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Case Report

Pelvic congestion syndrome due to agenesis of the infrarenal inferior vena cava

Terence Menezes[a], *Ehsan A. Haider*[a,b,*], *Faten Al-Douri*[a,b], *Mohamed El-Khodary*[a,b], *Ishaq Al-Salmi*[a]

[a] *Department of Radiology, St. Joseph's Healthcare Hamilton, Hamilton, Canada*
[b] *Department of Radiology, Faculty of Health Sciences, McMaster University, Hamilton, Canada*

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ABSTRACT

The inferior vena cava (IVC) is the main conduit of venous return to the right atrium from the lower extremities and abdominal organs. Agenesis of the IVC has an incidence of <1% in the general population [1], although it has been reported in the literature as occurring in up to 8.7% of the population [2]. Patients with absent IVC may present with symptoms of lower extremity venous insufficiency [6], idiopathic deep venous thrombosis [7], or pelvic congestion syndrome. To our knowledge there have only been a few cases reported in the literature of agenesis of the IVC associated with pelvic congestion syndrome [3,10,11]. We present another interesting case of pelvic congestion syndrome due to absent IVC.

1. Introduction

Our patient is a 34-year-old female who presented with gradually worsening pelvic pain. She had a medical history significant only for varicose veins in the lower extremities. No other significant previous medical history was elicited. No prior imaging was available. No prior ultrasound had been performed. She had never been pregnant and there was no significant obstetrical history, Gravidity Term Preterm Abortion Living (GTPAL). A contrast-enhanced magnetic resonance imaging (MRI) of the abdomen and pelvis was performed, and showed complete absence of the infrarenal inferior vena cava (IVC) with significantly tortuous and dilated vessels in the pelvis. Pelvic veins were dilated up to 2 cm. This case shows how absence of the infrarenal IVC can present as pelvic congestion syndrome.

2. Case report

A 34-year-old female presented with gradually worsening chronic, dull, and aching pelvic pain and menorrhagia over a few years. No back pain or radiculopathy was present. There was no history of thrombolysis, intervention, or surgery in the past. Gynecological examinations in the past were unremarkable. Her only relevant medical history was some lower extremity varicose veins that were treated conservatively. A

* Corresponding author at: Department of Diagnostic Imaging, St. Joseph's Healthcare Hamilton, 50 Charlton Avenue E, Hamilton, ON L8N 4A6, Canada.
E-mail addresses: tmenezes1@gmail.com (T. Menezes), ehaider@stjosham.on.ca (E.A. Haider), faldouri@stjosham.on.ca (F. Al-Douri), melkhoda@stjosham.on.ca (M. El-Khodary), dr.ish@hotmail.com (I. Al-Salmi).
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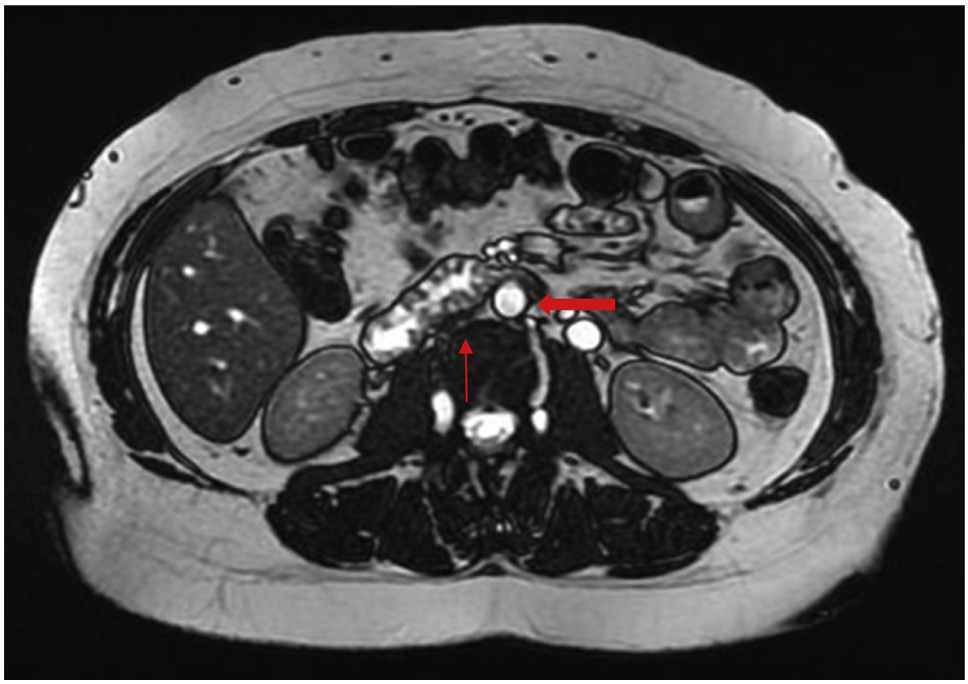

