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Case Report

Macroscopic Hematuria in Adolescents - Two Cases of Nutcracker Syndrome

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Abstract

Macroscopic hematuria is a common cause of referral to the Pediatric Emergency Department and can be due to a variety of etiologies, which can be identified through detailed clinical history and physical examination. Nonetheless, rare causes may go unnoticed with the most used diagnostic exams. Nutcracker syndrome describes left renal vein compression between the superior mesenteric artery and the abdominal aorta or between the abdominal aorta and vertebral column. The majority of the patients are asymptomatic, and when symptomatic, clinical manifestations include variable combinations of hematuria, proteinuria, left flank or abdominal pain. Diagnosis is usually made after the exclusion of other causes responsible for the symptoms, and it is confirmed on imaging results. Treatment remains controversial, with pediatric patients being typically managed conservatively due to spontaneous resolution in most cases. With these two cases, the authors aim to bring awareness to this entity so it can be easily identified and avoid unnecessary diagnostic workup.

Keywords: Hematuria; Nutcracker; Nephrology; Proteinuria; Kidney

Abbreviations

AA: Abdominal Aorta; ASLO: Anti-streptolysin O; CRP: C-reactive Protein; DUS: Doppler Ultrasound; ED: Emergency Department; HPF: High Power Field; LRV: Left Renal Vein; MRI: Magnetic Resonance Imaging; NCS: Nutcracker Syndrome; RBC: Red Blood Cells

Introduction

Macroscopic hematuria is a common cause of referral to the Pediatric Emergency Department (ED) and can be due to a variety of etiologies such as urinary tract infections, nephrolithiasis, and glomerulonephritis [1]. Detailed clinical history and physical examination are essential to identify the source of the hematuria, but rare causes may go unnoticed with the most used diagnostic workup.

Nutcracker syndrome (NCS) occurs when the left renal vein (LRV) becomes compressed between the superior mesenteric artery (SMA) and the abdominal aorta (AA) or between the AA and vertebral column [2]. Clinical manifestations include variable combinations of hematuria (micro or macroscopic), proteinuria, left flank or abdominal pain. Since it may also be asymptomatic, it can be an easily missed diagnosis.

We present two cases of adolescents with NCS who presented initially with macroscopic hematuria.

Case Reports

Case 1

A 13-year-old previously healthy male presented to the ED with gross hematuria since that day. He also had fever and sore throat for 48 hours, being medicated with amoxicillin and clavulanic acid the day before. He presented with normal blood pressure, mild palpebral edema, and tonsillar exudates. Laboratory tests revealed a sedimentation erythrocyte rate of 19mm, a C-reactive protein (CRP) of 75.6 mg/L, and 77 red blood cells (RBC) per high power field (HPF) on urinalysis. Complete blood count, urea, creatinine, electrolytes, complement component (C3, C4), and anti-streptolysin O (ASLO) were normal. He was diagnosed with post-infectious glomerulone-

phritis and discharged with the indication to stop physical activity. He was periodically evaluated until complete resolution of the symptoms but two weeks later he returned to the ED due to hematuria reappearance, associated with vomits and abdominal pain. He had reinitiated physical activity, having trained 2 times that week. Physical examination was normal, except for epigastric pain to palpation. Urinalysis revealed 47 RBC/HPF with 3% of dysmorphic erythrocytes, and blood tests were normal. Doppler ultrasound (DUS) revealed alterations suggestive of NCS. Posterior magnetic resonance imaging (MRI) described a reduction in the angle between the AA and the SMA (38%) and a loss of fat tissue around the route of LRV, confirming the diagnosis. Currently, he is 18 years old and maintains periodic evaluations in Nephrology consultations and intermittent episodes of microscopic hematuria and orthostatic proteinuria, sometimes related to physical exercise.

