Agenesis of the infrarenal inferior vena cava

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A 71-year-old man presented to the emergency room with complaints of progressive dyspnoe and pain in both lower extremities. His medical history consisted of multiple idiopathic deep vein thrombosis, familial deafness and cholecystolithiasis. Clinical examination revealed large varicose veins in both lower extremities. A following electrocardiogram, echocardiography and radiography of the thorax were normal (not shown). D-dimeren had risen for which an angio CT of the thorax was performed. This showed no pulmonary embolism (not shown). A venous duplex of the lower extremities revealed insufficiency of both the superficial and deep venous system (not shown). An abdominal CT with intravenous contrast injection showed absence of the infrarenal inferior vena cava (Fig. A), absence of the common iliac veins, enlarged ascending lumbar veins (Fig. B) and prominent anterior paravertebral collateral veins (Fig. C) which lead to a prominent azygos vein (Fig. C). A complex venous collateral circulation was found infrarenally (Fig. A) as well as in the abdominal wall (Fig. D). The suprarenal IVC was normal (Fig. E), formed by confluence of the renal veins. Multiple calcifications in the enlarged internal and external iliac veins confirm a history of deep vein thrombosis (Fig. F).

Comment

The normal inferior vena cava is composed of four segments during embryogenesis: hepatic, suprarenal, renal and infrarenal. The hepatic segment is derived from the right omphalomesenteric vein. The suprarenal segment is formed by the anastomosis of the right subcardinal vein and the right omphalomesenteric vein. The renal segment is derived from the anastomosis of the right subcardinal and the right supracardinal vein. The infrarenal segment is derived from the right supracardinal vein. The azygos and hemiazygos vein are respectively formed from the right and left supracardinal vein, whereas the common iliac veins are derived from the posterior cardinal veins. IVC anomalies are the result of an abnormal regression or persistence of these embryonic veins. Huntington and McLure proposed a classification system and suggested 14 theoretical variations of the IVC of which 11 have been observed to this day. The prevalence of agenesis of the IVC in the general population is thought to be 0.2%-1% (1). These patients are reported to have a higher risk of lower-extremity venous insufficiency and deep vein thrombosis, characteristically with a more proximal location, a younger age of onset and often bilateral involvement (1). IVC agenesis incidence in young people with lower limb deep vein thrombosis is estimated up to 5% (1). In our case the absence of the infrarenal IVC and the common iliac veins with preservation of the suprarenal segment suggests failure of development of the posterior cardinal and supracardinal veins. However some authors consider such an embryonic event unlikely, and suggest perinatal IVC thrombosis as etiology. The radiological importance of this anomaly is to avoid misdiagnosis of these vessels as retroperitoneal lymphadenopathies and to report it in preoperative assessment.

Reference
