

Case Report

Rare case of circumaortic left renal vein with fenestration and nutcracker phenomenon

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ABSTRACT

Circumaortic left renal vein (CLRV) is a rare venous anomaly. It is usually clinically silent until discovered accidentally during an operation, imaging or an autopsy. Vascular fenestrations are most commonly seen in the arterial system or cerebral vessels and rarely in the venous system. Nutcracker phenomenon is usually associated with CLRV anomaly. We have described a case of a 31-year-old female who presented to the Department of Nephrology with CLRV, along with retro aortic renal vein fenestration and anterior nutcracker phenomenon. Since this variation is rarely diagnosed, it is important to have knowledge about such anomalies to rule it out as a differential diagnosis of haematuria. This information will also prevent iatrogenic injuries during surgical procedures.

Keywords: Circumaortic left renal vein, Fenestration, Nutcracker phenomenon, Hematuria

INTRODUCTION

Anatomically, the renal veins connect the kidney to the inferior vena cava (IVC).¹ They are usually singular for each kidney and are usually located to the aorta anteriorly.² However, due to its embryological development, the anatomy of the renal veins can be variable.³ The prevalence of 3 main positions of renal vein has been reported: retro-aortic renal vein (3%), circum-aortic renal vein (3.5%) and multiple renal veins (16.7%). Most often, it remains clinically silent until it is discovered accidentally during an operation, imaging or an autopsy.⁴

Circumaortic left renal vein is an anomaly of the left renal vein where an accessory renal vein passes posterior to the aorta, while the normal renal vein passes anterior to the aorta.³ In most cases of circum-aortic left renal vein, compression of the pre-aortic left renal vein between the superior mesenteric artery and the aorta occur, which is termed as the anterior nutcracker phenomenon.⁵ Anterior

nutcracker phenomenon causes venous hypertension and is followed by urological problems, such as hematuria, varicocele, ureteropelvic junction obstruction, and pelvic congestion syndrome.⁴⁻⁶ In addition, left renal vein fenestrations are rarely seen, since vascular fenestrations are mostly found in the arterial system and cerebral vessels.⁷ In this article, we present a case of a circum-aortic left renal vein with fenestration, along with anterior nutcracker phenomenon, which leads to gross hematuria as a complication of the anomaly.

The article aims to highlight a rare case of an anomaly of the left renal vein (Circumaortic left renal vein with nutcracker phenomenon) and the significance of being familiar with such congenital anomalies.

CASE REPORT

On 26th of February, a 31-year-old female presented for a consultation in the department of nephrology with

complaints of gross hematuria. She noticed dark brown urine five times within the past year. She first consulted a urologist, who initially diagnosed her with acute cystitis, after finding increased erythrocytes in her urine analysis. The urologist assumed that it could be due to an infection she had and prescribed antibiotic therapy. However, the treatment did not improve her symptom. Hence, she visited the department of nephrology for further tests and treatment. The patient denied having a medical history of tuberculosis or viral hepatitis and confirmed having no recent travel history or contact with infected individuals. The patient has no history of allergies, transfusions or symptoms indicative of having intestinal infections in the past 10 days.

On physical examination, she did not have symptoms of peritonitis; her abdomen was soft with no palpable masses. The patient demonstrated painless urination with sufficient diuresis and an absence of peripheral edema. A kidney ureter bladder (KUB) ultrasound of the patient was performed on February 27th, which showed right kidney measuring 106×50 mm and left kidney measuring 107×47 mm, with normal bilateral renal parenchymal echotexture and preserved mobility. No calculi or dilation of ureters were observed. Volume of the bladder was 440 cc.

A computer electrocardiogram (ECG) was performed on 26th February, which revealed ectopic atrial rhythm, a normal electrical axis of the heart, and early ventricular repolarization syndrome, with a heart rate of 56 beats per minute. Laboratory investigations showed normal general blood and urine test results. Biochemical blood tests indicated a marginal increase in LDL level (3 mmol/l), an increase in creatinine level (107 mcml/l) and a mild decreased eGFR of 61.4 ml/min/1.73 m² (Calculated using CKD-EPI equation).

Finally, a computed tomography (CT) scan of the abdominal organs was performed with intravenous contrast on 5th March, which revealed an abnormal CLRv with aorto-mesenteric compression by the anterior renal vein (preaortic renal vein), along with a fenestrated posterior renal vein (retro aortic renal vein). The right kidney's blood supply consists of one artery, 5 mm in cross-section, arising from the aorta at the level of L2, and one vein at the level of L2. The left kidney's blood supply consists of one artery 5mm in cross-section, arising from the aorta at the level of L2, supplied by two veins. The pre-aortic vein (4 mm in diameter) from the left kidney is at the level of the upper edge of L2, bends around and in front of aorta, under the superior mesenteric artery. It is compressed at this level and drains into the IVC with the left ovarian vein (3 mm). The retroaortic vein, measuring 29 mm in length, has a fenestration behind the aorta at the level of the upper edge of L3. The upper part of fenestration measures 5×8 mm, and the lower part measures 8×18 mm, which then merge into one vein measuring 4×18 mm that drains into the IVC at the level of L3.



