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CASE REPORT

Posterior Nutcracker Syndrome with Retroaortic Left Renal Vein: A Rare Disease that Still Being Overlooked

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ABSTRACT

Posterior nutcracker syndrome or retroaortic left renal vein (RARV) is a rare disease and is still being overlooked. In our case, A 10-year-old boy with irrelevant medical history and physical exam presented with gross hematuria. The initial evaluation that included urine, blood, and ultrasonography were normal. When hematuria responded to conservative treatment, patient discharged home. When he presented again after 3 months with the same complaint, CT with contrast was requested that reported with no abnormalities. When patient came for the 3rd time with the same complaint, the previous CT angiography was reviewed and the diagnosis of RARV was overlooked in the CT report. When the diagnosis was established, conservative management was adopted because patient had stable Hb level with no other symptoms. Posterior nutcracker syndrome should be put in the differential diagnosis list in patients with recurrent and unexplained hematuria when the routine work-up shows no abnormalities.

Keywords:Nutcracker syndrome; retroaortic left renal vein; hematuria

INTRODUCTION

Unexplained gross hematuria with an unremarkable medical history and normal physical exam is a rare case scenario in clinical practice. When the routine initial laboratory tests and imaging reveal no abnormalities, the case becomes more challenging to urologists and an extensive work-up may be justified. Vascular anomalies among the rare possible causes that may cause unexplained gross hematuria. Nutcracker syndrome (NCS) is a vascular anomaly to the left renal vein (LRV)[1]. Although anterior NCS was reported frequently in the literature which describes compression of the LRV between the superior mesenteric artery and the aorta[2], posterior NCS is rare and infrequently reported in the literature, just a few cases reported. We present a case of posterior NCS, or retroaortic left renal vein (RLRV) that was overlooked and diagnosed lately.

CASE HISTORY

A 10-year-old boy presented with gross hematuria. Patient denied any recent history of trauma, fall from hight, or urologic interventions. No abnormalities were detected on clinical examination.At the time of presentation, renal function was normal and Hb was 12.8. The initial ultrasound (KUB) was normal. The patient was seen by a nephrologist and the immunologic profile was normal. Urine did not show any dysmorphic RBCs. On the 3rd day and with bed rest and plenty of fluids, hematuria improved. Urine became clear and the patient was discharged for routine follow up in the clinic. He came back after 3 months with recurrent gross hematuria, so CT KUB with contrast was ordered. It revealed a normal anatomy with no stones, masses or any visible abnormalities. The boy and his family were assured and he was discharged home. He presented again 5 months later with the same clinical picture, the CT was reviewed

and a retroaortic left renal vein (RLRV) type I confirmed the diagnosis of posterior Nut-cracker syndrome (PNCS) (see **Figure 1a, b**). Hb did not change and ranged between 12-13gm/dl.

A year after the last presentation, the patient has had no gross, but microscopic hematuria on routine urine analysis. No active intervention was done for him and follow up only was adopted because the hematuria was mild, intermittent and the serial Hb was unchanged.

