Nutcracker Syndrome: a Rare Cause of Hematuria

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ABSTRACT

In pediatrics, hematuria usually responds to benign etiologies. A rare cause is nutcracker syndrome, defined as compression of the left renal vein between the aorta and the superior mesenteric artery, resulting in elevated pressure in the left renal vein with the development of collateral veins and varicose dilatation. The prevalence of this syndrome is unknown. When symptomatic, it causes hematuria, proteinuria, and chronic pelvic pain. At puberty, the rapid growth and development of the vertebral bodies can produce a narrowing of the angle between the aorta and the superior mesenteric artery.
We describe the case of a teenage girl diagnosed with multiple metastatic mucoepithelial carcinoma of bone who presented anemia-producing hematuria. The diagnosis of nutcracker syndrome was arrived at with consideration of the therapeutic options. Finally, with shoe enhancement for scoliosis correction, hematuria was significantly lessened.

Key words: hematuria, nutcracker syndrome.

INTRODUCTION

Hematuria is a very frequent symptom in medical consultations of all specialties, as a routine finding or as a symptom reported in the clinic. Its etiologies in pediatrics are diverse but are frequently benign and transitory, except in adolescence, when they usually require further study.

Nutcracker syndrome is one of the less frequent causes of hematuria. It can be defined as a compression of the left renal vein between the aorta and the superior mesenteric artery, resulting in elevated pressure in the left renal vein and hematuria due to the development of collateral veins and varicose dilatations[1-3].

We describe the case of an adolescent girl diagnosed with metastatic carcinoma of the dorsal spine who presents profuse and anemia-inducing hematuria. She was diagnosed with nutcracker syndrome with an unusual presentation, and we discussed conservative treatment versus vascular stenting.

CLINICAL CASE

An adolescent girl was diagnosed with myoepithelial carcinoma of the distal tibia and left calcaneus, metastatic in lung, bone, skin, and lymph nodes, diagnosed at the age of 6 years.

She received two rounds of chemotherapy without oncologic response and continued with metronomic modality (vinorelbine and cyclophosphamide). It consists of administering chemotherapy at regular intervals, for long periods, and in low doses, which highlights its ease of administration and lower toxicity, with satisfactory antineoplastic potential.With this treatment, the primary lesion and pulmonary metastatic lesion remained stable. Two and a half years after this chemotherapy regimen, there was bone progression with dorsal 6 (D6) and dorsal 12 (D12) vertebral wedging. Oral sirolimus was added to his previous treatment, and she started a follow-up with orthopedics and traumatology, spinal unit, and kinesiology. Orthopedic enhancements were made for everyday footwear.

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With this new therapeutic scheme, she remained in her province of origin, Jujuy, with scheduled multidisciplinary check-ups in Buenos Aires. She maintained her schooling, attended classes regularly, and participated in recreational activities with his peers and family.

Three years later, when she was 13 years old, she consulted due to macroscopic hematuria of ten days of evolution shown in all micturitions, without dysuria or other associated symptoms. She had had her last menstrual period that same month.

On physical examination, she was in good general condition with clinical stability. Her anthropometric data showed a weight of 37.3 kg (Pc 3), height of 137.5 cm (Pc 10), P/T 94%, and body mass index (BMI) of 19.9. The gynecological examination ruled out the genital origin of the bleeding.She also had scoliosis secondary to vertebral metastases, for which she wore a 2 cm shoe enhancement, and there was an increase in deformity associated with a limb discrepancy, also accentuated by a right tibial metastatic metaphyseal lesion.

Initial evaluation included complete urine, complete blood count, creatinine, urea, urine culture, and abdominal and renal ultrasound.

We found red blood cells and proteinuria in the urine, a drop in hemoglobin value (9.4 g/dL for previous values of 12.2 g/dL), and normal urea and creatinine levels. The ultrasonography did not show any alterations.

The search for dysmorphic red blood cells in urine revealed negative results. We performed 24-hour urine with proteinuria (5.43 g/24 hours-137 mg/kg/day) and significant calciuria (2 g/24 hours). With these findings, no arterial hypertension, normal urea, creatinine, and albumin, and in the absence of dysmorphic red blood cells, we ruled out the possibility of hematuria of glomerular origin. Calciuria was associated with her extensive tumor bone involvement.

Except for anemia, the rest of the hemogram, the coagulogram, and the fibrinogen were within normal parameters.

In the abdominal tomography, both kidneys were of a size consistent with age, with an adequate concentration of the contrast material. There was no enlargement of the urinary tract.

Given the likelihood that the hematuria was secondary to the use of cyclophosphamide, the chemotherapy was discontinued.

We ruled out infections such as adenovirus and urinary BK virus.

The patient persisted with daily hematuria and sustained anemia with the periodic requirement of red blood cell transfusions.

