Case Report

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

Terence Menezes\textsuperscript{a}, Ehsan A. Haider\textsuperscript{a,b,*}, Faten Al-Douri\textsuperscript{a,b}, Mohamed El-Khodary\textsuperscript{a,b}, Ishaq Al-Salmi\textsuperscript{a}

\textsuperscript{a}Department of Radiology, St. Joseph’s Healthcare Hamilton, Hamilton, Canada
\textsuperscript{b}Department of Radiology, Faculty of Health Sciences, McMaster University, Hamilton, Canada

**Article Info**

Article history:
Received 4 March 2018
Accepted 2 April 2018
Available online 6 October 2018

Keywords:
Absence of IVC
IVC anomalies
Pelvic congestion syndrome
DVT
Varices

**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.

© 2018 The Authors. Published by Elsevier Inc. on behalf of University of Washington.

This is an open access article under the CC BY-NC-ND license. (http://creativecommons.org/licenses/by-nc-nd/4.0/)

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
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 infrahepatic 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 suprarecardinal veins [3]. The IVC is composed of four segments: hepatic, suprarenal, renal, and infrarenal. The hepatic segment is derived from the vitelline vein. The suprarecardinal segment develops from the right subcardinal vein by formation of the subcardinal-hepatic anastomosis. The renal segment derives from the right suprarecardinal and postsubcardinal anastomoses. The infrarenal segment develops from the right suprarecardinal vein. In the thoracic region, the suprarecardinal veins give rise to the azygos and hemiazygos veins. In the abdomen, the postcardinal veins are progressively replaced by the subcardinal and suprarecardinal 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 suprarecardinal 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 (ECT) 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 paraxilenterine 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.

REFERENCES

[1] Sneed D, Hamdallah I, Sardi A. Absence of the retrohepatic inferior vena cava: what the surgeon should know. Am Surg 2005;71:502–4.
[2] Cho BC, Choi HJ, Kang SM, Chang J, Lee SM, Yang DG, et al. Congenital absence of inferior vena cava as a rare cause of pulmonary thromboembolism. Yonsei Med J 2004;45(5):347–51.
[3] Bass JE, Redwine MD, Kramer LA, Huynh PT, Harris JH Jr. Spectrum of congenital anomalies of the inferior vena cava: cross-sectional imaging findings. Radiographics 2000;20(May–June(3)):639–52.
[4] Chuang VP, Mena CE, Hoskins PA. Congenital anomalies of the inferior vena cava. Review of embryogenesis and presentation of a simplified classification. Br J Radiol 1974;47:206–13.
[5] d’Archambeau O, Verguts L, Mylle J. Congenital absence of the inferior vena cava. J Belg Radiol 1990;73:516–17.
[6] Debeg E, Tielemans Y, Jolie E, Van den Brande P. Congenital absence of inferior vena cava. Eur J Vasc Surg 1993;7:201–3.
[7] Bass JE, Redwine MD, Kramer LA, Harris JH Jr. Absence of the infrarenal inferior vena cava with preservation of the suprarenal segment as revealed by CT and MR venography. AJR Am J Roentgenol 1999;172:1610–12.
[8] Chee YL, Culligan DJ, Watson HG. Inferior vena cava malformation as a risk factor for deep venous thrombosis in the young. Br J Haematol 2001;114:878–80.
[9] Knuttinen MG. Pelvic venous insufficiency: imaging diagnosis, treatment approaches, and therapeutic issues. AJR Am J Roentgenol 2015;204(February(2)):448–58.
[10] Nichols JL, Gonzalez SC, Bellino PJ, Bieber EJ. Venous thrombosis and congenital absence of IVC in a patient with menorrhagia and pelvic pain. J Pediatr Adolesc Gynecol 2010;23(1):e17–21.
[11] Wei Z, Wade R, Alan L, Li J. Successful surgical management of pelvic congestion and lower extremity swelling owing to absence of infrarenal inferior vena cava. Vascular 2005;13(6):358–61.
[12] Ramanathan T, Michael T, Hughes D, Richardson JA. Perinatal inferior vena cava thrombosis and absence of the infrarenal inferior vena cava. J Vasc Surg 2001;33:1097–9.
[13] Coakley FV, Varghese SL, Hricak H. CT and MRI of pelvic varices in women. J Comput Assist Tomogr 1999;23:429–34.
[14] Rundqvist E, Sandholm LE, Larsson G. Treatment of pelvic varicosities causing lower abdominal pain with extraperitoneal resection of the left ovarian vein. Ann Chir Gynaecol 1984;73(6):339–41.
[15] Zhou W, Rosenberg W, Lumsdon A, Li J. Successful surgical management of pelvic congestion and lower extremity swelling owing to absence of infrarenal inferior vena cava. Vascular 2005;13(6):358.