A 62-year-old man had relapsing episodes of deep vein thrombosis of the lower extremities. The first episode had occurred almost 40 years earlier, followed by at least 7 similar episodes. He was taking anticoagulants orally. He had no other concomitant disease.

On clinical examination dilated veins could be seen on the lateral abdominal wall and periumbilically (Medusa’s head) (Fig. 1). Both lower extremities showed signs of chronic venous insufficiency, with enlarged varicose veins, hyperpigmentation and skin atrophy (Fig. 2).

Chest x-ray, routine hematologic test results and the electrocardiogram were within the normal range. CT of the abdomen showed absence of the inferior vena cava (IVC) below the level of the liver. MR venography confirmed the presence of an extremely narrow structure below the confluence of the right renal vein to the IVC and was considered as the continuation of the IVC (hypoplastic IVC) (Fig. 3). Theazygos and hemiazygos venous systems as well as the left lumbar vein were massively dilated. The dilated left lumbar vein received enlarged collateral veins from the external iliac vein and common femoral vein.

Congenital malformations of the IVC occur in 2%–3% of the population, which reflects the complexity of its embryogenesis especially in its postrenal segment. The commonest congenital anomalies include left positioning and duplication of the vein. The presence of spontaneous, recurrent, usually bilateral, deep vein thromboses of the lower extremities in young patients, which tend to extend to the iliac veins, should alert the physician to this anomaly. However, the development of the deep vein thrombosis as a paraneoplastic manifestation or as a result of a hypercoagulation must be ruled out. Absence or hypoplasia of the IVC can be accompanied by other congenital abnormalities such as dextrocardia or other congenital heart disease. Treatment consists of lifelong anticoagulant therapy.

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References