

CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Vascular Diseases Quiz – Case 28

A 45-year-old female has been presented with distended and engorged veins, visible on the anterior abdominal wall around the umbilicus. These veins have appeared 5 years ago and gradually developed to the present state (fig. 1).

The patient has been referred to the vascular unit by a general practitioner after a complete blood and biochemistry check; both checks had returned normal results. The patient's past medical history included an episode of deep vein thrombosis (DVT) of the lower extremity. Otherwise she was fit and well.

The patient's skin was of normal color and without skin lesions or rash. Clinical examination revealed superior flow of the distended abdominal wall veins. No abdominal distension or tenderness was present. Liver was of normal consistency, with a soft edge, regular surface, and with normal dimensions. Spleen was not palpable. A number of slightly dilated veins were visible on both the anterior femoral surfaces.

The patient underwent a computed tomography (CT) angiography of the chest and the abdomen (fig. 2).

What is the diagnosis?

Comment

Caput medusae, also known as palm tree sign, is a clinical sign that consists of dilated or engorged periumbilical veins. Causes vary widely from liver pathology and portal hypertension to superior or inferior vena cava obstruction.

In our case, the patient had normal blood biochemistry, a negative spleen and liver palpation and no skin signs; all possible signs of liver pathology. Since liver pathology was to some extent excluded and the patient's medical history included an episode of DVT, an investigation for possible vein pathology was performed. The CT angiography showed enlarged superficial anterior abdominal wall and the inferior vena cava (IVC) was not visible. Vein blood flow from the lower half of the abdomen and the lower extremities is redirected through the abdominal wall collaterals and the azygos-hemiazygos system.

Agenesis of the IVC is the result of either developmental failure of the posterior cardinal and supracardinal veins, or an event of perinatal IVC thrombosis. IVC agenesis is a rare condition and the incidence is approximately 5% in patients under the age of 30 with past history of DVT, and general population prevalence lower or equal to 1%. IVC agenesis is related to thrombophilia, autoimmune

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N. Patelis,¹
M. Doulaptsis,²
C. Klonaris¹

¹First Department of Surgery, Vascular Division, "Laiko" General Hospital, National and Kapodistrian University of Athens, Medical School, Athens
²Second Propedeutic Department of Surgery, Vascular Division, "Laiko" General Hospital, National and Kapodistrian University of Athens, Medical School, Athens, Greece



Figure 1

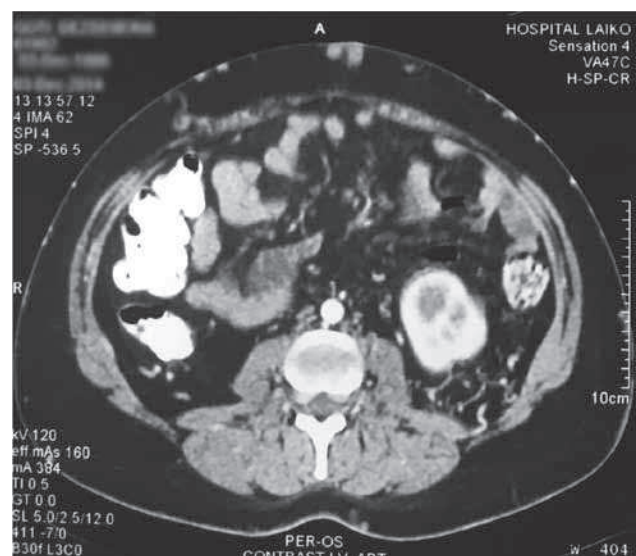


Figure 2

diseases, pro-thrombotic medication and neoplasms. IVC agenesis is usually diagnosed with the use of contrast-enhanced CT scan, as abdominal ultrasound scan has been reported to give ambiguous results. IVC agenesis is related to higher incidence of DVT of the lower extremities, but pulmonary embolism (PE) is not a common complication. No therapy consensus exists, but most reports conclude that the use of IVC filter is not necessary, as PE incidence in these patients is very low. Anticoagulation has showed promising results, but the treatment regime is still to be clarified; elastic stockings should also be used along anticoagulation.

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Corresponding author:

C. Klonaris, First Department of Surgery, Vascular Division, "Laiko" General Hospital, National and Kapodistrian University of Athens, Medical School, Athens, Greece
e-mail: chris_klonaris@yahoo.com