Case study

Double Vena Cava In Haemorrhagic Polycystic Kidney Disease Patient

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ABSTRACT

Aims: We review the literature~~s~~ pertaining to the implications of having the rare anomaly of double vena cava.
Presentation of Case: A 20 year old ~~gentleman~~ male treated for polycystic kidney disease went for a computed tomography renal for microscopic haematuria and incidentally noted left sided vena cava separated from the right.
Discussion: Double inferior vena cava is a rare congenital venous anomaly. It usually does not cause any symptoms and incidentally seen during radiological imaging such as computed tomography for other medical conditions.
Conclusion: The discovery of this rare embryological feature depicts variety risks to patient during retroperitoneal surgery.

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 venous system for the development of IVC involved the posterior cardinal, subcardinal and supracardinal veins during the 5th week of gestation. The posterior cardinal veins remains 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 this process will end up with an anatomic ~~anomalies~~ anomaly of the IVC. [3] As in our case, the duplication of IVC likely results from persistence of the right and the left supracardinal veins.

We incidentally found a double IVC in a young man who was investigated for microscopic hematuria. We did literature review and discussed the implication of having this anomaly.

2. PRESENTATION OF CASE

A 20-year-old ~~gentleman~~ male was diagnosed with polycystic kidney disease following investigation for his sudden onset of difficulty to micturate. Physical examination was unremarkable. Laboratory examination 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 haemorrhagic cysts. The CT also showed a duplicated left sided inferior vena cava which extend 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)

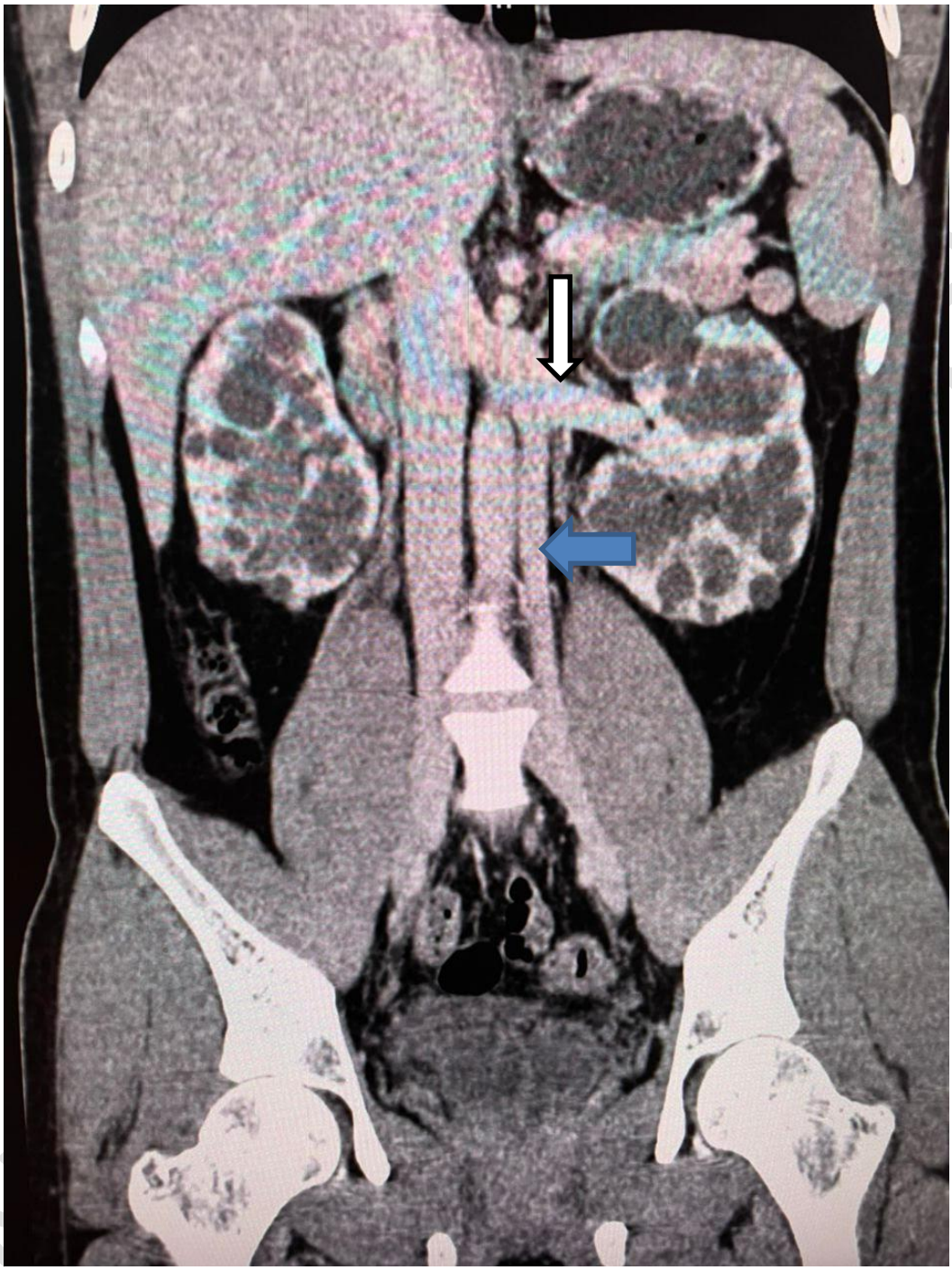

Figure 1. CT scan demonstrating left-sided vena cava (blue arrow) draining into the left renal vein (white arrow). The right-sided IVC can also be visualized separately from the left-sided vena cava.

Patient was prescribed with some urals and the symptoms eventually resolved. We set up a routine follow up for the patient as to assess the progress of the hemorrhagic polycystic

kidney disease which in the event of renal failure or extensive bleeding may require surgery. It is imperative for the operating surgeon to be aware of the rare venous anomaly as to avert any unfavorable outcomes throughout the operation.

3. DISCUSSION

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

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 disease have been reported in patients with a duplicated IVC, as noted in our literature search. Milani et al. reported that a patient with double IVC alone may not be specifically predisposed to development of acute thromboembolism as it could also be influenced by other routine pre-existing conditions, such as malignancy, contraception hormones, and genetic predisposition. [5] To date, there is no existing data to guide the anticoagulation treatment for thrombosis in patients with congenital anomaly. In attempting caval filtration, a different strategy is needed depending on the presentation of illness and there are few literatures reported. 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]

Radiologists should also be aware of the different types of anomalies of the IVC in order to avoid misinterpreting radiological imaging. There are reports that mistakenly diagnosed double vena cava 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 afforded these patients.

When performing abdominal surgery, the presence of double IVC needed to be acknowledged as it can lead to significant bleeding in an open abdominal aortic aneurysm surgery. It might also be mistakenly ligated during a nephrectomy for transplantation or other purposes. 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 hemorrhagic polycystic kidney disease. 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.

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