Fig. 1 – Axial 2D FIESTA image at the level of the infrarenal abdominal aorta shows no adjacent IVC to the right (thin arrow). Instead there is some T2 hypointense soft tissue that may represent atretic IVC or collaterals. A normal positioned and normal caliber aorta is seen to the left (solid arrow).

contrast-enhanced MRI of the abdomen and pelvis was performed, and showed a complete absence of the infrarenal IVC (Fig. 1). Tortuous dilated vessels were seen in the myometrium and pelvis (Figs. 2 and 3). Pelvic veins were dilated up to 2 cm. The right external and internal iliac veins joined and then immediately drained into a large right lumbar collateral (Fig. 4). The left external and internal iliac veins appeared to drain into small paravertebral venous channels. The left gonadal vein was dilated measuring 12 mm (Figs. 4 and 5). It drained into the left renal vein. The right gonadal vein was replaced by multiple tortuous vascular channels, which appeared to drain into the right renal vein. Both renal veins drained into the infrahepatic IVC. The infrarenal IVC was absent and replaced by multiple tortuous vascular channels, which communicated with the paravertebral and ascending lumbar venous plexuses. No filling defects or thrombus was identified. The remaining portions of the visualized abdomen and pelvis were unremarkable. The intrahepatic IVC was incompletely imaged and the suprahepatic IVC was out of the field of view.

3. Discussion

Agenesis of the IVC has an incidence of <1% in the general population [1], although it has been reported in the literature occurring in up to 8.7% of the population [2]. IVC developmental abnormalities occur at 6–10 weeks of gestation when the infrahepatic IVC develops from three pairs of embryonic veins: the postcardinal, subcardinal, and supracardinal veins [3]. The IVC is composed of four segments: hepatic, suprarenal, renal, and infrarenal. The hepatic segment is derived from the vitelline vein. The suprarenal segment develops from the right subcardinal vein by formation of the subcardinal-hepatic anastomosis. The renal segment derives from the right supra-subcardinal and postsubcardinal anastomoses. The infrarenal segment develops from the right supracardinal vein. In the thoracic region, the supracardinal veins give rise to the azygos and hemiazygos veins. In the abdomen, the postcardinal veins are progressively replaced by the subcardinal and supracardinal veins but persist in the pelvis as the common iliac veins [4]. Absence of the entire posthepatic IVC implies that all three-paired venous systems failed to develop properly. Absence of the infrarenal IVC suggests failure of development of the posterior cardinal and supracardinal veins. It is difficult to identify a single embryonic event that causes either of these scenarios, which leads to controversy as to whether these conditions are true embryonic anomalies or the result of perinatal IVC thrombosis [5].

Patients with absent IVC may present with symptoms of lower extremity venous insufficiency [6], idiopathic deep venous thrombosis [7], or pelvic congestion syndrome. Although patients with the absence of infrarenal IVC are generally asymptomatic, the most common clinical symptom is Deep Vein Thrombosis (DVT) [8], which is typically treated with anticoagulation, though in our patient no signs or symptoms of DVT were elicited. Reduced venous flow, venous hypertension, and thrombophilia are felt to play a role in the development of DVT in these cases. Our patient did, however, have

