

Complex Malformation of the Inferior Vena Cava

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Rezumat

Malformație complexă de venă cavă inferioară

Malformațiile venei cave inferioare sunt rar raportate la copil, ele fiind de obicei asimptomatice. Aceste malformații sunt generate de o perturbare survenită în timpul dezvoltării embrionare a sistemului venos sau apar datorită trombozei venoase perinatale cu afectarea secundară a dezvoltării sistemului venos. Prezentăm cazul unui pacient în vârstă de 14 ani internat pentru extirparea unui pachet varicos localizat la nivelul peretelui abdominal. Pacientul prezenta inegalitatea membrelor inferioare (lungime și perimetre). Examenle de laborator au evidențiat valori normale; la ecografia abdominală s-a observat hipoplazia segmentului retrohepatic al venei cave inferioare. Angiografia CT a permis stabilirea diagnosticului de malformație complexă a sistemului venos abdominal profund (atrezia segmentului retrohepatic al venei cave, anastomoză cavo-cavă prin venele azygos, formarea anormală a venei cave inferioare cu absența venei iliace stângi). Prezența dilatațiilor varicoase abdominale trebuie să indice necesitatea investigării complexe a pacientului în vederea depistării unor posibile malformații ale venei porte și/sau venei cave.

Cuvinte cheie: malformație venă cavă inferioară, copil

Abstract

Malformations of the inferior vena cava (IVC) are rare presentations, exceptional in children, and are usually asymptomatic. They are caused by disturbances in the embryological formation of the venous system or can develop as a result of perinatal venous thrombosis with secondary impairment of the venous development. We report the case of a 14 year old boy, admitted for pediatric evaluation before undergoing plastic surgery in order to remove superficial varicose veins of the lower abdomen. The patient presents with inequality in circumference and length of the legs. Laboratory investigations are normal and the abdominal ultrasound describes hypoplasia of the retrohepatic segment of the inferior vena cava. The diagnosis of complex malformation of the abdominal deep venous system (retrohepatic vena cava atresia, cavo-caval anastomosis through azygos veins, abnormal formation of the inferior vena cava with the absence of the left iliac vein) was established through a CT angiography. The presence of abdominal varicose dilations should indicate the necessity to closely look for malformations of the portal and/or caval venous systems.

Key words: anomalies of inferior vena cava, child

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Introduction

Venous malformations may occur in the deep venous system, the superficial one or both may be affected. Inferior vena cava (IVC) malformations are rare (0.2-0.3% in healthy individuals, with very high prevalence limits, depending on the specialty of the reporter: 0.07%–8.7%) (1,2). They can be totally asymp-

tomatic or they may be the result of an acute thrombosis. However, superficial varicose dilatations are not an expected outcome of this condition. Since the development of cross-sectional imaging, congenital anomalies of the IVC and its tributaries have become more frequently encountered in asymptomatic patients (3). Vascular structures are usually easily identified on computer tomographic (CT) scans of the abdomen and pelvis, obtained by administering intravenous contrast material.

Case report

We report the case of a 14 year old boy, admitted for pediatric evaluation before undergoing plastic surgery in order to remove superficial varicose veins of the lower abdomen.

The patient's history is unremarkable, with inequality of the legs observed since infancy, for which he was repeatedly evaluated by the Pediatric Surgical Department. A group of suprapubic varicose dilations was present since birth, initially small in size, but which grew in time, becoming cosmetically disturbing (Fig. 1). Plastic surgery was at this moment taken into consideration. The clinical examination documented the presence of the suprapubic group of varicose dilations and unequal legs (in length and circumference): ankle circumference left/right = 26/25 cm, leg circumference left/right = 32/31cm, thigh circumference left/right = 42/41cm.

The laboratory investigations were unremarkable: normal blood count and blood chemistry, negative inflammation markers. The thoracic X-ray revealed an enlargement of the middle mediastinum; this anomaly was not initially integrated in the clinical context.