Figure 1: CT angiogram of 31-year-old female with CLRv with fenestration associated with nutcracker phenomenon. The image shows 2 left renal veins. A pre-aortic renal vein (blue arrow) and a retro-aortic renal vein (red arrow) leaving the left renal hilum.



Figure 2: CT angiogram of 31-year-old female with CLRv with fenestration of retroaortic left renal vein. The image shows the retroaortic left renal vein (red arrow) passing between aorta and lumbar vertebrae, which then continues to split into a fenestration (blue arrow).

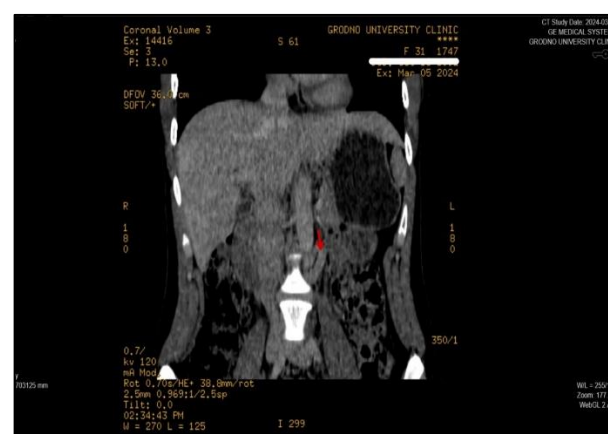


Figure 3: CT scan of 31-year-old female with CLRv with fenestration of retroaortic left renal vein. The image shows fenestration in retroaortic left renal vein (red arrow).

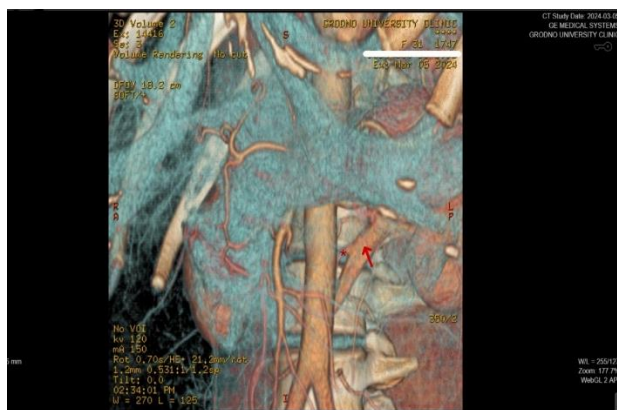


Figure 4: Three-dimensional reconstruction of the CT angiography of 31-year-old female with CLRV with fenestration associated with nutcracker phenomenon. The image shows the retroaortic left renal vein (red arrow) with fenestration (red *).

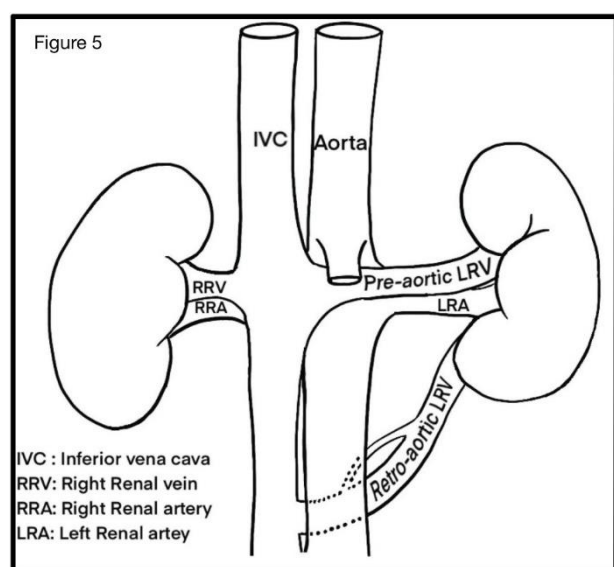


Figure 5: Schematic illustration of the present case of 31-year-old female with CLRV (Type II) with retroaortic vein fenestration.

Since all laboratory examinations revealed collectively normal findings with spontaneous resolution of hematuria, the patient was advised to be under the observation of a physician, to comply with the correct water-salt regimen, and to avoid environments with cold temperatures and strenuous physical activity. An ultrasound every six months and consulting a nephrologist in case of recurrence of hematuria were also recommended. In the event of multiple recurrences of hematuria and any further complications related to the nutcracker phenomenon, it was planned to refer the patient to the surgical department for surgical interventions.