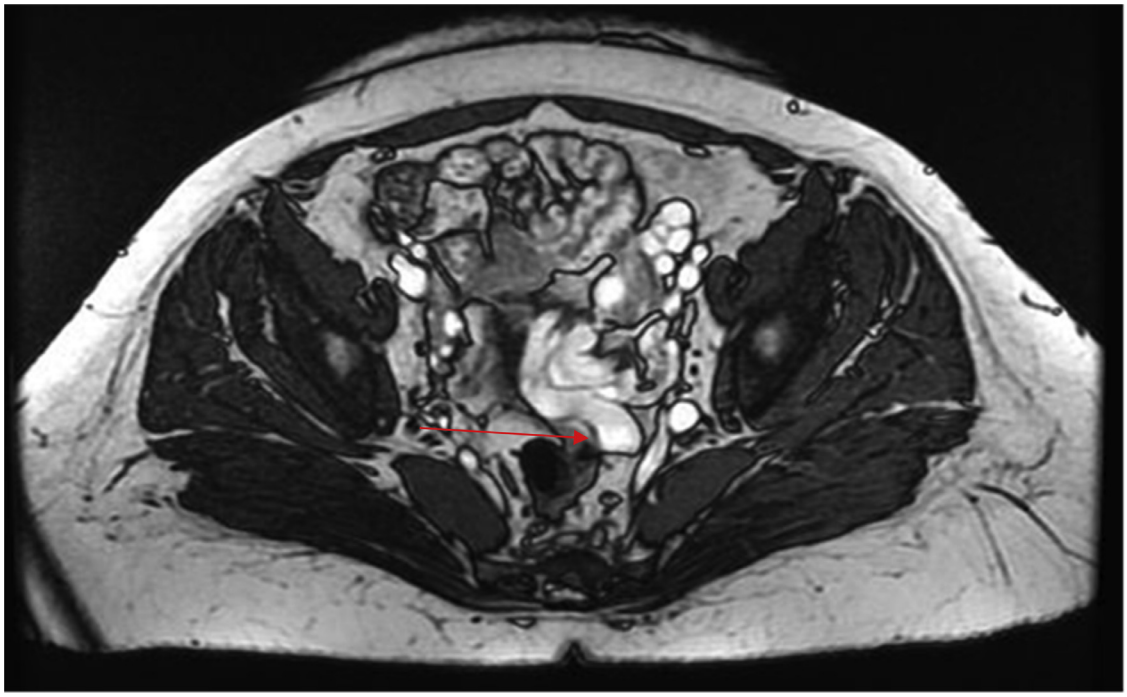

Fig. 2 – Axial 2D FIESTA shows dilated parametrial veins up to 2 cm (arrow). On T2-weighted magnetic resonance images, pelvic varices appear as multiple hyperintense dilated tubular structures around the uterus, ovaries, and pelvic sidewall.

