Case Series

Congenital Absence of Suprarenal Inferior Vena Cava

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Abstract

We describe here 2 cases of congenital agenesis of the suprarenal inferior vena cava (IVC), which is a rare and often complex abnormality, in the setting of iliocaval deep venous thrombosis. This disorder often presents via a wide variety of symptoms and treatment modalities range from pharmacological thrombolysis to open venous reconstruction.

We will review the embryology of the IVC and outline the presentation of 2 cases of its congenital absence. These cases highlight the significance of considering this abnormality in the setting of deep venous thrombosis involving the iliocaval system to avoid diagnostic pitfalls. We recommend the use of appropriate imaging modalities to delineate this pathology and discuss the various therapeutic approaches to this problem.

Key Words: Inferior vena cava, Congenital, Absence

Introduction

Congenital agenesis of the inferior vena cava (IVC) is a rare abnormality. Its embryogenesis is a complex process involving the development of multiple anastomoses between three primitive paired embryonic veins1. This defect can lead to considerable variability in the configuration of venous return from the abdomen and lower limbs. Some of these anomalies have significant clinical implications. We will discuss two cases of agenesis of the suprarenal IVC.

Case 1

A 60 year old man presented on 14/5/2013 with bilateral lower limb swelling with associated back and abdominal pain over 4 days. His surgical history featured cervical fusion following an axial loading injury as a teenager. He was a non smoker and had no risk factors of venous thromboembolism. He was born at 34 weeks of gestation.

This builder experienced sharp lower back pain of sudden onset 4 days prior to his presentation, while at the top of a ladder. He sought attention from his naturopath and physiotherapist without much relief. He developed lower abdominal and right sided knee pain over the following 2 days. He presented to the emergency department with both abdominal and lower back pain, as well as altered sensation down both legs. He developed bilateral lower limb swelling over the 6 hours that he was in the emergency department.

On examination, he was an elderly man of slim built, lying uncomfortably in bed with grossly swollen and tender thighs and calves. A purple coloration was noted throughout his lower limbs. He had no abdominal tenderness and no varicosities were found.

Computed tomography of his abdomen and pelvis revealed absence of the superior segment of his inferior vena cava with dilated collateral vessels contributing to azygos and hemiazygous continuations(Fig1) . Doppler ultrasound of his lower limbs showed occlusive thrombus within his distal IVC, right common iliac vein and external iliac vein, throughout the common femoral vein to the popliteal vein. On the left, there was a suggestion of partially occlusive thrombus within the left iliac veins and no flow through the left common femoral vein.

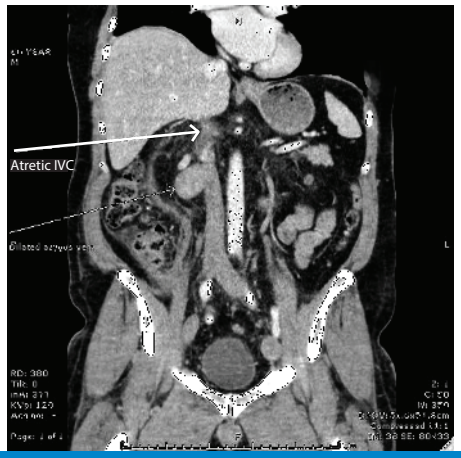

Fig 1 - CT abdomen and Pelvis showing absent IVC

His thrombophilia screen and tumour markers were negative.

He was managed with bed rest, leg elevation and a heparin infusion acutely. He underwent catheter directed thrombolysis on the next day. Unfortunately he developed a small retroperitoneal psoas haematoma after 24 hours and his thrombolysis was stopped prematurely.

He was initiated on life long anticoagulant therapy with warfarin. He was discharged from the hospital when stable and was reviewed 7 months later in the outpatient clinic. He had been symptom free since discharge and had returned to work.

Case 2

This 19 year old female German student presented on 24/2/2009 with a 4-day history of lower back and abdominal pain, which radiated down her right thigh and calf. She had travelled by bus from Sydney to Melbourne a few days prior to her presentation. Her past medical history included asthma. She was a non smoker and was on oral contraceptive pill.

She was initially investigated with ultrasound and was found to have extensive ilio-femoral deep venous thrombosis. Specifically, a combination of venography and CT angiography revealed thrombosis in the right common femoral, external iliac, internal iliac and common iliac veins (Fig2). The left internal iliac and left common iliac veins were also occluded.

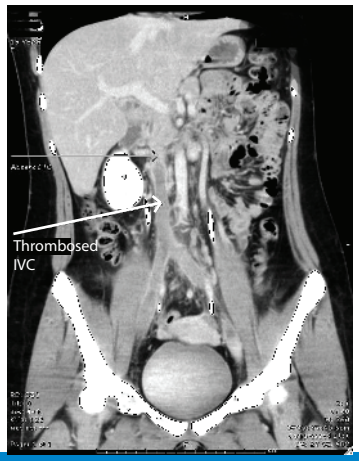

Fig 2 - CT abdomen and Pelvis showing thrombosed IVC

The inferior vena cava was occluded to the level of the right renal vein and the right renal vein drained through the azygos system. The left renal vein did not cross the midline and drained through well established hemiazygos veins. The hepatic veins drained into the suprahepatic portion of the inferior vena cava. Markedly enlarged azygos and hemiazygos veins were demonstrated. The IVC was not visible above the level of the renal veins and it was suggested that she had agenesis of the superior segment of her IVC.

