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RARE CASE OF CIRCUMAORTIC LEFT RENAL VEIN WITH FENESTRATION AND NUTCRACKER SYNDROME

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Introduction. Anatomically, the renal veins connect the kidney to the inferior vena cava (IVC). They are usually singular to each kidney and are usually located to the aorta anteriorly. However, due to its embryological development, the anatomy of the renal veins can be variable [1, 2]. Various positioning of renal vein has been reported and circumaortic left renal vein is one of them. Circumaortic left renal vein **is** an anomaly of left renal vein when an accessory left renal vein passes posterior to the aorta, along with normal renal vein passing anterior to the aorta. Prevelance of main anatomical variants of renal vessels vary in different scientific literatures. According to the previous studies, the prevalnce of circumaortic left renal vein has been reported to be between the range of 1% to over 15%. Hence, occurrence of circumaortic renal vein can be assumed to be a rare venous abnormality. Mostly, it remains clinically silent until it gets discovered accidentally during an operation or imaging.In most cases of circumaortic left renal vein, compression of the the pre-aortic left renal vein between the superior mesenteric artery and the aorta occur, which is termed as the nutcracker phenomenon [3, 4]. In addition, left renal vein fenestrations are seen rarely since vascular fenestrations are mostly seen in the arterial system and cerebral vessels.In this article, we present to you, a case of circumaortic left renal vein with inferior renal vein fenestration, along with anterior nutcracker syndrome which lead to macroscopic hematuria as a complication of the anomaly.

Aim of the study. The article aims to highlight on a rare case of an anomaly of renal veins called circumaortic left renal vein and importance of being familiar with such congenital anomalies.

Materials and methods. On 26th of February, a 31 year old female presented herself for a consultation to the Department of Nephrology with complaints of

macroscopic hematuria. She noticed her urine being in dark brown colour 5 times within the past 1 year. She first consulted to 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 gave antibiotic therapy. However, the treatment did not improve her symptom. Hence, she visited the department of Nephrology to do further tests and get treatment. Patient denied having a medical history of tuberculosis or viral hepatitis and confirmed having no recent travel history or contact with infectious individuals. Patient has no history of allergies, transfusions and symptoms indicative of having intestinal infections in the past 10 days. Patient demonstrated normal, painless urine output with sufficient diuresis and absence of edema.

Electrocardiogram (ECG) was done on 26th February which revealed Ectopic atrial rhythm, Normal electrical axis of the heart (vector) position, with heart rate of 56 beats per minute. Laboratory investigations revealed a marginal increase in level of lymphocytes (40.7%) from the complete blood count test. Urine analysis according to Nechiporenko did not show any leukocytes and erythrocytes . Biochemical blood tests showed increase in creatinine level (107 mcmol/l). GFR = 60 ml/min/1,73м^2. Kidney Ureter Bladder (KUB) ultrasound of the patient was done on 27th February, which showed right kidney measuring 106x50mm and left kidney with the size of 107x47mm, normal bilateral renal parenchymal echotexture with preserved mobility. No calculi or dilation of ureters were seen. Volume of the bladder was 440 ml.

Finally, a Computer Tomography (CT) scan of abdominal organs was performed with intravenous contrast on 6th March, which revealed circumaortic left renal vein with aorto-mesenteric compression by the superior renal vein (Nutcracker phenomenon), with fenestrated inferior renal vein. Right kidney one artery, 5mm in cross section from the aorta at the level of L2, one vein at the level of L2 and left kidney one artery 5mm in cross section from the aorta at the level of L2, supplied with 2 veins. Superior vein (4 mm diameter) from left kidney is at the level of upper edge of L2, bends around and in front of aorta, under the superior mesenteric artery. It is compressed at this level and follows into IVC with left ovarian vein (3mm). Inferior vein passes (29mm in length) with fenestrations from left and behind the aorta, at the level of upper edge of L3 (3x18mm) and is compressed to 3mm (diameter). Lower part of fenestration at L3 (8x18mm) and is compressed to 4mm (diameter). Both the superior and inferior veins merge into one vein with 4x18mm and flows into IVC at the level of L3.

Patient was advised to be under observation of a physician, to be in compliance with correct water-salt regime, to avoid environments with cold temperature and strenuous physical activity. Patient was advised to do ultrasound once in every 6 months. Patient was then discharged on 6th March 2024.

Results and discussion. It is important to understand the anatomy and congenital defects of the renal vein to prepare for retroperitoneal surgeries. During the surgical procedures, accidental injury to these venous anomalies can lead to the patient having severe hemorrhage, nephrectomy, and sometimes even death. In

addition, knowing the information about the patient having this anomaly may also provide safety guidelines for endovascular procedures.

To understand the development of this variation, it is important to consider the embryogenesis of the renal vein system. By 8 weeks of embryogenesis, the bilateral supracardinal veins and bilateral subcardinal veins, which drain the upper and lower half of the body respectively , join together to form circumaortic venous ring ("renal collar"). As development continues, the dorsal aspect of this renal collar obliterates and the ventral aspect is left to mature and leads to complete development of normal renal vein. In the present case, both the dorsal and ventral aspect of the renal collar persisted, and formed circumaortic left renal vein.

Although circumaortic left renal vein is clinically silent in most cases, possible symptoms include hematuria, left flank pain,inguinal pain and pelvic congestion syndrome. In this patient, hematuria was the only symptom that presented clinically. Compression of the superior renal vein between aorta and superior mesenteric artery (Nutcracker Syndrome) may have caused macroscopic hematuria. 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. Hence why, this patient was advised to be observed by the physician, and to do ultrasound once every 6 months. Left inferior renal vein fenestration may also be one of the reasons for why the patient has hematuria. Although venous fenestrations are rare, it is possible to see such fenestrations in venous system.

Conclusion. Having the anatomic knowledge of renovascular structures and venous variations of patients is important, to increase its detectability and to prevent possible iatrogenic injury during surgical procedures and interventions. Furthermore, it is important for surgeons to keep in mind the possibility of an accidental injury to a missed venous anomaly, at instances of persistent, "unexplained" bleeding. If fatal complications are to be avoided, appreciation of such anatomical variants, a high index of suspicion, careful reading of the preoperative CT scan when available, and safe operative technique are all vital.

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