Anomalous iliac vein development: a case report and literature review

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Introduction

Pelvic venous anomalies currently form a rare subset of congenital vascular malformations set apart from commonly known defects of vena cava development. With the advent of cross-sectional imaging, these venous variations have become more identified in live subjects. The embryology governing the development of the pelvic drainage system is complex with anomalous growth defects present in nearly 20% of subjects [1]. Proper vascular evolution requires the orchestration of various moving systems with both gross tactile and fine signaling recognition patterns utilized for cell-specific tissue generation. Alterations within this precise developmental pathway can lead to derangement of the pelvic outflow system, including inferior vena cava (IVC) anomalies and variations in iliac vein development. Morita et al. [2] reported seven cases of anomalous drainage of a separated right internal iliac vein (RIIV) into the left common iliac vein. We describe an additional case of a separated right internal iliac vein with drainage into the proximal left common iliac vein.

Case Report

30-year-old male presented to our institution with complaints of abdominal discomfort and constipation of four
days duration. He reported associated abdominal bloating and pressure but denied nausea, vomiting, melena, hematochezia, and weight loss. MiraLax and Dulcolax suppositories were used without relief. There were no aggravating factors. Prior medical and surgical history was significant only for a previous appendectomy.

Physical exam revealed an afebrile patient with normal vital signs. Abdominal exam was significant for abdominal fullness and decreased bowel sounds; there was no tenderness with palpation. Routine hematologic, biochemical, and urinalysis results were all within normal limits. Upright and supine anteroposterior abdominal radiographs demonstrated non-dilated small bowel loops with air-fluid levels in the right lower quadrant. Computed tomography (CT) exam of the abdomen and pelvis with intravenous contrast was subsequently performed, demonstrating no acute intra-abdominal abnormality. Incidentally, a separated RIIV draining into the proximal left common iliac vein was discovered (Figures 1 and 2). A duplicated right renal collecting system was also noted with cortical scarring of the lower pole moiety and associated caliectasis.

The patient improved clinically while in the emergency department without intervention. He was discharged home in stable condition with instructions for a soft oral diet and early re-evaluation with his primary care physician.

Figure 1. Coronal CT abdomen and pelvis with IV contrast. Demonstrates the separated trunk of the RIIV draining into the left common iliac vein. (White arrow = right common iliac vein. Red arrow = right common iliac artery. Green arrow = separated trunk of the RIIV. Blue arrow = left common iliac vein. Black arrow = left common iliac artery.)
Discussion

In the fifth week of gestation, three primary venous groups - omphalomesenteric, umbilical, and cardinal veins - dictate the broader development of the body’s venous system \[3\]. With respect to the pelvic region, the cardinal veins begin to show differentiation in the seventh week characterized by subgroup anastomosis between three primary draining systems: subcardinal veins collecting from the kidneys, sacrocardinal veins gathering from the lower extremities, and supracardinal veins receiving blood from the body walls \[4\]. Contiguously, segments of the IVC begin to add structure on the right as they form common junctures with cardinal subsets on the left. Distal to this region, an oblique transverse anastomosis between sacrocardinal segments provides basis for creation of the left common iliac veins. The evolution of the iliac venous system diverts an escalating volume of blood into the right longitudinal veins. In doing so, the leftward drive for blood flow diminishes thereby potentiating the eventual disappearance of the left venous collecting system \[5\]. Throughout this larger process gained by gross development of vasculature, the overarching signaling pathways encoded by genetic markers, such as Eph4, cause the release of vascular endothelial growth factor to better guide the building blocks of vessels \[6\]. Oftentimes, the evolution of these vessels can become distorted leading to anomalous vein development.

Some of the most documented and well-studied pathologies of pelvic venous drainage leading to severe dysfunction regard congenital IVC formation \[7-9\]. IVC hypoplasia can arise from a multitude of locations. Typically, this occurs in the subhepatic region surrounding the anastomosis of common iliac and retrohepatic veins \[4, 10-11\]. This abnormality can lead to dysfunction of venous hemodynamics causing trophic ulcers, peripheral venous thrombosis, and renal hydronephrosis. Furthermore, it may be associated with congenital heart disease.

Previous methods of detecting vascular anomalies in the abdomen and pelvis relied on post-mortem cadaver dissection, which was largely not helpful in guiding clinical decision-making for a population. Intravascular venography is an invasive exam that defines venous pathology well; however, it may not at times clearly distinguish accessory or anomalous veins aside from the dominant draining outflow vein. Current imaging technology with ultrasonography (US), CT, and magnetic resonance imaging (MRI) allows for a non-invasive means of vascular evaluation. US is useful for evaluation of peripheral neck, arm, and leg veins; however, deep chest or abdominal pelvic veins are not well seen with US. Cross-sectional imaging with either CT or MRI can provide detailed evaluation of the intra-abdominal and pelvic veins, as well as assess for other malformations that can be seen with hereditary disorders such as Klippel-Trenaunay-Weber syndrome.

Correct identification and classification of pelvic venous abnormalities is important to the clinical approach of patients diagnosed with deep venous thrombosis (DVT). For example, a patient with a newly diagnosed DVT and duplicated IVC who cannot receive anticoagulation therapy may require placement of a suprarenal IVC filter or placement of two filters – one in each IVC – to prevent pulmonary embolism \[12\]. There are also important secondary surgical implications to consider when identifying anomalous pelvic veins, including but not limited to anterior lumbar interbody fusion, renal transplantation, hysterectomy, aortoiliac surgery, and intravascular procedures, and when staging large retroperitoneal masses \[5, 12-17\].

Previously, Morita et al. \[2\] described seven cases of pelvic vein malformation characterized by separation of the trunk of the RIIV draining into the central left common iliac vein. In each of these cases, the separated right internal iliac venous trunk led to the development of a second RIIV originating proximal to the bifurcation. We report the eighth case to add to the literature of RIIV duplication caused by truncal
separation manifested by proximal origin. While the patient in this case presented with non-specific abdominal symptoms, incidental discovery of the separated RIIV may help guide future clinical management of this patient.

Conflicting interests

The authors have declared that no conflict of interests exist.

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Author Contributions

P.R. performed the original reading of the patient’s Computed tomography scan. P.T.R. performed the literature review and analyzed the case. P.T.R. and P.R wrote the manuscript. P.R. (Poyan Rafiei), P.T.R. (Patrick T. Reeves).

Abbreviations

IVC: Inferior vena cava; RIIV: Right internal iliac vein; CT: Computed tomography; US: Ultrasound; MRI: Magnetic resonance imaging; DVT: Deep vein thrombosis.

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