An unusual cause of lumbar pain after physical exercise: Caval vein duplicity and its detection by ultrasound

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Abstract This paper deals with two recent cases of Inferior Caval Vein (IVC) duplicity, congenital anomaly often but not always asymptomatic that should be considered in the differential diagnosis of flank pain when other more common disorders have been excluded. Here are described the clinical and surgical significance together with the typical sonographic findings useful for the diagnosis of IVC duplicity. Particular attention is given to a peculiar and easily repeatable sonographic aspect associated with IVC duplicity, not previously reported in Literature and that we called the “Neptune’s sign”.

Keywords Color doppler sonography · Inferior vena cava · Caval vein duplicity · Congenital anomalies

Sommario In questo lavoro vengono riportati 2 recenti casi di duplicità della Vena Cava Inferiore (VCI) che è un’anomalia congenita spesso, ma non sempre, asintomatica e che deve essere considerata nella diagnosi differenziale di dolore al fianco, quando le altre cause più comuni sono state escluse. Oltre all’importanza clinica e chirurgica della duplicità della VCI ne vengono descritti gli elementi ultrasonografici utili per la diagnosi. Particolare

attenzione è dedicata ad un aspetto ecografico peculiare, facilmente ripetibile, associato con la duplicità della VCI e non precedentemente riportato in letteratura che abbiamo chiamato il “segno di Nettuno”.

Introduction

The IVC derives from a complex process of regressions and anastomoses between different embryonic veins. Its improper completion is not so rare and, as a consequence, leads to the genesis of anatomic anomalies [1].

Among them the IVC duplication is the most common and though generally described as asymptomatic it can be responsible for back pain, mimicking a renal colic as described here below. Moreover, it can lead to thrombosis and can determine anomalous circulation of blood to the heart [2].

As in abdominal, retroperitoneal or aortic vascular surgery an excessive bleeding is often caused by erroneous venous injury, the IVC duplication has always to be ruled out before attempting these procedures [3, 4].

The recurrent pulmonary embolism after caval filters placement is another complication related to the lack of awareness and pre-procedural recognition of the IVC duplicity [5].

The abdominal color Doppler sonography plays a pivotal role in the study of the IVC and may allow the early detection of its duplicity and other congenital anomalies often associated with this venous variant.

The purpose of this series of clinical cases is to describe the sonographic features of IVC duplicity, emphasizing the importance of the abdominal ultrasound examination in not neglecting IVC imaging.

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Case descriptions

Case 1

A 33-year-old man reported to our Unit a history of occasional right flank pain exacerbated by physical activity and interpreted in the past as a consequence of reno-ureteral colic episodes.

A direct abdominal X-ray was performed excluding any form of lithiasis urinary disorders.

The abdominal sonography, performed with an Acuson S3000TM ultrasound system (Siemens Healthcare, Germany), confirmed regular kidneys and the absence of lithiasis or hydronephrosis.

A color Doppler ultrasonography revealed no renal or hepatic pathological findings.

An abdominal transverse scan identified two circular structures on either side of the abdominal aorta with a major diameter in the right one (Fig. 1a); the color Doppler sonography showed the presence of flow inside both structures leading to the suspect of a venous abdominal anomaly (Fig. 1b).

With the left coronal scan (Fig. 1d) was detected a “trident” aspect of the abdominal vessels with two IVC distinct segments each on the upper and lower side of the aorta leading to the diagnosis of IVC duplication.

CT scans with contrast confirmed the anatomical anomaly with the left IVC segment originating from the left external iliac vein and being tributary of an ectatic left renal vein while the left internal iliac vein was instead confluent in the right IVC segment (Fig. 1c, e).

The IVC right segment was confirmed to be dominant.