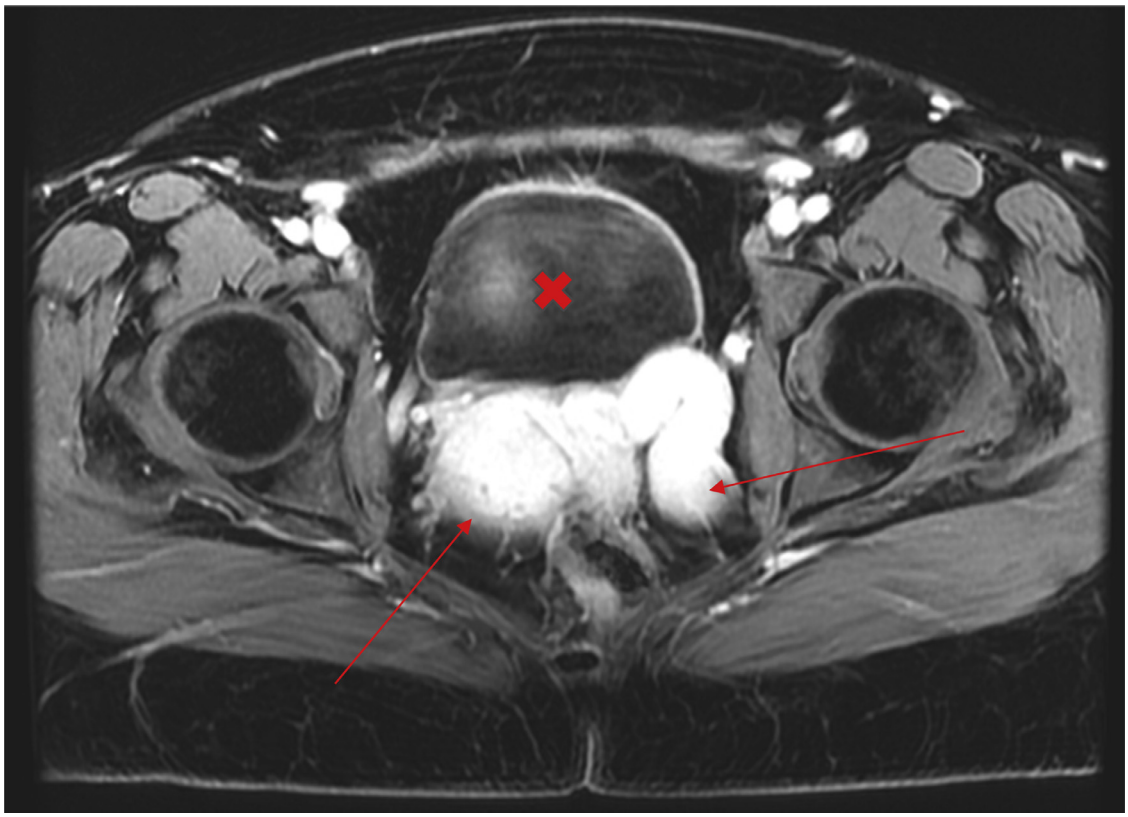

Fig. 3 – Contrast-enhanced, fat-saturated T1 image showing dilated tortuous vessels in the pelvis. The bladder is seen anteriorly (x).

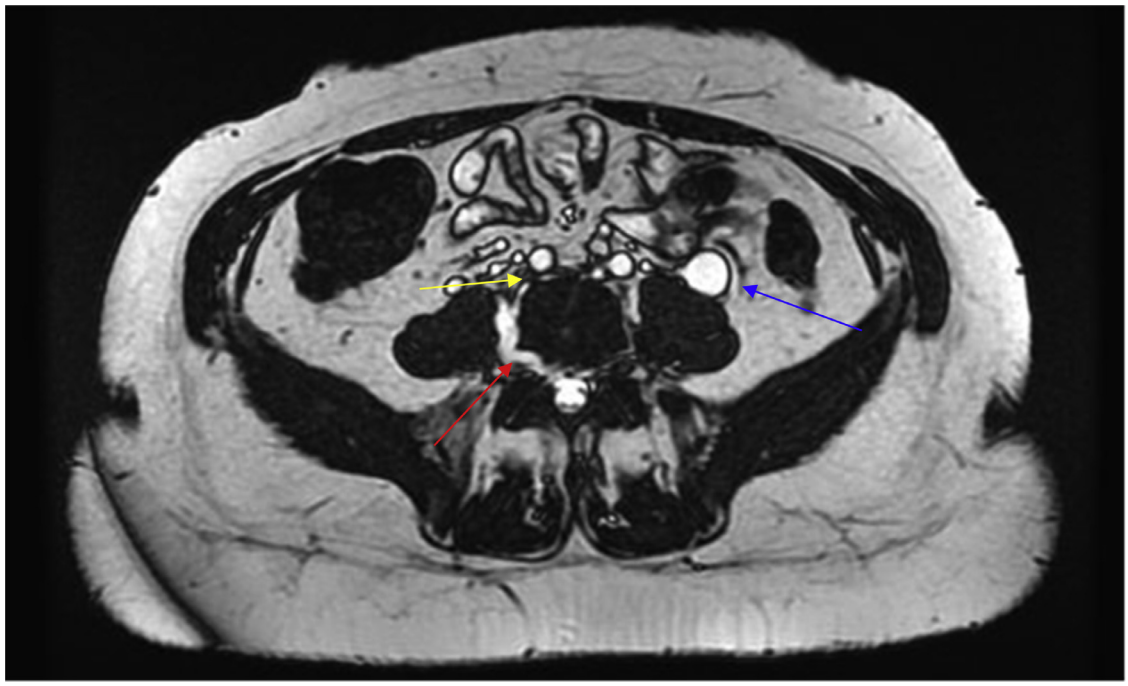

Fig. 4 – Axial 2D FIESTA image at the expected confluence of the right internal and external iliac veins shows no right common iliac vein (yellow arrow). Instead the right internal and external iliac veins drain into a large right lumbar vein (red arrow). A significantly dilated left gonadal vein measuring 12 mm in diameter is also noted (blue arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

