SCIENTIFIC LETTER

# AGENESIS OF INFERIOR VENA CAVA

J. L. RIZZARDI
A. BUSCEMI
D. SCHIAVONI
J. FORNASO
A. G. CRUZ
L. PIRE
R. AUN

CORRESPONDENCE:
jlrizzardi@hotmail.com

Dorrego 2392. CP 2000.
Rosario. Hospital Español de Rosario.

The topography of the inferior vena cava (IVC) is divided into 3 segments: prerenal, renal and postrenal. In the embryo, there are 3 pairs of venous vessels combining fusion and resorption. Given the complexity of the process, different anomalies may occur, some of them with great predisposition to thrombosis. This anomaly was first explained in 1973 by Abernethy, who described the azygos continuation of the IVC associated with a mesocaval shunt and dextrocardia in a 10-month-old child. These alterations are rare and occur between the sixth and eighth weeks. The frequency of congenital anomalies of the inferior vena cava is 0.3%-0.5% in the healthy population and 2% in patients with cardiovascular disease. Known anomalies are retroaortic left renal vein, circumaortic left renal vein, double infrarenal IVC, azygos or hemiazygos continuation of the IVC, and absent IVC. Although IVC congenital anomalies have low prevalence, multislice tomography has increased their frequency of appearance. In particular, multislice computed axial tomography (CAT) images have the benefit of establishing the diagnosis of this rare pathology and, if the surgical approach to the retroperitoneum is needed, allow to plan access and avoid damaging abnormal vessels.

This paper refers to the case of IVC agenesis in a 28-year-old male patient with no previous venous thrombosis who sought care for edema of the lower limbs. In the physical examination, he presented soft and painless edema extended bilaterally from the foot to the knee, with 2/4 intensity and no skin color change. No pedal arterial pulses were palpated. He did not show genital edema or collateral venous circulation. The first study was a *venous Doppler scan*, which revealed the presence of thrombi from the popliteal veins to the iliac veins. It was complemented with multislice CAT, confirming the presence of thrombosis in the said venous axis and the absence of the infrarenal IVC (Figure 1). Oral anticoagulant treatment with dicoumarol was decided, succeeding in resolving the edema in a horizontal position.

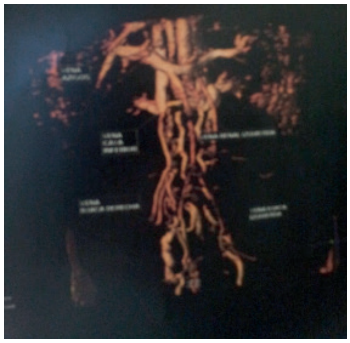

**Figure 1.** Occlusion of the inferior cava vena (on the right) with collateral circulation from the illiac veins to the vena cava at the level of the renal veins.

In subsequent controls with elastic support, he presented edema with induration and increased intensity with daily activity. He did not accept the alternative treatment with fibrinolytics or the anatomical reconstruction of the IVC.

Anomalies of abdominal veins are described according to the embryological sector involved: subcardinal, supracardinal or poscardinal[1] (Table 1). Each of the above vascular structures corresponds to a pair of veins, in reference to the absence of the IVC. In vein formation s, the iliac veins and the vena cava are conditioned by processes of atrophy or develop from them. For this reason, it is observed that the vena cava and the iliac vein resulting from this anomaly are very rare, with their etiopathogenesis being established in the absence of supracardinal development. The other hypothesis is thrombosis followed by fibrosis. The venous bridging in the pelvic area has at the lower end the external and internal iliac veins, which connect with the lumbar veins, which in turn connect with the azygos and hemiazygos system. As a result of venous hypertension and slow venous flow, venous insufficiency and thrombosis occur. This is what the said patient presented when he appeared. The clinical presentation is often asymptomatic, and azygos drainage may be associated with cardiovascular or renal malformations. In young people with thrombosis in both femoral-iliac axes, this determines a strong inclination to infer the presence of the IVC agenesis variant of venous malformations[2-4].

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|---|---|
| Anomalies of the postcardinal veins | Retrocaval/circumcaval uréter |
| Anomalies of the subcardinal veins | Interruption of the inferior vena cava with azygous/hemiazygous continuation |
| Anomalies of the supracardinal veins | Persistence of the left supracardinal vein - Left inferior vena cava |
| | Double inferior vena cava - Persistence of both left and right supracardinal veins |
| Anomalies of the renal segment | Circumaortic venous ring |
| | Retroaortic renal vein |
| | Multiple renal veins |

**Tabla 1.** Classification of anomalies of the inferior vena cava[1].

Doppler scanning is an extremely valuable tool to establish this situation and is complemented with the study of retroperitoneal vessels. Multislice CAT, which reveals with accuracy the malformation present, completes the imaging study[5]. The patient under consideration was young and had both deep venous axes compromised. The first imaging study was a Doppler scan, which showed the occlusion of the femoral-iliac veins. This causes a rise in the base pressure, already increased by the absence of the inferior vena cava.

Renal alterations are important in the surgical treatment of aortic or kidney pathology. Not knowing about this pathology may cause traumatic lesions of venous vessels with hemorrhages. In the course of surgery of ruptured aneurysm of the abdominal aorta, we have found 2 patients with retroaortic left renal vein and one with circumaortic. In the first case, the hematoma of the retroperitoneum made it difficult to visualize the structures and they were damaged causing large hemorrhages difficult to control. The transperitoneal approach is highly likely to lead to this situation, while the retroperitoneal approach averts such possibility due to the displacement of the left retroperitoneum. The detection of this rare anomaly prevents an incorrect diagnosis of retroperitoneal adenopathies or masses and is useful for the surgeon to plan the surgical approach. This situation does not occur in the case of a ruptured aneurysm because multislice CAT scanning is not always possible and due to the presence of blood in the retroperitoneum[4].

In conclusion, the presence of venous thrombosis in young patients, particularly men, should lead to infer the presence of vascular malformations. The thrombotic occlusion of the femoropopliteal vein is associated with this. The absence of IVC should be inferred in the case of young patients with bilateral thrombosis and extension in the femoral-popliteal-iliac axis. ■

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