Case 2

A 16-year-old male with no significant past medical history presented to the ED with persistent painless gross hematuria for 2 days. He denied any episodes of trauma, vigorous exercise, recent infections, or weight loss. On physical examination, he had a blood pressure of 120/84 mmHg and mild lower abdominal pain to deep palpation. Urinalysis revealed hematuria (1066 RBC/HPF, some of them dysmorphic). Blood tests, including complete blood count, urea, creatinine, electrolytes, albumin, total proteins, and coagulation tests were normal, except for a mildly elevated CRP (39.4 mg/L). ASLO, immunoglobulin A, M, and G, complement component, antinuclear, and anti-double-stranded DNA antibodies were negative. DUS revealed compression of the LRV posterior to the SMA, with an elevated peak flow velocity at this level, compatible with NCS. According to the clinical status, laboratory tests, and imaging findings, a conservative approach was considered appropriate for this patient. He was discharged and advised to avoid vigorous physical activity. Subsequent MRI revealed a diminished angle between the SMA and the AA and compression of the LRV by the SMA, confirming the diagnosis. He is periodically observed in Nephrology consultations, remaining clinically stable, with no medication, and no episodes of gross hematuria.

Conclusion

NCS is a rare cause of hematuria in children, probably underdiagnosed. It describes LRV compression between the SMA and the AA (anterior NCS) or between the AA and vertebral column (posterior NCS), causing a rise in LRV pressure and collateral vein development [3]. The exact prevalence of NCS is unknown and it can be diagnosed at any age, but appears to be more common in young adults (second or third decade) [4,5]. Although it has been reported to be more prevalent in females, recent studies have shown an equal prevalence in both genders [6].

This entity can go easily unnoticed since the majority of the patients are asymptomatic and diagnosed incidentally through imaging exams. When symptomatic, a wide range of manifestations may occur such as abdominal and left flank pain, orthostatic proteinuria, and hematuria, which appears to be the most common clinical feature [7]. It can also present with pelvic congestion syndrome (lower abdominal pain, dysmenorrhea, dyspareunia, and dysuria), and gonadal varices (varicocele or ovarian vein syndrome), due to the development of collateral circulation secondary to LRV hypertension [5]. These symptoms are usually aggravated by physical activity and stress [4].

Due to the variety of non-specific symptoms and the absence of clinical diagnostic criteria, NCS is difficult to diagnose and requires a high index of suspicion. Diagnosis is usually made after the exclusion of other causes responsible for the symptoms, and it is confirmed on imaging results [8]. DUS is an easy, fast and inexpensive exam, and is recommended as the first diagnostic test, with a sensitivity of 69-90% and a specificity of 89-100% [6]. Since it is not diagnostic, due to the highly variable index and range of values, axial imaging is usually necessary. Computed tomography and MRI are capable of demonstrating reduction of the aortomesenteric angle (less than 45º) and collateral circulation in the renal hilum [8]. Retrograde venography remains the gold standard for diagnosis, but it is not commonly used in the pediatric age due to radiation exposure and its invasive nature. It confirms the anatomical alterations and measures the pressure gradient across the entrapped area, making a definitive diagnosis if this gradient exceeds 3 mmHg [9].

Treatment remains controversial and should be based on the patient's age and the severity of the symptoms. Patients under the age of 18 with mild symptoms are typically managed conservatively since NCS appears to resolve spontaneously in 75% of young patients with physical development or weight gain [2,4,7]. This seems to be explained by the growth of connective tissue and fat close to the LRV, together with the formation of collateral veins, which alleviate compression and venous hypertension. This approach may be maintained for 24 months. Analgesics can be used for pain control, angiotensin-converting enzyme inhibitors for relieving orthostatic proteinuria, and low-dose aspirin for improving renal perfusion [6,8]. If after this period the patient has persistent symptoms that interfere with his day-to-day activities, surgery becomes the next treatment [7]. The surgical approach can be the first choice

of treatment in cases of severe symptoms, such as severe pain, persistent macroscopic hematuria, and progressive renal function impairment [3]. Endovascular stenting of the renal veins is usually the initial procedure of choice and may be accompanied by embolization of the gonadal vein. Other procedures, such as nephropexy, renocaval reimplantation or auto-transplantation, transposition of the LRV or SMA, renocaval bypass, or even nephrectomy may be needed [8]. In both of our cases, given the mild symptoms with no interference in the quality of life of the patients, we decided on a conservative approach.

In conclusion, NCS is a rare and probably underdiagnosed condition that should be considered in the differential diagnosis of hematuria. Both of these adolescents presented in our ED initially with gross hematuria and mild abdominal pain. In the second case, an accurate and fast diagnosis was possible, due to a high index of suspicion and a request for specific imaging tests.

With these two cases, the authors aim to bring awareness to this entity so it can be easily identified and avoid unnecessary diagnostic workup.

Conflict of Interest

No financial interest or any conflict of interest exists.

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