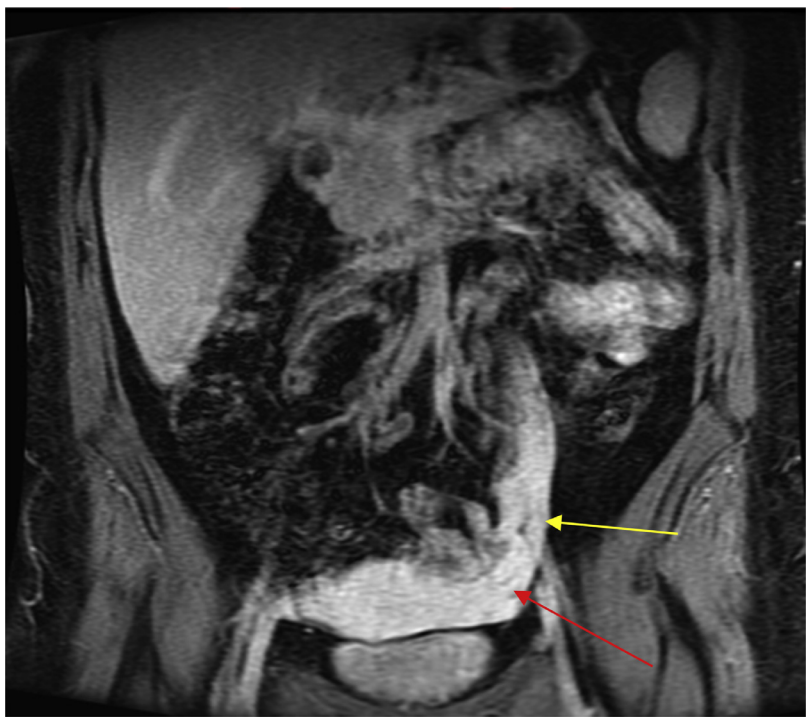

Fig. 5 – Coronal LAVA FLEX sequence shows enhancing dilated pelvic vessels (red arrow) with a dilated left ovarian vein (yellow arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

lower extremity varices, which were attributable to an absent IVC.

Pelvic venous congestion is a relatively common and overlooked condition that can be painful and debilitating for many women. It was first described in 1857 by Louis Alfred Richet to describe a chronic, dull pelvic pain, pressure, and heaviness that persisted for >6 months [9]. These symptoms are attributable to dilated, tortuous, and congested pelvic veins. Symptoms are exacerbated with prolonged standing, menses, and increased abdominal pressure. The symptoms are usually worse at the end of the day.

The association between absence of IVC and pelvic varices causing PCS is weak and has been documented only in a few case reports [3,10,11]. Congenital anomalies of the venous system leading to obstruction of venous drainage are a well-recognized cause of this condition. Absence of IVC leads to development of multiple collaterals predominantly via four large routes. These include the gonadal venous system, the paravertebral venous plexus, the hemorrhoidal plexus, and the superficial pathway through superficial abdominal veins. All these pathways ultimately drain into the Superior Vena Canva (SVC) or the portal venous system (hemorrhoidal plexus) [12]. In our patient, the main collaterals were the gonadal venous system and the paravertebral venous plexus.

Cross-sectional imaging plays a vital role in the diagnosis of absent IVC and other associated developmental variations. Although Contrast Enhanced Computed Tomography (CECT) imaging in venous phase provides anatomical variation in details, multiplanar pelvic MRI also has excellent image quality, providing high tissue contrast and spatial resolution in depiction of pelvic anatomic detail and vasculature. MRI may reveal evidence of other causes of chronic pelvic pain, such as endometriosis, which may not be visible at ultrasound, or other uterine, adnexal, urologic, gastrointestinal, or musculoskeletal causes of pain. The diagnostic criteria for MRI and computed tomography proposed by Coakley et al. [13] consist of at least four ipsilateral parauterine veins of varying caliber, at least one measuring >4 mm in diameter, or an ovarian vein diameter greater than 8 mm.

Treatment options include coil embolization of the gonadal vein and surgical ligation of the ovarian vein [14]; however, the presence of multiple collaterals between iliac and ovarian venous plexuses may cause recurrence of symptoms. In patients with an absent IVC, embolization or ligation may not be a viable option as the gonadal veins may be the only source of collateral flow back to the central venous system. An optimal treatment option for symptomatic relief is open surgical bypass to decompress pelvic collateral vessels by diverting flow from the lower extremities. Several case reports have shown the efficacy of vena cava bypass. For example, Zhou and colleagues have successfully treated a similar case with a common femoral vein to the suprarenal IVC bypass using a bifurcated polytetrafluoroethylene graft, with rapid symptom resolution, and the patient remained symptom free 6 months later. This was the first reported case describing a surgical strategy for isolated infrarenal IVC absence in a symptomatic patient [15].

4. Conclusion

Complete absence of IVC is rare entity and can be a cause of pelvic congestion syndrome. Our patient presented with features of pelvic congestion syndrome. Cross-sectional MRI can play a role in diagnosis of absent IVC and associated pathology. The mainstay of treatment is surgical bypass therapy. Our case is one of the few reported in the literature.

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