ABSENT SUPRARENAL INFERIOR VENA CAVA REVEALED ON CT SCAN IN A PATIENT WITH ACUTE APPENDICITIS

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Abstract

Inferior vena cava agenesis (IVCA) is an uncommon congenital anomaly which was first described in the seventeenth century. The detection of such an anomaly is often incidental and patients are asymptomatic until the third to fourth decade of life. Due to the scarcity of these cases, there is no consensus on the subsequent treatment and surveillance of incidental asymptomatic IVC anomalies. Herein, we report on a young male with incidental findings of suprarenal IVC agenesis identified on contrast enhanced computed tomography (CT) scan of the abdomen in the course of treatment for acute appendicitis.

Keywords: *Inferior Vena Cava, Agenesis, Deep-Vein Thrombosis, Appendicitis*

Introduction

Inferior vena cava agenesis (IVCA) is an uncommon congenital anomaly with an incidence of 0.0005 % in the general population (1). This malformation is usually discovered on contrast enhanced computed tomography (CT) scan of the abdomen or identified intraoperatively during abdominal surgery (2). Frequent association of deep vein thrombosis (DVT) during the third to fourth decade of life is observed in the majority of patients (3). Herein, we report a case of the incidental finding of suprarenal IVC agenesis during the course of treatment for acute appendicitis.

Case Presentation

A 21-year-old male presented to the emergency department due to right lower abdominal pain for one day which was colicky in nature and radiated to the umbilicus. There was a presence of fever and anorexia without any prior symptoms suggestive of urinary tract infection. Subsequent history revealed that the patient was an active and fit man, with a non-significant birth history and without any prior hospitalizations. Abdominal examination revealed a tender right iliac fossa with rebound tenderness. Parameters of the full blood count were within normal limits with the exception of a raised white cell count of 13.9 x 1000/mm^3. A contrast enhanced CT of the abdomen was performed as the patient had experienced less severe right iliac fossa colicky pain six months prior, and the results showed features consistent with a diagnosis of acute appendicitis. An incidental finding, apart from a dilated appendix with surrounding fat stranding, was an absent suprarenal Inferior Vena Cava (IVC) with collateralization of the azygos and lumbar venous system (Figure 1-3). The left and right renal venous drainage consisted of multiple tortuous dilated vessels which drained to the lumbar vessels and azygos veins (Figure 2 and 3). There was continuation of the infrarenal IVC below the renal hilum to the bifurcation of the common iliac vein (Figure 4). The venous drainage of the remaining infrarenal IVC was towards the lumbar veins, which were dilated on the CT scan (Figure 3). The patient underwent an emergent laparoscopic appendicectomy with intraoperative findings of suppurative appendicitis that was confirmed on the final histopathological examination. Recovery was uneventful and the patient was discharged on post-operative day 1. In light of the incidental finding of an absent suprarenal IVC, further history revealed that the patient had no prior history of bilateral calf pain to suggest deep vein thrombosis. To the patient's best knowledge, there was no family history of similar findings of an absent IVC. Recovery was uneventful and the patient went back to his normal active lifestyle prior to surgery. The patient is currently under the follow-up of general surgery and declined to be followed-up in a vascular unit for risk of developing deep vein thrombosis.

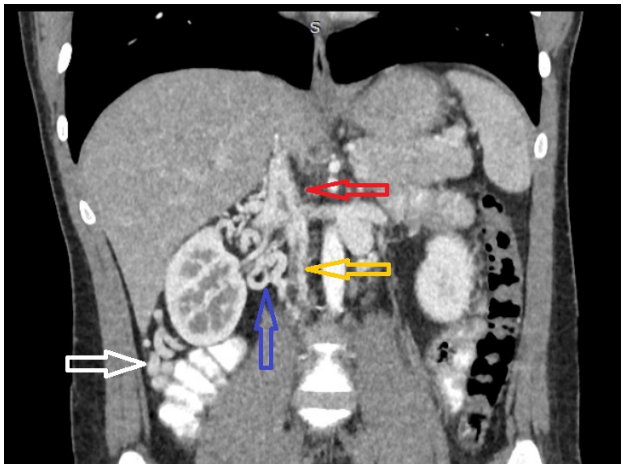

Figure 1: Coronal view of CT scan of IVCA showing incomplete IVC from suprarenal (red arrow), multiple tortuous collateral draining the right kidney (blue arrow), tortuous lumbar veins (yellow arrow) and multiple tortuous retroperitoneal veins (white arrow)