Fig. 1 **a** Transverse grayscale sonogram showing two circular structures on either side of abdominal aorta. *RIVC* right inferior vena cava, *LIVC* left inferior vena cava, *AA* abdominal aorta. **b** Color Doppler sonogram confirming vascular nature of these structures in the context of IVC duplication. *RIVC* right inferior vena cava, *LIVC* left inferior vena cava, *AA* abdominal aorta. **c** CT abdominal transverse scan at the same level. **d** Color Doppler sonogram left coronal scan showing two segments of IVC on either side of abdominal aorta. Similarity with a trident can be appreciated (“Neptune’s sign”) right at the top. Legend: *RIVC* right inferior vena cava, *LIVC* left inferior vena cava, *AA* abdominal aorta. **e** CT abdominal coronal multi planar reformation. IVC duplication with the dominance of the right IVC segment can be evaluated

Fig. 2 **a** Color Doppler sonogram right coronal scan showing the two IVC segments and their anatomical relationship with the renal arteries along with Neptune's sign. Legend: *RIVC* right inferior vena cava, *LIVC* left inferior vena cava, *AA* abdominal aorta, *RRA* right renal artery, *LRA* left renal artery; **b** Spectral Doppler shows a venous signal inside one of the segments. **c** NMRI coronal scan (BTFE) shows Neptune's sign Legend: *RIVC* right inferior vena cava, *LIVC* left inferior vena cava, *AA* abdominal aorta, *BTFE* balanced steady-state free precession, *EIA* external iliac artery, *EIV* external iliac vein

Case 2

A 26-year-old man came under our observation for an history of back pain and intermittent hematuria appearing after physical exertion and explained in the past as renal colic induced by microlithiasis.

While a bilateral kidney grayscale sonography was unremarkable, a transverse gray scale sonogram performed in the epigastric region revealed the presence of 2 paired and apparently cystic structures on either side of abdominal aorta while coronal scans clarified the tubular forms of these structures (Fig. 2a).

The color Doppler showed the presence of a flow in all these structures confirming that they were abdominal vessels and evoking the aspect of a “trident” when they were insolated on a right coronal plane together with the abdominal aorta (Fig. 2a). The spectral Doppler ultrasonography identified the presence of a venous signal in both of them (Fig. 2b); therefore an anomaly of the IVC with 2 component parts could be suspected and then better defined by the recognition of anatomical relationships between double IVC and renal arteries (Fig. 2a).

NMRI sagittal and coronal scans were then performed to clarify the complete course of the double IVC which, on the IVC left side, has a segment originating from the left external iliac vein and terminating at the level of the left renal veins (Fig. 2 c).

Discussion

The IVC duplication, though generally asymptomatic, can be clinically relevant because it can be responsible for back pain sometimes related to physical activity, as detected in both patients. It can rarely lead to thrombosis (with about only 6 cases reported in literature) but can determine anomalous circulation of blood to the heart [2].

The IVC is the result of a complex embryogenetic process during the period from 6 to 10 weeks of gestation when the development, anastomosis, fusion and regression of different vascular segments (the vitelline vein, the paired posterior cardinal, supracardinal and subcardinal veins) take place.

Considering the complexity of this process, it's not so rare its improper completion and, as a consequence, the genesis of at least 14 anatomic anomalies, the most common of which are the duplication of the inferior vena cava (IVC), its transposition, the retroaortic left renal vein and the circum-aortic left renal vein. [1, 6].

The IVC duplication, as described above, has a prevalence varying from 0.2–0.3 to 2.8 % according to different authors [7, 8] and it derives from the persistence of both supracardinal veins; it can be partial or total leading to an asymmetric or symmetric duplication, respectively [7].

In most cases, the absence of regression of the left caudal supracardinal vein determines a communication between the left common iliac vein and the left renal vein.

Although many variations have been reported, the IVC duplication commonly occurs below the renal veins with the formation of two IVC infrarenal segments which can be different in size.

The two IVC parts run on both aorta sides, then join in the suprarenal IVC at the level of the renal arteries, while the IVC ipsilateral duplication is very rare.

The two IVC segments generally communicates with the right IVC segment, it being the dominant one, and with the left IVC that ends in the renal vein [7].

While in the past anatomical entities like IVC duplicities were recognized only during operative procedures on the retroperitoneum, in recent years pre-operative diagnoses are even more common [9, 10], thanks to Computed Tomography (CT) scans and Ultrasound (US) examinations.

