Case study

Double Vena Cava In Hemorrhagic Polycystic Kidney Disease Patient

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ABSTRACT

Aims: We review the literature pertaining to the implications of the rare anomaly of double vena cava.
Presentation of Case: A 20 year old male treated for polycystic kidney disease went for a computed tomography (CT) renal for microscopic hematuria and incidentally noted a double inferior vena cava.
Discussion: Double inferior vena cava is a rare congenital venous anomaly. It usually does not cause any symptoms and is incidentally seen during radiological imaging such as computed tomography for other medical conditions.
Conclusion: The discovery of this rare embryological feature in a patient during retroperitoneal surgery may increase the risk of surgery and intraoperative complications.

Keywords: inferior vena cava, congenital, kidney disease, embryology, imaging

1. INTRODUCTION

Double inferior vena cava (IVC) is a rare congenital venous anomaly with a reported incidence ranging from 0.2 to 3% [1]. It was first described by Lucas in 1916 [2]. It does not cause any symptoms and is usually found incidentally when an imaging is done for other medical conditions, just like our present case.

The embryological development of IVC involves the posterior cardinal, subcardinal and supracardinal veins during the 5th week of gestation. The posterior cardinal veins remain in the pelvis to become common iliac veins. The right supracardinal vein persists to form the infrarenal IVC. The right subcardinal vein persists to develop into the suprarenal IVC segment. The left subcardinal vein and the left supracardinal vein regress completely. An alteration of any one of this process will end up with anatomical anomaly of the IVC. [3] As in our case, the duplication of IVC likely results from persistence of the left supracardinal vein which fails to regress.

We incidentally found a double IVC in a young man investigated for microscopic hematuria. A literature review was done on the implication of this anomaly.

2. PRESENTATION OF CASE

A 20 year old male was diagnosed with polycystic kidney disease following investigation for his sudden onset of difficulty to micturate. Physical examination was unremarkable. Laboratory investigation showed normal level of urea (4.8 mmol/L) and creatinine (95 umol/L). Urinalysis revealed traces of blood. A multiphase renal CT was obtained and showed bilateral enlarged multiple cysts of varying sizes in both kidneys, some with wall calcifications and some with non-enhancing hyperdense component within which is in keeping with hemorrhagic cysts. The CT also showed a duplicated left sided inferior vena cava which extended from the left common iliac vein joining the left renal vein near the renal hilum, crossing anterior to the aorta to join the right sided inferior vena cava. (Figure 1)