The abdominal ultrasound visualized normal liver and kidneys, echogenic images in both adrenal glands (calcifications), normal vena porta and supra-hepatic veins. IVC has a decreased retro-hepatic calibre. Multiple lacunar images are present in the IVC area. The IVC is dilated (17 mm in diameter) at the entrance into the right atrium and can be visualized only on a 3.9 cm portion. Distally, the IVC has a small calibre (7 mm) and it appears to be compressed by a number of homogenous, polycyclic echogenic images, with a 9 mm diameter (Fig. 2 A, B, C). Cardiac ultrasound and ECG were normal. The bone marrow biopsy was normal, revealing



Figure 1. Group of dilated veins in the inferior region of the abdomen

no atypical cells.

At this point the differential diagnosis included IVC compression (abdominal aortic aneurism, solid tumors: lymphoma, renal or colonic cancer) or IVC obstruction through venous thrombosis or malformation. The patient presented with no specific signs and symptoms related to a neoplasia (fever, weight loss, pallor, and fatigue), no lymph nodes enlargement, hepatosplenomegaly was not observed, the inflammation markers were negative and the bone marrow biopsy indicated no atypical cells. In order to exclude venous thrombosis, coagulation tests were ordered and specific prothrombotic factors were also found to be normal (antiphospholipid antibodies, S protein, C protein, antithrombin III).

An abdominal and pelvic CT angiography was performed and revealed: IVC atresia from above the right renal vein to below the suprahepatic venous confluent, compensatory dilation of the azygos vein, anastomotic perivertebral veins and in the paravertebral muscles, significant anastomosis in the pelvis and subcutaneous, malformation of the IVC with the absence of left iliac vein (Figs. 2, 3).

Therefore the diagnosis of intraabdominal complex deep venous system malformation was established: retro hepatic IVC atresia, cavo-caval anastomosis through the azygos



Figure 2. Abdominal ultrasound. (A): Lacunar images in the IVC area; (B): Dilated IVC at the entrance of the right atrium; (C): IVC compressed by polycyclic echogenic images

