Intrahepatic agenesis of the vena cava causing venous thrombosis of the lower limbs

Agenesia intrahepática de la vena cava como causa de trombosis venosa de miembros inferiores

To the editor:

Inferior vena cava agenesis (IVCA) is a congenital anomaly of low incidence, associated in its diagnosis with a first episode of deep vein thrombosis (DVT) or as a causal finding after an imaging test.

This is the case of a 27-year-old male patient with no personal history of interest, who consulted the emergency department for generalized pain in the left

leg of 6 hours of evolution, which he related to the previous intramuscular administration of an anti-inflammatory drug in the buttock, for the treatment of low back pain. Examination revealed pain, edema, induration, increased caliber and violaceous coloration of the left lower limb, with preserved sensitivity and strength and femoral, popliteal and pedial pulses present. A D-dimer of 7.5 ng/mL and CRP > 90 mg/L were identified. A venous color Doppler ultrasound of the lower limbs was requested, which showed bilateral venous thrombosis of the lower limbs. This was followed by an abdominal computed tomography (CT) scan, which showed thrombosis of the bilateral inferior cava, iliac and femoral veins (Figure 1A) and a prominent azygos vein (Figure 1B). All these findings are pathognomonic of intrahepatic agenesis of the inferior vena cava. During admission, the study of occult neoplasia was negative, the molecular study of the PAI1 gene (plasminogen activator inhibitor 1) showed heterozygosity of the 4G/5G polymorphism, related to an increased risk of thrombotic phenomena. After discharge and despite treatment with acenocoumarol, he was readmitted 10 days later for a new DVT of the left lower limb.

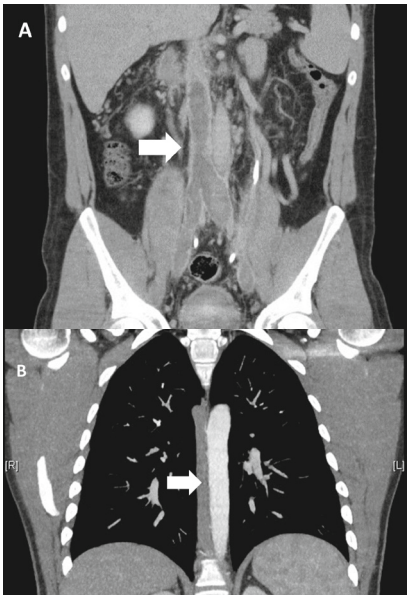

Figure 1. A: Coronal section of the abdomen: thrombosed vena cava and iliac veins. B: Coronal view of the thorax: very enlarged azygos vein.

Intrahepatic agenesis of the inferior vena cava is a rare congenital vascular anomaly, with a very low population incidence (< 0.01%)[1]. It occurs as a result of aberrant development of the venous system during embryogenesis, between the sixth and eighth week of gestation[2]. The embryological alteration that results in agenesis of the hepatic segment of the inferior vena cava is the absence of the hepatic subcardinal anastomosis, which causes atrophy of the right subcardinal vein. As a consequence, blood flow is diverted to the azygos vein through the suprasubcardinal anastomosis.

This anomaly is generally observed in asymptomatic young patients, as an accidental finding, and is sometimes associated with other congenital cardiac or abdominal abnormalities. It is an important risk factor for DVT and pulmonary thromboembolism, with an incidence of 5% in patients under 30 years of age with idiopathic DVT, especially in men and with clinical manifestations of DVT after intense physical exercise[3]. Although bilateral iliacofemoral involvement is less than 10% in DVT, in patients with inferior vena cava malformation it reaches up to 50%, as in the present case, despite the initial clinical presentation of the left leg only[4].

Treatment consists of anticoagulation in the acute episode (sodium or low molecular weight heparin), although the duration of treatment is not well established. Most authors argue for lifelong anticoagulation[5], with the use of elastic support, postural measures, exercise and avoidance of all additional risk factors, such as prolonged immobilization or the use of contraceptives, although all of this has a low level of evidence. In some cases, reconstructive treatment by endovascular bypass has been used, with good results in the published series[4-6].

Carolina Gómez-Hernández[1],
María Isabel Fuentes García[2],
Celestino Hernández-García[3]

[1]*Emergency Department, Hospital Universitario de Canarias, Tenerife, Spain.*
[2]*Radiodiagnostic Department, Hospital Universitario de Canarias, Tenerife, Spain.*
[3]*Cardiology Department, Hospital Universitario Ntra. Sra. de La Candelaria, Tenerife, Spain.*

cgomezh84@gmail.com

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References

1 Parsa P, Lane JS 3rd, Barleben AR, Owens EL, Bandyk D. Congenital agenesis of inferior vena cava: a rare cause of unprovoked deep venous thrombosis. Ann Vasc Surg. 2015;29:1017.e15-8.
2 Lambert M, Marboeuf P, Midulla M, Trillot N, Beregi JP, Mounier-Vehier C, et al. Inferior vena cava agenesis and deep vein thrombosis: 10 patients and review of the literature. Vasc Med. 2010;15:451-9.
3 Gayer G, Luboshitz J, Hertz M, Zissin R, Thaler M, Lubetsky A, et al. Congenital Anomalies of the Inferior Vena Cava Revealed on CT in Patients with Deep Vein Thrombosis. AJR. 2003;180:729-32.
4 Cruz IE, Ferreira P, Silva R, Silva F, Madruga I. Inferior Vena Cava Agenesis and Deep Vein Thrombosis: A Pharmacological Alternative to Vitamin K Antagonists. Eur J Case Rep Intern Med. 2019;6:001310.
5 Reslan OM, Raffetto JD, Addis M, Sundick S. Congenital Absence of Inferior Vena Cava in a Young Patient with Iliofemoral Deep Venous Thrombosis Treated with Ultrasound-accelerated Catheter-directed Thrombolysis: Case Report and Review of the Literature. Ann Vasc Surg. 2015;29:1657.
6 Sarmento JA, Granja S, Madureira AJ, Casanova J, Moreira J. Congenital systemic venous drainage obstruction: A case report. J Card Surg. 2020;35:1703-7.