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Case Report

Recurring hematuria revealing absence of infrarenal segment of the inferior vena cava [☆]

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ABSTRACT

Abnormal development of the inferior vena cava is a relatively rare condition. We report the case of a 47-year-old woman presenting with an absent infrarenal segment of the inferior vena cava revealed by recurring episodes of gross hematuria. This entity probably resulted from perinatal acquired thrombosis rather than from a congenital anomaly and is associated with compensatory dilation of collateral venous pathways. This extremely rare hemorrhagic presentation is presumably caused by rupture of small dilated renal or vesical veins. Ignorance of this atypical presentation can lead to erroneous or delayed diagnoses.

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Introduction

Abnormal development of the inferior vena cava (IVC) is quite rare and is often associated with congenital malformations. When symptomatic, it usually manifests through thrombotic events. Exceptionally, it can manifest through a hemorrhagic presentation and become a real diagnostic challenge for clinicians.

Case presentation

A 47-year-old woman was referred to our urology department by her general practitioner for recurring hematuria. She experienced at least 3 episodes of gross hematuria in the last 2 years. Those episodes were associated with acute pelvic pain and were self-limited. No clear causal factor was found. She had a history of urolithiasis and multiple superficial and deep

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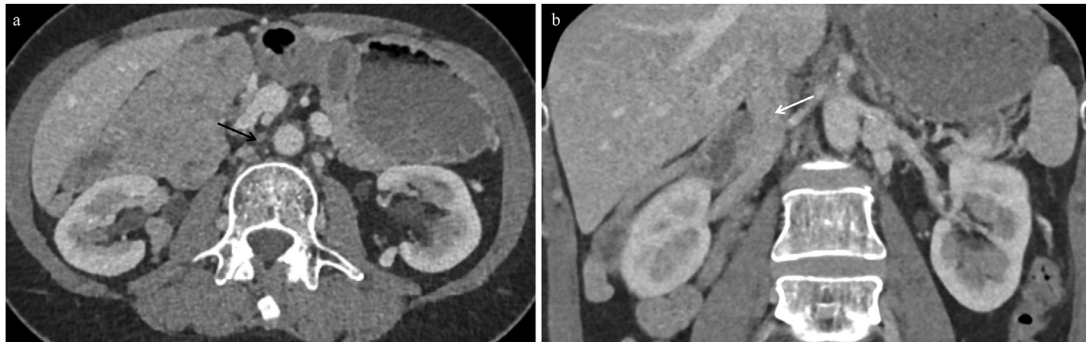


Fig. 1 – (A, B) Portal phase contrast-enhanced CT revealing (A) absence of the infrarenal segment of the IVC (black arrow) and (B) preserved patency of its superior segments (white arrow).

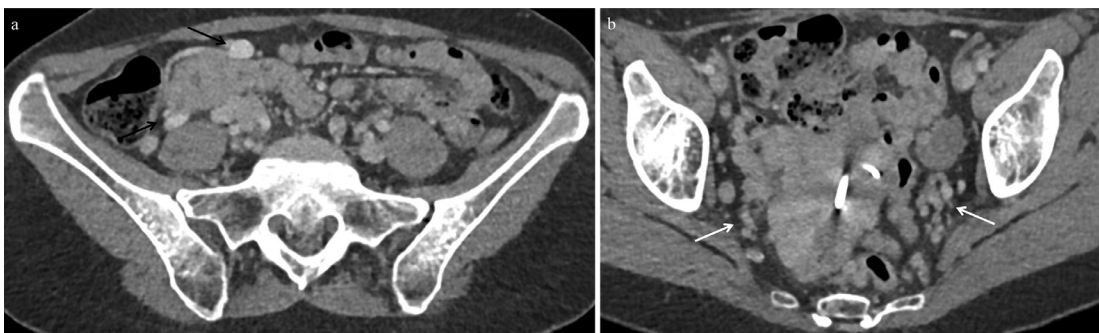


Fig. 2 – (A, B) Portal phase contrast-enhanced CT showing dilated collateral venous circulation: (A) mesenteric veins (black arrows) and (B) iliac veins (white arrows).

vein thrombosis related to factor V Leiden thrombophilia. Physical examination was normal. After ruling out leading causes of hematuria (laboratory tests including chemical and microbiological urinary analysis were irrelevant), the patient was referred by her urologist for a computed tomography (CT).