Even if US depends on the operator and sometimes is limited in the study of the infrahepatic IVC for the presence of overlying bowel gas, it can show the flow in the IVC in normal and pathological conditions and its role is crucial for the study of caval segments and their congenital variants in pediatric patients [11].

With the abdominal gray-scale sonogram in transverse plane, 2 paired anechoic and apparently cystic structures can be appreciated on either side of abdominal aorta [9] but in coronal or sagittal sections the tubular structure can be shown together with the course of IVC segments (Fig. 1). The dilatation of IVC after the Valsalva maneuver can confirm the venous nature of this double structure.

If a coronal scan in a plane passing both through the abdominal aorta and IVC segments is performed, an aspect that resembles the shape of a trident can be identified leading to the definition of what we call “the Neptune’s sign”.

With the use of color Doppler the presence of flow inside both the caval system and the adjacent aorta can be confirmed making the “Neptune’s sign” even more clearly visible (Figs. 1d, 2a).

The spectral Doppler analysis reveals the phasicity and pulsatility which are typical of central veins in each of two IVC segments.

IVC duplication can be associated with many other congenital anatomical abnormalities, some of them well recognizable with abdominal US, such as abdominal aneurysm, congenital absence of the right kidney and anomalies of the right renal vein course.

Other congenital anomalies to be considered are the congenital heart disease, the retrocaval or transcaval ureter and the rectal ectopia, as reported in association with the double IVC [7].

Although the IVC duplicity can be fairly well recognized with an ultrasound by experienced operators aware of its existence, it can be missed or even misinterpreted if the IVC is not correctly visualized in a sagittal or coronal plane.

As a consequence, the IVC variant placed on the left of the abdominal aorta can be confused with a mass or a lymphadenopathy, a saccular aortic aneurysm, a left pyeloretic dilatation, a retroperitoneal cyst and an abnormal form of a gonadal vessel [7, 10, 11].

From its first description in 1793, there has been a growing awareness of the importance of the existence of IVC duplicity and its pre-operative recognition in order to reduce abdominal vascular surgery complications [1, 12].

For the surgeon a double IVC that usually joins the aorta anterior part, can represent a technical challenge because of the exposure of the aorta just below the origin of the renal arteries. As a consequence, it has to be considered before aortic vascular surgery in order to prevent serious hemorrhagic complications [1]. Both nephrectomy, adrenalectomy, and sympathectomy may be dangerous if performed in patients with unsuspected IVC duplicity and IVC ligation because misinterpreted as aberrant gonadal vessel.

IVC duplication requires further precautions for the surgeon in particular settings such as laparoscopic live donor nephrectomy. Whereas the left-sided kidney is usually preferred (for the longer length of its vein), the presence of double IVC may change this scenario shortening the left renal vein and changing the planning in favour of the right kidney [1, 13, 14].

With the exception of surgical procedures in other specific conditions such as when attempting placement of caval filters, IVC duplicity has to be preventively excluded. If confirmed, alternative placement locations or additional filters have to be chosen to prevent pulmonary embolism relapse (for the presence of collateral pathways for emboli) and to avoid the possibility of uselessness of the procedure [5].

Conclusions

IVC duplication, just like other IVC anatomic congenital anomalies and their tributaries, may remain undetected. The lack of awareness and recognition of these clinical entities among surgeons and radiologists can have serious clinical consequences.

The prompt diagnosis of IVC duplicity by US can be crucial to identify the cause of flank pain (especially when it appears after physical exertion) when other more common disorders have been excluded, can reduce hemorrhagic complications before retroperitoneal and abdominal vascular surgery and can prevent recurrent pulmonary embolism before vena cava filter placement.

Compliance with ethical standards

Conflict of interest The authors Pierpaolo Di Nicolò, Luca Zanoli, Michele Figuera, Antonio Granata have no conflict of interest.

Human and animal studies The clinical cases reported in this article do not contain studies with human or animal subjects performed by any of the authors.

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