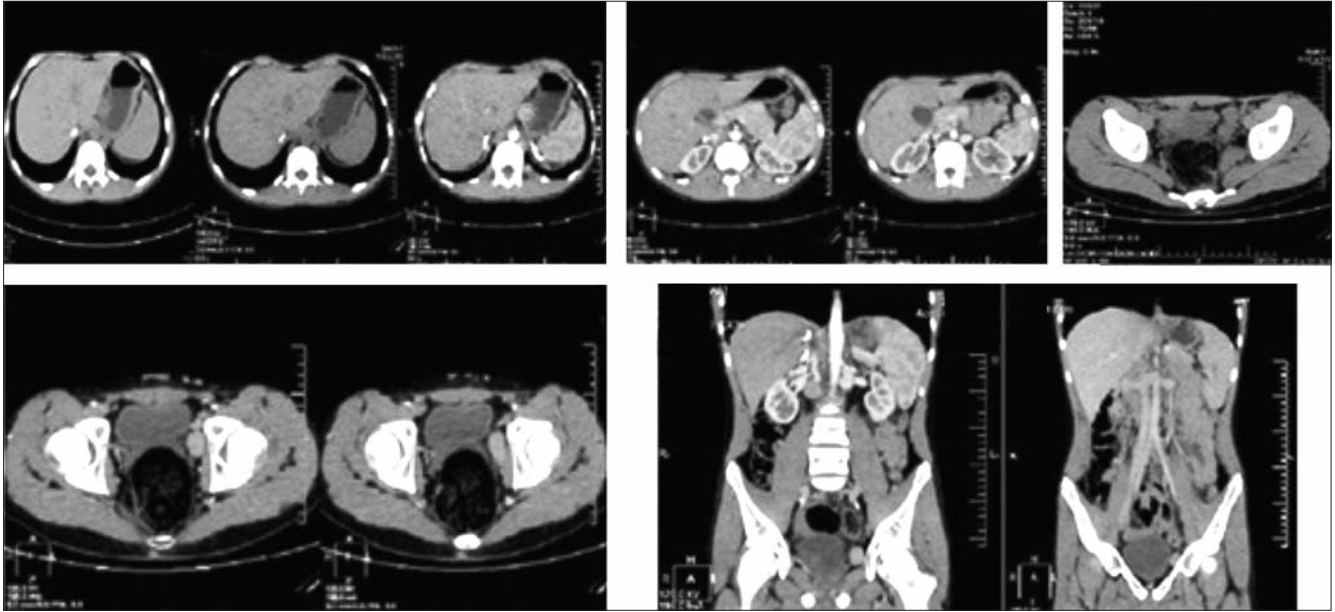


Figure 3. (A, B, C, D, E): Abdominal and pelvic angioCT

veins, varicose dilations of the pelvic deep venous system and of the parietal pubic veins, malformation of the IVC and absence of the left iliac vein.

Discussions

Malformations of the IVC may be: malrotation and malposition on the left side of the IVC, IVC continued with intrathoracic azygos vein (a variant of this malformation was diagnosed in our case), circumaortic left renal vein, duplication of the IVC with normal intrathoracic route or duplication of the IVC continued with azygos or hemiazygos veins, absence of the infrarenal IVC or of the entire IVC.

The superficial varicose veins of the lower abdomen are suggestive for abdominal vascular impairment with collateral circulation development. One of the most common and helpful clues in establishing the diagnosis of IVC malformations is well developed and possibly dilated intrathoracic hemiazygous and/or azygos continuations. These collateral circulations as well as other retroperitoneal venous pathways are usually well developed before symptoms present (4,5,6).

Therefore the suspicion of vascular malformation was the only viable one and, in order to establish a diagnosis, the decision to perform an angio-CT was taken (4).

Two hypotheses could be taken into consideration regarding the origin of the malformation: changes during the process of embryogenesis or perinatal thrombosis that consecutively influences the postnatal development of the venous system, with compensatory collateral circulation.

In our case we suspect a perinatal thrombosis with its consequences because of the presence of calcifications seen on the abdominal ultrasound in both adrenal glands (7,8).

Hemodynamically, when there is a malformation /obstruction of the IVC, the following changes take place:

compensatory dilatation of the azygos venous system develops in order to aid the venous return from the lower body, cavo-caval shunts may also arise. The cavo-caval shunts may determine vascular dilatations of the superficial subcutaneous vessels, dilatation of paravertebral plexus or dilatation and disturbed circulation of intraabdominal vessels (2).

The patient with IVC malformations may present symptoms of lower extremity venous insufficiency (legs inequality) or idiopathic deep venous thrombosis. If the deep venous collateral system is sufficiently developed, reflecting the compensated long duration of the process drains the venous blood from the lower extremities. Venous stasis secondary to inadequate blood return through collaterals is likely to be prevented (9).

Our patient had a very large collateral pool draining the lower body venous blood through intra-abdominal and subcutaneous varicose packs, without thrombosis at the time of diagnosis.

Regarding the treatment (the patient was referred for investigations before performing plastic surgery), surgery of the varicose pack is firmly contraindicated, the ligation of collateral vessels might have major circulatory consequences and could even lead to death. Unidentified anomalies of the inferior vena cava can lead to significant morbidity during the surgical exploration. This knowledge can prevent hemorrhagic complications from occurring during surgery (8,10).

A second therapeutic specification is the need for prophylactic anticoagulant therapy. Authors have different opinions, but there is a tendency to recommend anticoagulant therapy only in those cases with venous thrombosis (11).

Conclusions

The presence of abdominal varicose dilations should warn the

pediatrician regarding possible cavo-caval shunts and/or caval malformations. These patients should be carefully investigated (especially through imaging studies) in order to establish an accurate diagnosis and to determine the indication for surgical intervention.

Imaging studies represent the key of the diagnosis, complete data being given by contrast computer tomography or angiography.

Surgery should not be recommended without prior investigations, because removing anastomoses may have major hemodynamic consequences and may even lead to death. Regarding prophylactic anticoagulation, there is no clear indication. The recommendation is for patients with venous thrombosis.

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