Imaging findings

A multiphase contrast-enhanced CT of the abdomen and pelvis was performed but it found no urinary tract anomaly and no urolithiasis in particular. However, CT revealed the absence of the infrarenal segment of the inferior vena cava and preserved patency of its superior segments (Fig. 1). In addition it showed dilated azygos and hemiazygos veins, lumbar veins, renal veins, gonadal veins, iliac veins, both superior and inferior mesenteric veins, rectal venous plexuses and epigastric veins (Fig. 2). Of note, the right gonadal vein drained into the right renal vein and the left renal vein drained into the hemiazygos vein (Fig. 3). We schematized the abnormal abdominal venous circulation of our patient (Fig. 4). The rest of the examen was irrelevant to the case.

Discussion

Abnormal development of the inferior vena cava is seen in approximately 4% (0.07-8.7) of the population. Among this group,

IVC agenesis is the rarest, reported in only 0.0005%-1% [1,2]. In this subcategory, isolated infrarenal agenesis of IVC is the rarest. Indeed, partial IVC agenesis most commonly involves the suprarenal segment (90% of cases) and is usually associated with other congenital malformations (mainly the heart or the spleen) [1-3].

Inferior vena cava is classically divided in 4 segments, from upstream to downstream: infrarenal, renal, suprarenal and hepatic. Embryologically, it is formed between the sixth and the eighth week of gestational age by the successive development and involution of 3 pairs of embryonic veins: in order, posterior cardinal veins, subcardinal veins and supracardinal veins [4,5]. In summary, the hepatic segment is derived from the right vitellin vein, the supra-renal segment is derived from the anastomosis between the right vitellin vein and the right subcardinal vein, the renal segment is derived from the anastomosis between the right subcardinal vein and the right supracardinal vein, and the infrarenal segment originates from the right supra-cardinal vein although the exact origin of this last segment remains controversial.

Development of collateral circulation is the typical anatomic compensation for absence or thrombosis of a venous segment. Our patient CT shows all of the possible redistribution pathways of the venous flow in case of absence of the infrarenal segment of the IVC: deep pathway through the azygos, hemiazygos veins and ascending lumbar veins; median pathway through the renal and gonadal veins; portal pathway through the inferior mesenteric vein; and superficial pathway through the epigastric veins.

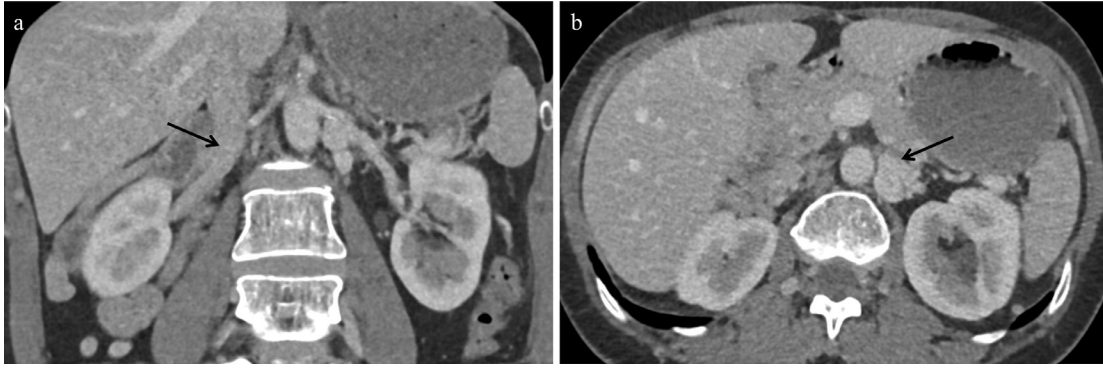


Fig. 3 – (A, B) Portal phase contrast-enhanced CT showing renal veins drainage (arrows): into the IVC for the right renal vein (A) and into the hemi-azygos vein for the left renal vein (B).

