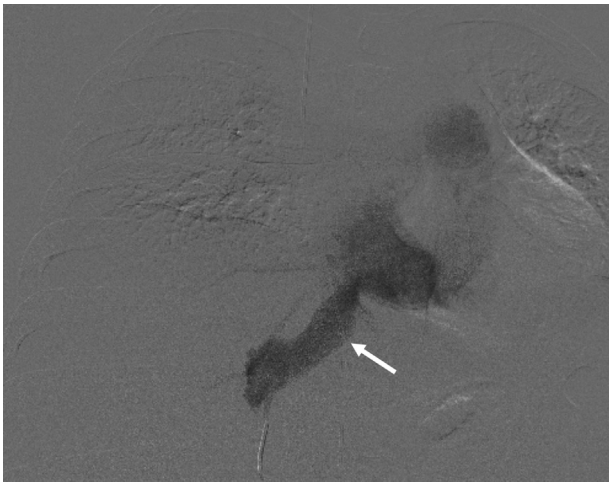

Figure 6. Right renal vein venography showing direct drainage into the right ventricle through a collateral (white arrow).

Agenesis of the IVC is one of the most uncommon anomalies, with an estimated prevalence of 0.0005%—1%.[2] This anomaly may be found as two different variants, complete agenesis or segmental agenesis. Agenesis of the suprarenal segment is the most common, while agenesis of the hepatic and infrarenal segment constitutes only 6% of all cases.[3] It is thought that the majority of cases remain asymptomatic because of the extent of the collateral venous network in the abdomen and LE; however, there will always be venous stasis compared with the general population despite the drainage.[2]

Patients that present with agenesis of the infrarenal segment need drainage through the azygous, hemiazygous, lumbar, para-vertebral, and abdominal wall veins, as seen in both of the presented cases.[4,5] The common iliac veins can drain directly into the lumbar venous system.[4] Inherently, IVCA leads to venous stasis and endothelial damage; hence, any risk factor that causes hypercoagulability can lead to DVT.[6] These risk factors were the secondary precipitant factors in both cases (anabolic steroids, major physical exertion, and prolonged immobilisation).

The incidence of DVT in patients over 40 years of age is 1 in 1,000; meanwhile in patients between 20 and 40 years, the incidence decreases to 1 in 10,000. DVT has a multifactorial aetiology that is significantly associated with genetic and environmental risk factors that affect the coagulation state.[7] Approximately 5% of patients below 30 years of age with DVT have complete or segmental IVCA.[8]

A homogeneous profile has been detected in patients with IVCA:[4,5]

- young patients, $<$ 40 years of age
- male gender
- unilateral or bilateral DVT.

IVCA commonly presents with abdominal and lumbar pain, unilateral or bilateral LE swelling and paresthesia, chronic venous insufficiency, and abdominal wall venous dilation. PE has been reported through the collateral circulation,[6,9] as seen in patient two, where both renal veins drained into the pulmonary circulation through collaterals.

CTA is recommended for all patients with idiopathic DVT above the inguinal ligament, in combination with magnetic resonance imaging or venography, the latter being the more sensitive.[7] Thrombophilia screening is also recommended to rule out a hypercoagulability state.[8]

A consensus has not yet been established for the treatment of patients with DVT and IVCA. Most cases have been effectively managed with anticoagulation and compression stockings, and, in some cases, thrombolysis.[8] Catheter directed thrombolysis may be considered; however, it is

known to be challenging because of the anatomy.[9] There are five prior reports of DVT/IVCA treated by catheter directed thrombolysis; of these, only three were treated successfully, with a total dose of 20—60 mg of alteplase.[8] In both of the presented cases, thrombolysis was achieved with UET with excellent results and a 100% technical success. There are no reports in the literature regarding the use UET and angioplasty alone in this scenario (extensive DVT + PE + IVCA). The use of UET allows for a shorter length of stay, greater technical success, and decreased morbidity, with the use of a much smaller dose of thrombolytic (10—20 mg alteplase).

Owing to the young age of the patients and extensive DVT/IVCA, treatment with UET was chosen to decrease the considerable risk of post-thrombotic syndrome.

In the majority of cases, anticoagulation has been achieved with LMWH for the first few days, with bridging to vitamin K antagonists or factor Xa inhibitors. Prolonged treatment is recommended. There is evidence of recurrent DVT in patients that stop oral anticoagulation within the first 25 months,[3,8] (probably because of permanent mechanical venous stasis, altered venous drainage through collaterals, and previous thrombosis). Patients were discharged on long-term oral anticoagulation and compression stockings. The 3 and 12 month follow up showed no new thrombotic episodes.

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CONFLICTS OF INTEREST

None.

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