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# Rare congenital anomaly of the inferior vena cava

Congenital anomaly of the inferior vena cava (IVC) is a well described phenomenon [1,2]. These anomalies include complete absence, partial absence or duplication of the IVC [3]. Such anomalies of the IVC are seen more frequently in those with other congenital cardiac anomalies (0.6%–2%) [4].

This congenital condition can be discovered incidentally, or due to symptoms of associated congenital heart disease, asplenia, polysplenia, congenital kidney anomalies or deep venous thrombosis [1,2,3].

Figs. 1 and 2 are the CT angiography images of the venous system of the lower extremities and inferior vena cava of a 44 year old Caucasian male who presented with a long history of intermittent edema of both lower extremities. His clinical examination was perfectly normal. CT angiography of the lower extremities and IVC demonstrated patent popliteal, femoral and iliac veins with a completely absent IVC. Massively dilated collateral vessels were visible with large, dilated azygos and hemi-azygos veins. The azygos vein drained into the superior vena cava. In addition the lumbar veins were dilated with multiple collaterals present in the pelvis and abdomen. He tested heterozygous for the factor V Leiden mutation (R506Q).

Although congenital anomaly of the IVC is a well known clinical entity, combined absence of the suprarenal and infrarenal IVC (as demon-

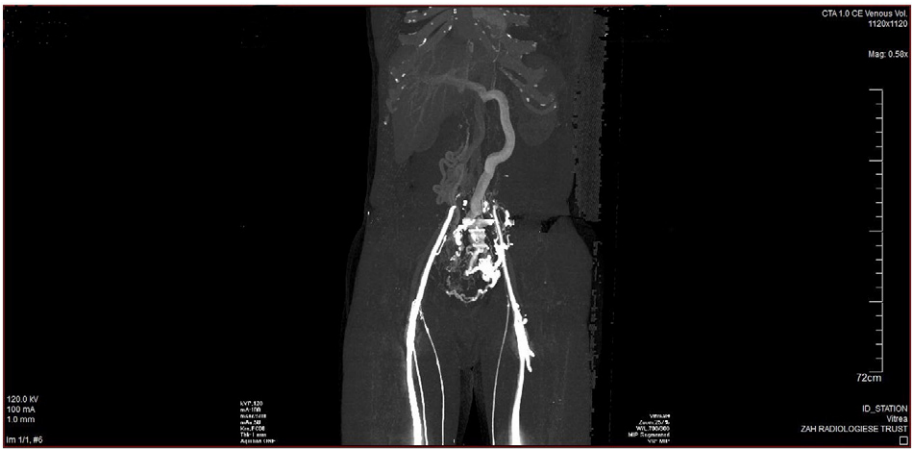


**Fig. 1.** CT angiography image demonstrating absence of the infra- and suprarenal portions of the inferior vena cava with numerous collateral vessels.

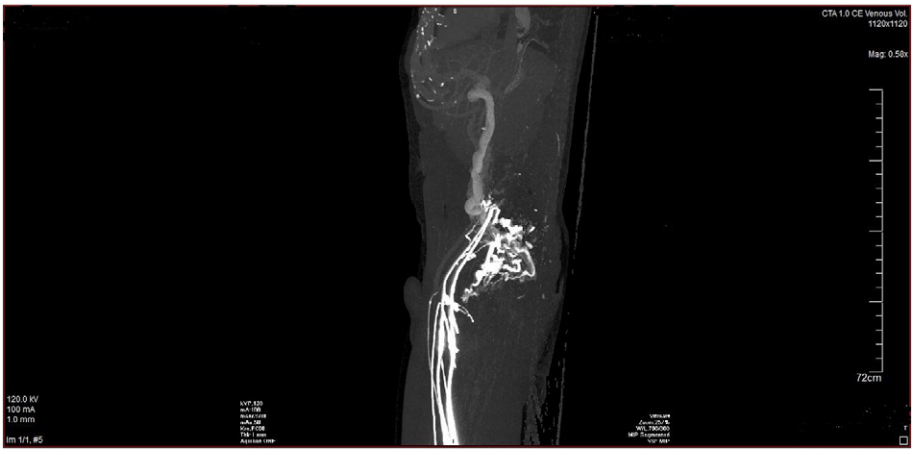


**Fig. 2.** CT angiography image demonstrating absence of the infra- and suprarenal portions of the inferior vena cava with numerous collateral vessels from a lateral view.

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strated in this particular case) has been reported in only eight cases [1]. Furthermore, whether the absence of all or only a segment of the IVC is an embryonic anomaly or the result of perinatal thrombosis with regression and subsequent disappearance of the affected segment is currently a controversial topic [5].

The clinical implications of this interesting congenital anomaly include the following: 1) it can lead to an erroneous diagnosis of deep venous thrombosis [6], 2) the azygous continuation may mimic a right paravertebral or tracheobronchial tumour [6] and an accessory hemiazygos vein may mimic an enlarged aortic knuckle [6], 3) a fatal outcome after ligation of the azygos vein in an undiagnosed patient with an absent IVC during surgery has been reported [7], and 4) congenital absence of the IVC has been described to create difficulties for catheter ablation of arrhythmias via the femoral vein approach [8]. It has been recommended that all patients with an IVC anomaly be screened for a thrombophilic disorder; as in a series by Gayer et al. [2] 7 out of 9 patients with an IVC anomaly and deep venous thrombosis (DVT) had a positive thrombophilic screen [2]. Ruggeri et al. [9] also found that congenital absence of the IVC may be a potential independent risk factor for DVT in the young [9].

In light of this the patient was started on Rivaroxaban (Xarelto®) 10 mg daily.

Isolated absence of the IVC rarely occurs in isolation and is usually associated with other cardiac and/or visceral anomalies, such as dextrocardia, polysplenia or malrotation of abdominal viscera [6].

In summary, this particular case of congenital absence of IVC is unique due to the fact that no other cardiac and/or abdominal visceral anomalies are present. Furthermore, a combined absence of the supra- and infrarenal portions of the IVC is present and as stated only eight cases are found in the literature.

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