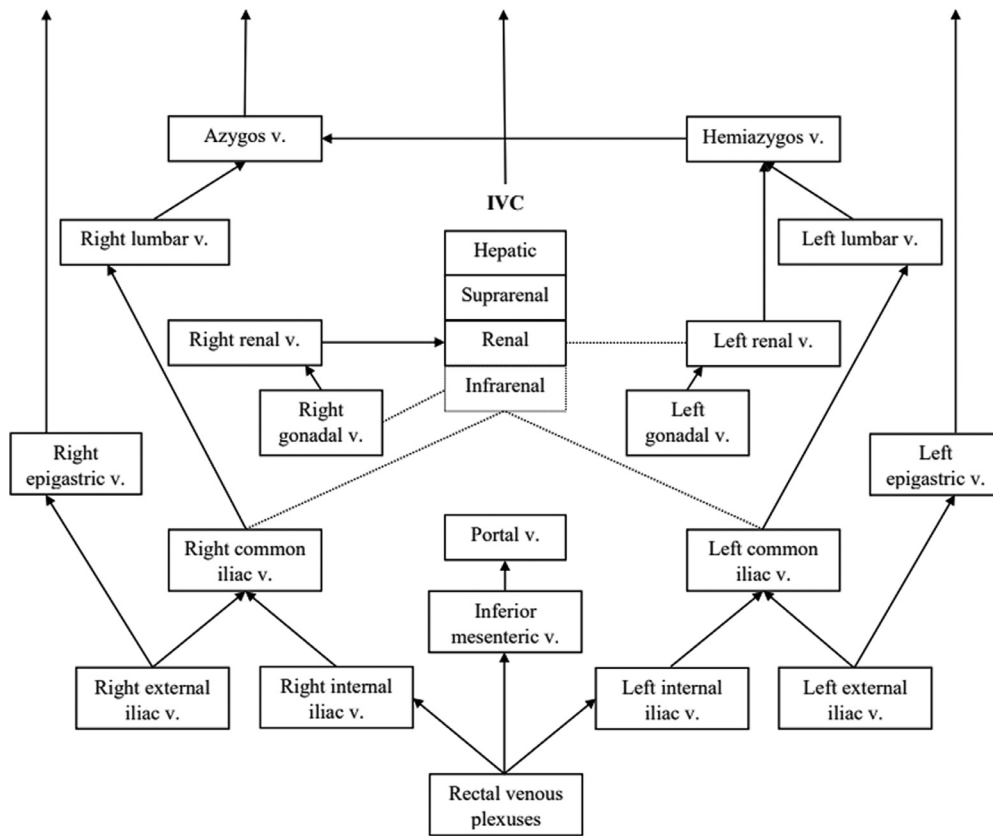


Fig. 4 – Schematized venous circulation of our patient showing missing infrarenal IVC segment (dotted borders) and absent/non-patent venous pathways (dotted lines). IVC, inferior vena cava; v., vein(s).

Partial IVC agenesis cases most often are asymptomatic. When symptomatic, they usually present with proximal deep vein thrombosis of the lower limbs. Rarely, symptoms are related to compression of adjacent structures – such as lumbar or sacral nerve root compression, intestinal tract compression... – secondary to venous hypertension within the aforementioned derivation pathways [1-3]. Exceptionally, venous hypertension can lead to rupture of small veins and subsequent hemoptysis or hematuria as previously illustrated in the literature [6]. We reasonably assumed that was the same

mechanism for hematuria in our patient in the absence of any other cause of hematuria.

Our patient had heterozygous factor V Leiden thrombophilia. This mutation is more often found in patient with IVC agenesis than in the general population: 16.4% vs 4.8% respectively [2,7]. This is in agreement with the common theory that the absence of the infrarenal segment of the IVC would result from perinatal acquired IVC thrombosis. This hypothesis is based on the fact that no single embryonic event can explain the absence of the infrarenal segment of the IVC. Accordingly,

Ramanathan et al. reported the sole case to our knowledge of regression of a properly formed infrarenal segment of the IVC in association with a perinatal IVC thrombosis [3,8]. Moreover, our patient left renal vein drained into the hemiazygos vein. This is consistent with flow redistribution through the reno-azygo-lumbar arch, suggesting an absent or non-patent connection between the left renal vein and the renal segment of the IVC. As the left renal vein and the infra-renal segment of the IVC originate from different embryonic structures, the most probable explanation is an originally normal connection between IVC and renal veins with an acquired thrombosis of the infrarenal and renal segments of the IVC, and the left renal vein. Interestingly, Mc Donald et al. reported already in 1974 an association between neonatal IVC and renal vein thrombosis. Secondary to the thrombosis, patency was probably maintained or rapidly restored between the right renal vein and the IVC whereas the infrarenal IVC and the connection between the left renal vein and the renal IVC remained occluded and underwent sclerosis with subsequent opening of the reno-azygo-lumbar collateralization [7,9].

In conclusion, we report a case of absent infrarenal IVC segment revealed by recurring gross hematuria presumably caused by rupture of small dilated renal or vesical veins. Knowing and understanding this extremely rare association can prevent erroneous or delayed diagnoses.

Patient consent

We obtained written and informed consent from the patient for publication.

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