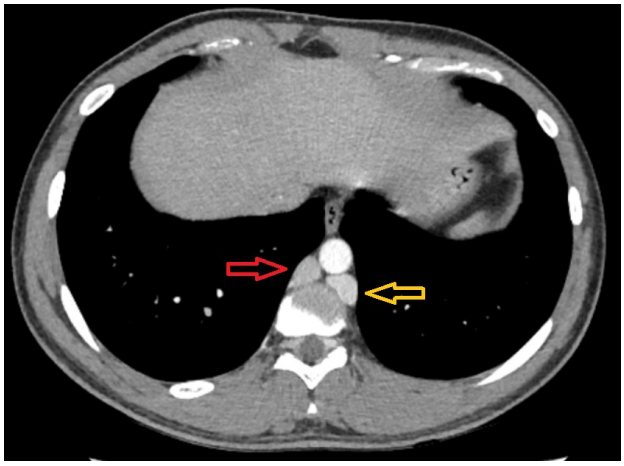

Figure 2: Axial view of contrast enhanced CT scan of IVCA showing dilated right azygos vein (red arrow) and dilated left hemiazygos vein (yellow arrow)

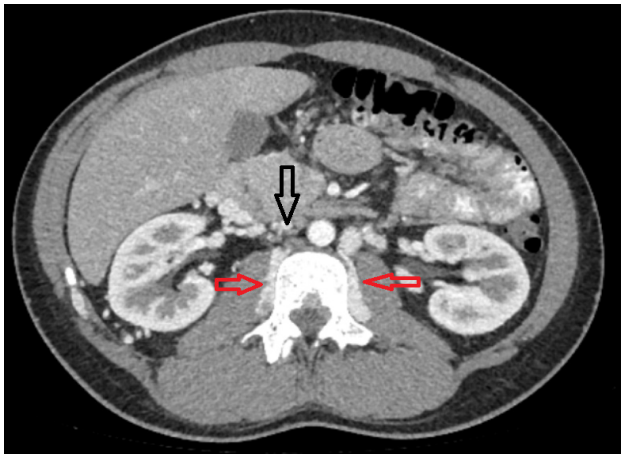

Figure 3: Axial view of CT scan of IVCA showing absent IVC (black arrow), dilated right and left lumbar veins (red arrow)

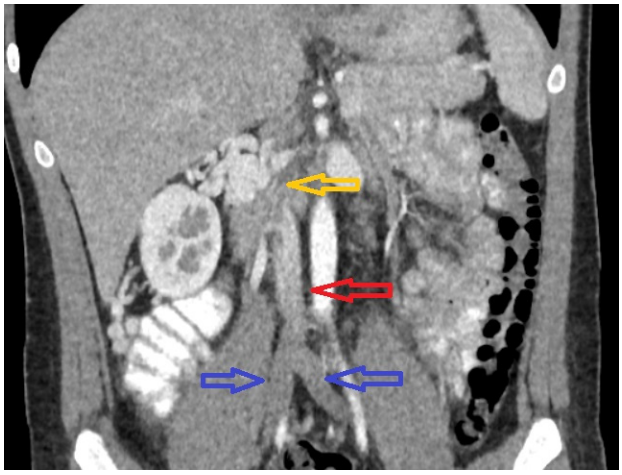

Figure 4: Coronal view of CT scan of IVCA showing continuation of IVC after the renal hilum (yellow arrow), continuation of infrarenal IVC (red arrow) and bifurcation of bilateral iliac vein (blue arrow)