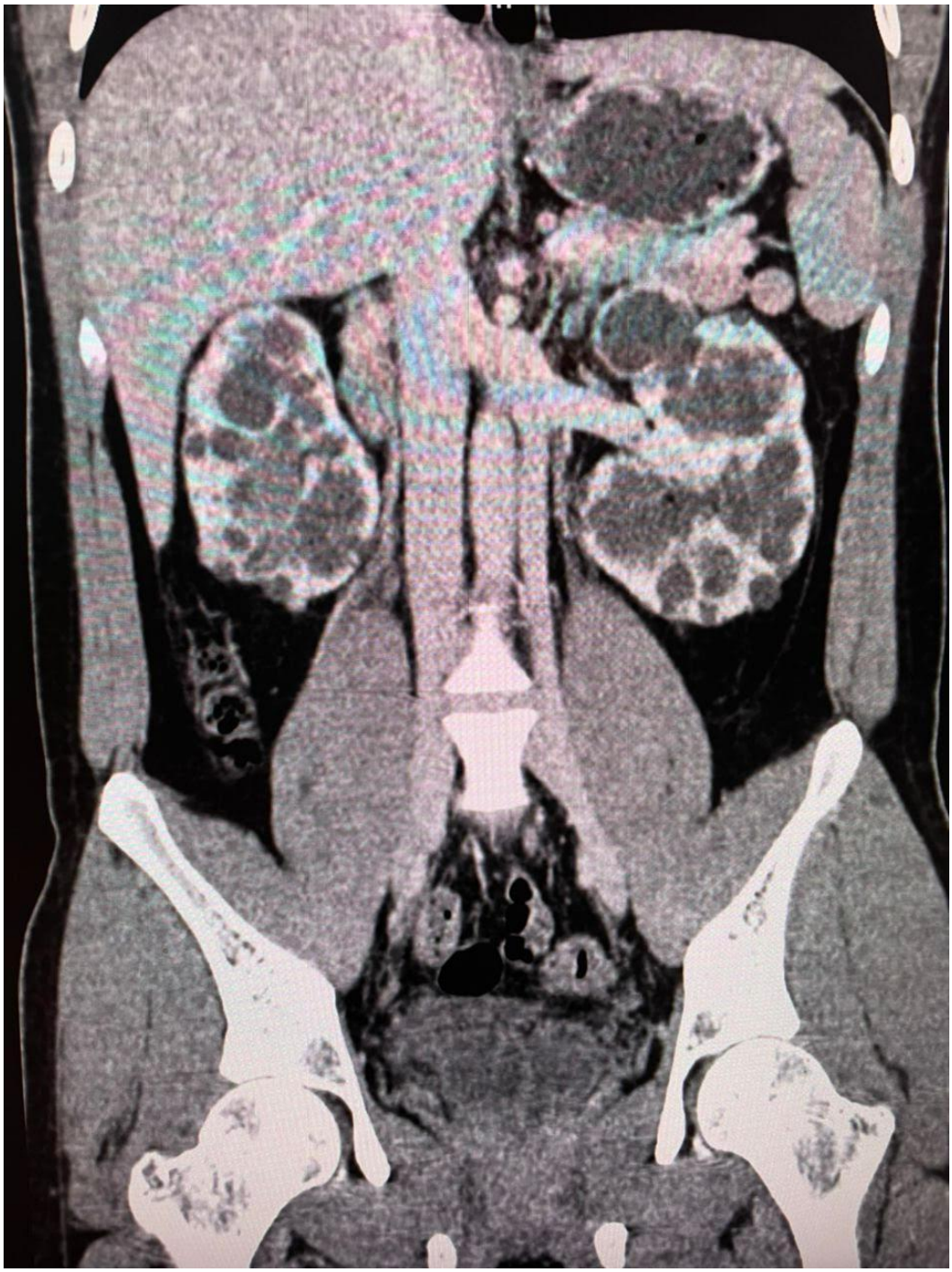

Figure 1. CT scan demonstrating left-sided vena cava (blue arrow) draining into the left renal vein (white arrow). The right-sided IVC can also be visualized separately from the left-sided vena cava.

Patient was prescribed with ural sachets and the symptoms eventually resolved. Our patient has the possibility of developing renal failure or extensive bleeding from the hemorrhagic polycystic kidney disease which may require surgery. Therefore, it is imperative for the operating surgeon to be aware of the rare venous anomaly of IVC as to prevent any unfavorable outcomes throughout the operation.

3. DISCUSSION

There are various structural anomalies that can occur to the IVC. Double IVC is among one of them. Bass et al described other anomalies such as left IVC, retroaortic left renal vein, circumaortic left renal vein and the absence of hepatic segment of IVC with azygos continuation of IVC among others. [1] In some cases, more than one variation can co-exist with double IVC. In most circumstances, IVC developmental anomalies are strongly associated with renal vein developmental anomalies.[9]

In these anomalous veins, blood flow can become altered as they become dilated and tortuous. As a consequence, they are more likely to undergo thromboembolic events. Several case reports of thromboembolic diseases have been reported in patients with duplicated IVC, as noted in our literature search. However, Milani et al. reported that patients with double IVC alone may not be specifically predisposed to development of acute thromboembolism, as other routine pre-existing conditions, such as malignancy, contraceptive hormones, and genetic predisposition, may predispose the risk. [5] To date, there is no existing data to guide the anticoagulation treatment for thrombosis in patients with congenital anomaly. In attempting caval filtration, literature reported that different strategies are needed, depending on the presentation of illness. Asma et al suggested that placement of single filter at suprarenal IVC via the right jugular venous approach is safe and effective to prevent pulmonary embolism in patients with extensive venous thromboembolism or those contraindicated from use of anticoagulation. [6]

Another possible implication that may occur is the nutcracker phenomenon in which the left renal vein (LRV) is entrapped by the abdominal aorta (AA) and the superior mesenteric artery (SMA). A retroaortic LRV or a circumaortic double LRV may be entrapped by the abdominal aorta and the vertebral column. This phenomenon has commonly manifested as hematuria, flank or pelvic pain, congestion syndrome in women, or varicocele. Although extremely rare, the coexistence of double vena cava and nutcracker syndrome may result in clinical symptoms that often require medical intervention. Anna et. al reported a case of nutcracker syndrome in a patient of left IVC, where the diaphragmatic movements during respiratory phase can cause reduction in the angle between AA and SMA. [10] The diagnosis is made when the angle between the abdominal aorta and the superior mesenteric artery is less than 35° from CT or magnetic resonance imaging (MRI).[8] Treatment options include open surgery or endovascular approach if symptoms worsen as it may lead to renal failure if left untreated.

Radiologists should also be aware of the different types of anomalies of the IVC in order to avoid misinterpretation in imaging. There are cases reported that double vena cava was misdiagnosed as lymphadenopathy, aortic aneurysm, retroperitoneal cysts and transposition of IVC. Klimberg et. Al reported a duplicate inferior vena cava that mimicked retroperitoneal lymphadenopathy in a patient with testicular embryonal cell carcinoma. [7] Misinterpretation in imaging may significantly affect the treatment for these patients.

When performing abdominal surgery, the presence of double IVC needs to be acknowledged as it can lead to significant bleeding in a retroperitoneal surgery, as it might be accidentally injured if not identified. It might also be mistakenly ligated during a nephrectomy for

transplantation or other purposes. Hence, Kennealy et al emphasized the importance of reviewing venous phase imaging during the evaluation of a living donor pair for kidney transplantation as discovery of double IVC in the operating room could lead to an unexpectedly short vein and serious technical difficulties in certain recipients [4].

Our patient might be a candidate for renal transplant due to his underlying polycystic kidney disease which may be further complicated by renal failure and hemorrhage. At the moment, there is no specific intervention planned for him. However, if he has to undergo a retroperitoneal surgery or any intervention to his vena cava in the future, the physician has to take note of his condition and be extra careful as poor planning can lead to a major disaster.

4. CONCLUSION

A high index of suspicion of double vena cava existence is important to avoid important diagnostic pitfalls and also for smooth preoperative surgical and interventional radiological planning.

CONSENT

All authors declare that written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images.

ETHICAL APPROVAL

Ethical approval is not required at our institution to publish an anonymous case report. Images were provided by the Department of Radiology and Diagnostic Imaging, Hospital Segamat, Johor, Malaysia, following permission from the Head of the Radiology Department.

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