Surgical interventions should be considered only when symptoms of this phenomenon are severe or persistent and when patients fail to respond to conservative treatment

after several months.⁵ The interventions should aim to decompress the left renal vein and decrease the left renal vein hypertension, as well as address pelvic venous reflux.⁵ Such interventions include left renal vein transposition, gonadocaval transposition, nephropexy, left renal vein to inferior vena cava bypass and nephrectomy as a last resort.^{5,8,9} Hence, this patient was advised to be observed by the physician, and to undergo ultrasound once every 6 months.

DISCUSSION

To understand the development of the circumaortic left renal vein (CLR), it is important to consider the embryogenesis of the renal vein system.⁹ By 8 weeks of embryogenesis, the bilateral supracardinal veins and bilateral subcardinal veins, which drain the upper and lower halves of the body respectively, join together to form a circumaortic venous ring ("renal collar").^{2,4,9} As development continues, the dorsal aspect of this renal collar obliterates, and the ventral aspect is left to mature, leading to the complete development of the normal renal vein.^{2,9} In the present case, both the dorsal and ventral aspects of the renal collar persisted and formed a circumaortic vein. Very limited case reports are available regarding the abnormality of renal veins.

According to the classification made by Natsis et al in their study, there are 3 different forms of CLRV. In type I, a single left renal vein branches into a preaortic and a retroaortic vein, which drains separately into IVC.¹⁰ In type II, there are 2 left renal veins from the left kidney draining into the IVC (one is preaortic and the other is retroaortic).¹⁰ Type III is a complex form of CLRV. In this form, 2 renal veins that are anastomosed in a common trunk drain from the kidney, which then separates again into 1 preaortic and 1 retro-aortic branch, and drains into the IVC.¹⁰ In this form, there may be anastomoses between the preaortic and retroaortic veins (being multiple or not) or multiple preaortic or retroaortic renal veins without anastomoses, forming a venous plexus around the aorta.¹⁰ The present case falls into type II of this classification of CLRV along with fenestration on the retroaortic branch.

Although CLRV is clinically silent in most cases, possible symptoms include hematuria, left flank pain, inguinal pain and pelvic congestion syndrome.¹¹ In this patient, hematuria was the only symptom that presented clinically. Compression of the preaortic renal vein between aorta and superior mesenteric artery (Anterior Nutcracker Phenomenon) may have caused gross hematuria. Left retroaortic renal vein fenestration may also be one of the reasons why the patient had hematuria. Although venous fenestrations are rare, it is possible to observe such fenestrations in the venous system.⁷ Hence, it is important to rule out anatomical abnormalities of renal vasculature as possible causes of hematuria. The diagnostic methods for detecting left renal vein anomalies are renal venography, color Doppler ultrasonography, CT angiography (CTA), and magnetic resonance angiography

(MRA).¹² Since renal venography is an invasive and expensive method, most preferred methods are non-invasive techniques such as color Doppler ultrasound, CTA and MRA.¹³ Computed tomography is a frequently used diagnostic method for imaging the kidney and retroperitoneal region.⁴ Studies show that 1.5-3.5% of abdominal CT imaging detects renal vein anomalies.⁴ Multidetector computed tomography (MDCT) is a diagnostic method that can identify changes in revascularization more easily.¹³ With advances in CT technology, MDCT has replaced conventional angiography and venography in most clinical conditions.¹⁴ It is a reliable, easily applicable, and noninvasive tool for demonstrating abdominal organs and vascular structures.¹⁴

It is important to understand the anatomy and congenital defects of the renal vein to prepare for retroperitoneal surgeries such as radical nephrectomy, living donor kidney transplantation, and procedures for abdominal aortic aneurysm repair.¹² During the surgical procedures, accidental injury to these venous anomalies can lead to severe haemorrhage which further complicates the surgery leading to nephrectomy, and sometimes even death.¹² In addition, knowing about the patient having this anomaly may also provide safety guidelines for endovascular procedures, such as abdominal venous sampling, renal venography, spermatic vein embolization, and placement of an IVC filter.¹²

CONCLUSION

Having knowledge of the anatomic structures of the renovascular system and venous variations in patients is important to increase detectability and to prevent possible iatrogenic injury during surgical procedures and interventions. It is essential for surgeons to keep in mind the possibility of accidental injury to a missed venous anomaly, in instances of persistent, inexplicable bleeding. If fatal complications are to be avoided, appreciation of such anatomical variants, careful reading of the preoperative CT scan when available, a high suspicion of such incidents, and safe operative techniques are all vital. It is also crucial to consider venous anomalies as a differential diagnosis for symptoms such as unexplained hematuria.

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