Discussion

Inferior Vena Cava (IVC) anomalies, which are commonly observed in men, was first recognized more than 200 years ago. Abernethy et al described the first case, in 1793, in a patient with a mesocaval shunt and direct continuation of the IVC into the azygos vein (5). To date, reported aberrations involving the IVC consist of five (5) major variations which include: 1. Retroarotic left renal vein; 2. Left infrarenal IVC; 3. Circumaortic left renal vein; 4. Double IVC, Azygos continuation of IVC and; 5. Absence of infrarenal IVC (5). Collectively, these anomalies are termed IVC aplasia, IVC atresia, IVC agenesis, IVC hypoplasia or absent IVC (3, 6). Despite the numerous variations in terms reported, the estimated prevalence of IVC anomalies is only 0.0005 – 1 %, with an incidence of 1 per 10000 in the general population (6). Formation of the IVC in-utero begins from the sixth and eighth gestational week leading to the anastomosis and posterior regression of the subcardinal, supracardinal and postcardinal veins (4). The two proposed etiologies are embryological dysgenesis or intrauterine venous thrombosis, which lead to these anomalies involving the IVC, with the former being the most commonly accepted theory (3). The majority of these patients are asymptomatic and detected on incidental discovery following abdominal surgery or contrast enhanced CT scan. In a minority of patients, the main presenting symptoms are bilateral lower extremity dull aching pain, edema, ulcers and occasional non-specific lower abdominal pain. These symptoms normally occur in the third to fourth decade of life after physical exertion. This is likely due to the inability of the collaterals to cope with increasing blood flow during physical exertion, which leads to venous stasis and deep vein thrombosis (7). Frequent anatomic sites of DVT occurrence are in the infrarenal IVC, external, internal iliac veins and femoral veins bilaterally. To cope with the increasing demands of blood flow, venous drainage is redistributed within the deep venous pathways via the lumbar, intervertebral and into the azygos veins

(6). Lambert et al (3) published a descriptive study of 62 patients with IVC dysgenesis and identified several key points which are pertinent for subsequent management: 1. Risk of DVT was 35.4 % in patients with IVC agenesis; 2. Diagnosis of IVC anomalies was more likely to be detected by contrast enhanced CT scan of the abdomen; and 3. A total of 18. 6% of known thrombophilia patients with IVC agenesis had concomitant DVT. Due to the possible risks, it is advocated that thrombophilia screening be carried out in all patients presenting with IVC anomalies and DVT (7). From the relatively small number of reported cases, subsequent surveillance with imaging and primary prophylaxis options should be mainly on an individual case to case basis. In patients with deep venous thrombosis, therapeutic oral anticoagulants should be commenced. Further individual risk assessment is required for lifelong continuation or discontinuation of oral anticoagulation after the resolution of DVT (3). In asymptomatic patients without DVT, adequate counselling with regards to risk factor prevention (prolonged immobilization, oral contraceptive use) and lifestyle modification (obesity and smoking) in relation to DVT should be given. Usage of thromboembolic deterrent stockings and leg elevation is a logical and safe approach to the prevention of DVT in patients with asymptomatic IVC anomalies (3). The risk of pulmonary embolism is low due to the trapping of possible emboli within the azygos/hemiazygos system before reaching the pulmonary circulation (7). In relation to this case report, ample advice was given with regards to DVT prevention as the patient was young, active and asymptomatic. However, due to personnel reasons, the patient declined a subsequent vascular surgery follow-up. Thrombophilia screening was not performed as this patient was asymptomatic without any family history of thrombophilia.

Conclusion

Inferior vena cava agenesis is an uncommon congenital anomaly which is usually identified in asymptomatic patients. Due to the scarcity of reported cases, treatment of asymptomatic patients remains on a case to case basis with emphasis on the prevention of deep vein thrombosis. Albeit the risk of pulmonary embolism is low, the development of DVT (from IVC anomalies) in a young patient is debilitating due to its accompanying symptoms and foreseen long-term treatment with oral anticoagulants.

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Competing Interests

The authors declare that they have no competing interests.

Informed Consent

Written informed consent was obtained from the patient who participated in this study.

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