Her thrombophilia screen, angiotensin converting enzyme level, B2 microglobulin, serum electrophoreis and tumour markers were all essentially normal. There was no evidence of paroxysmal nocturnal haemoglobinuriaon flow cytometry.

She was started on a heparin infusion initially and was treated with intravenous urokinase for 5 days. Most of the thrombus was dissolved except for a small residual amount in the right common iliac vein which was presumed to be long standing thrombus.

Her inpatient stay was complicated with a further bout of thrombosis in her right external and common femoral veins over the next 3 days and she had a further 5 days of thrombolysis. This returned flow from her lower limbs to her heart via an extensive collateralised azygos system.

No thrombophilic cause was found and she was anticoagulated with warfarin at the end of her thrombolysis. She returned to Germany with a medical escort and was anticoagulated with warfarin for 1 year. She did not have any further episodes of venous thrombosis.

Discussion

Anomalies of IVC formation are rare, with an incidence in the general population of 0.005 to 1 %, but can have serious complications[2]. An awareness of such venous malformations can avoid diagnostic pitfalls.

The IVC is formed during weeks 6 to 8 of gestation and this involves a series of consecutive anastomoses among three pairs of veins from the cardinal system (supracardinal, posterior cardinal, and subcardinal veins). It is theorized that agenesis of the IVC results from failure of the right subcardinal vein to connect to the liver and shunting of blood occurs instead into the right supracardinal vein. Caudad blood arrives at the heart via the azygos vein and SVC while the hepatic vein reaches the right atrium at the site of the IVC.

A single case of regression of the IVC post-natally has been documented after formation of thrombus in the iliocaval system at 48 hours of life[4]. Although our first case was born at 34 weeks of gestation, there was no indication that he had any venous or thrombotic abnormalities at birth.

The majority of individuals with IVC agenesis present without symptoms. The most common presentation is deep venous thrombosis in the iliofemoralregion, mainly among young males[5]. It is noted that the chance of pulmonary embolism in this category of proximal DVT is low as thrombi have to navigate through a dense network of collateral vessels[6,7].

Since its embryology is so complex, there are multiple alternate pathways for venous drainage of the lower half of the body as well as several opportunities for congenital anomalies (situsinversus, asplenia, or polysplenism)[3]. Typically, collaterals establish themselves through the gonadal or azygous-hemizygous systems; or paravertebral or haemorrhoidal plexi[6]. Case reports detailing symptoms from those avenues are few – lower limb neurological symptoms[8,9,13], abdominal complaints[10,11] and shock from rupture of a venous aneurysm10. Both our patients presented with abdominal and lower limb pain.

Neoplastic disease as well as other causes for DVT should be sought with imaging modalities of the abdo

men and pelvis. In both our patients, CT scanning were used. Additionally, they underwent venography. Hereditary thrombophilia was excluded in both our patients despite there being only 2 reported associations with congenital absence of the IVC[11,12].

Both our patients initially received thrombolytic therapy and were anticoagulated with heparin before being started on lifelong warfarin. While Dougherty et al. describe venous bypass surgery, this was not considered in either patient given the resolution of their symptoms and lack of functional impairment[3].

These 2 cases highlight the importance of considering congenital vena cava abnormalities in the setting of iliocaval thrombosis, especially in presentations with associated abdominal or neurological symptoms. Suitable imaging modalities such as computed tomography or contrast venography should be employed and long term anticoagulation therapy is considered appropriate management[14].

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If it's MRSA, Wait! Don't Rush.

A new study conducted in Cedars-Sinai Medical Center, California, suggests that treating MRSAinfected-patients with beta lactam antibiotics is not a bright idea as it makes the patient's condition worse. MRSA is a relatively common organism responsible for life threatening infections including hospital acquired ones. It is found in the skin of 1% of healthy individuals. In US, it is the commonest cause of skin and soft tissue infections. Since MRSA culture report takes a couple of days, the patient is likely to be treated in the interim with most commonly used first line antibiotic, beta lactam group. Beta lactam activates mecA gene (the very gene that is responsible antibiotic resistance) and damages MRSA's cell wall releasing a powerfully immunogenic substance. The latter triggers an adverse immune response that makes the infection worse. The moral is, if you have a least bit of suspicion of MRSA infection, wait for culture report. Don't rush with beta lactams!(Cell Host & Microbe. Volume 18, Issue 5, p604–612, 11 November 2015)

- Dr. K. Ramesh Rao