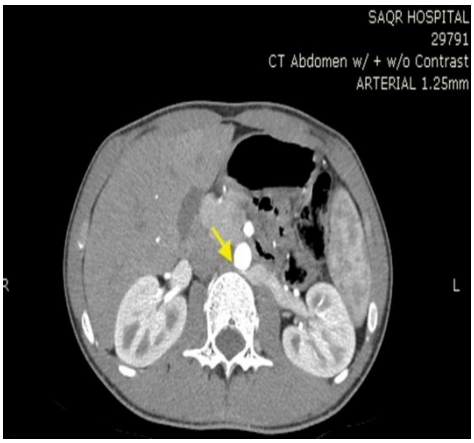

Figure 1a

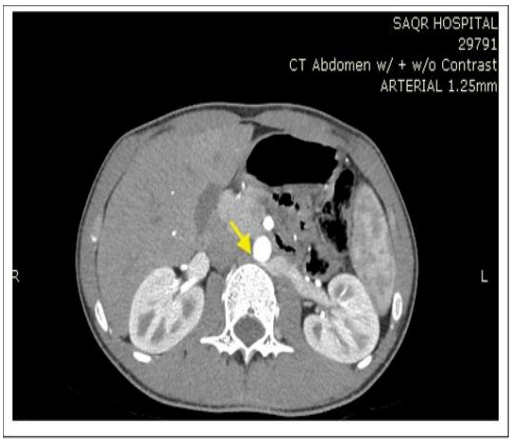

Figure 1b

DISCUSSION

Vascular anomalies should be suspected in younger patients with gross spontaneous hematuria after other common causes have been ruled out. A vascular renal malformation, like A-V malformation, pseudoaneurysm, A-V fistula usually induce severe gross hematuria and a significant drop in Hb. Our patient had a nearly stable serial Hb that was within the normal range and the radiologist initially reported CT angiography with no renal or vascular pathology.

Nutcracker phenomena / syndromes (NCP / NCS) are among the rare causes of unexplained hematuria. Both terms are used interchangeably, but NC phenomenon refers to a radiologic finding only. When it is associated with symptoms, it's given the term NC syndrome. It is also known as left renal vein (LRV) entrapment which is characterized by impeded outflow from the left renal vein into the inferior vena cava (IVC) due to extrinsic LRV compression[1].

The extrinsic compression on the left renal vein could occur anteriorly or posteriorly. Because the anterior compression is more common (80% of cases reported) and occurs when the LRV is compressed between the superior mesenteric artery and the abdominal aorta, NCS traditionally refers to this phenomenon[2]. Our radiologist could preclude this possibility on the initial report. However, the LRV could be compressed posteriorly between the aorta and the spine, which is known as retroaortic left renal vein (RLRV)-which is less common and could be overlooked.

Type one is the most common type of RLRV (6/9= 67%), and our case belongs to this type. Whatever the type, the compression impairs the venous return from the LRV with subsequent distal engorgement.RLRV is generally rare and could be completely asymptomatic and could be discovered incidentally only in a CT for an unrelated disease. Karman et al respectively evaluated 1,856 CT, and 68 (3.6%) were found with RLRV, 35% of them were found with no symptoms [3].

Symptoms may include: flank pain, hematuria either microscopic or gross, left-sided lower abdominal pain, or pelvic congestion and dyspareunia in females**.** In men, it may lead to varicocele which is quite common and reported in 10% of those patients. Our patient did not have palpable varicocele by physical exam.Our patient had gross hematuria which is the most frequent symptom that was reported in many published series. Hematuria is due to a rupture of the thin-wall varices into the collecting system, secondary to elevated venous pressure[4]. Spontaneous resolution of NCP has been described in children, sometimes after several years of persistence.

RLRV is less common and less frequently reported in the literature. In a series that included 9 cases by Nam et al [5], the mean age was 46y, with symptoms of hematuria or flank pain. 6 of them (67%) were of type I. Although gross hematuria improvs in the vast majority of patients, microscopic hematuria may persist for longer time in those patients. Our patient's age was younger than the mean age of the Nam et al published series, and has had no recurrent gross hematuria, but still had persistent microscopic hematuria. The treatment of RLRV varies based on patients' symptoms, with most of the published series recommending no intervention especially in patients like our case. Our patient had infrequent gross hematuria (2-3 times/ year), had no flank pain, with a stable Hb, so we adopted conservative management as it carries more than a 75% resolution rate as reported.

Conclusion

Posterior nutcracker syndrome or retroaortic left renal vein is a very rare disease and is still being overlooked. It should be put in the differential diagnosis list in patients with unexplained hematuria when the routine work-up shows no abnormalities. CT angiography diagnoses the disease and conservative management can be adopted if gross hematuria is infrequent, and the patient has a stable Hb level with no other symptoms.

Legends to figures:

Figure 1 (**a:** Arterial and **b:** Portal phases): Contrast enhanced computed tomography revealed a type I retro-left renal vein (RLRV) between abdominal aorta and vertebra showing focal narrowing of its diameter (arrows) compatible with posterior nutcracker syndrome

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