A cystoscopy was performed, which showed a bladder with obstructed visualization due to hematuria, a bladder wall undamaged, and an orthotopic ureteral meatus with no evidence of bleeding. Angiotomography showed left renal vein with a change of caliber at the level of the aortomesenteric compass, compass angle 46 degrees, distance from the aorta to the superior mesenteric artery 5.5 mm, and scoliosis of left convexity with inferior dorsal vertebra with central crushing and signs of vertebral disc degeneration (Figs. 1 and 2).

With a diagnosis of nutcracker syndrome, we reviewed the therapeutic options. We considered the magnitude of the symptoms, the high transfusion requirement, the interference in the quality of life, and, on the other hand, the oncologic status of an incurable but stable disease, which did not generate specific symptoms for the time being. The alternatives evaluated were:

- Conservative treatment, nutritional plan with weight gain associated with orthopedic shoe enhancement.
- Invasive surgical treatment: renal autotransplantation.
- Minimally invasive surgical treatment: insertion of a vascular endoprosthesis.

Due to the girl's current situation, her positive quality of life before the intercurrent, her oncologic stability with the therapy received, and her progression-free survival, together with the need to control the bleeding as soon as possible, it was decided in a multidisciplinary agreement to offer a minimally invasive procedure that would achieve permeabilization of the left renal venous compression. We suggested endovascular stent placement.

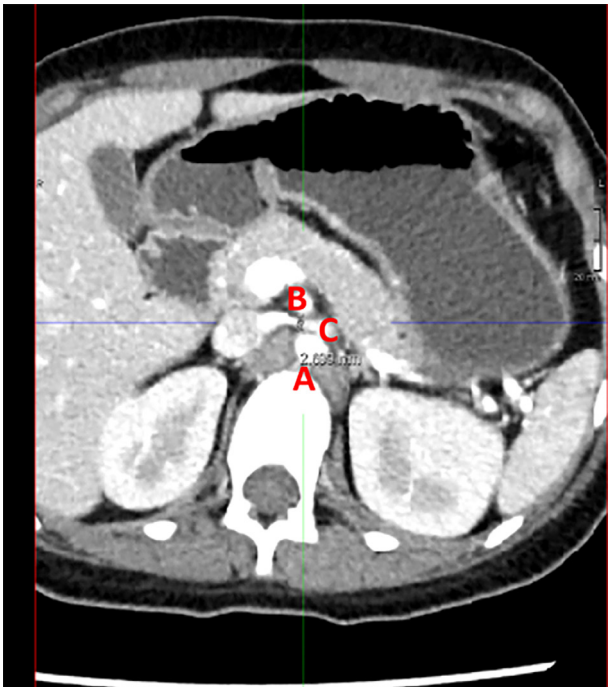

Figure 1. Abdominal angiotomography. Axial section. The mouth of the left renal vein can be seen in the inferior vena cava; its passage is narrowed by the compass formed by the aorta (A) and the superior mesenteric artery (B). At renal hilum level, there is proximal venous dilatation (C)..

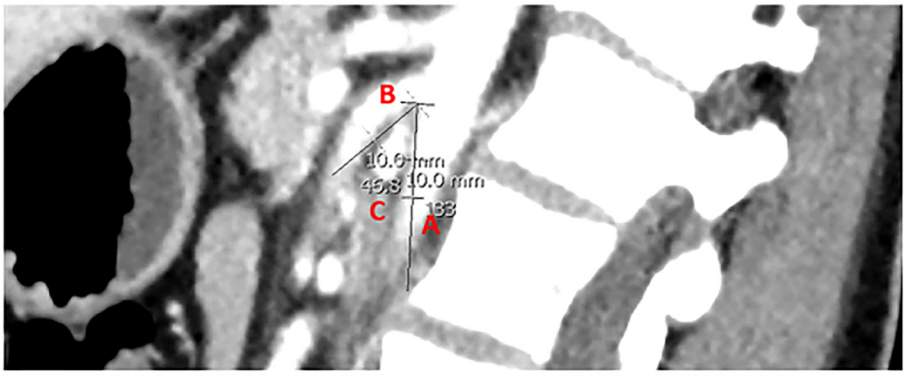

Figure 2. Abdominal angiotomography. A sagittal section at L2-L3 level. Aortomesenteric compass of 46 degrees. Note the intimate relationship of the left renal vein with the arteries that form the compass at this level.

Before this procedure, the Traumatology and Orthopedics Service corrected the scoliosis enhancement.

Immediately after wearing the correct footwear, the patient presented progressive improvement of the bleeding until macroscopic resolution.

She continued with periodic check-ups and resumed her oncologic medication. At one year of follow-up, although she presented intermittent bleeding, hemoglobin values remained stable. The present study was conducted following the guidelines established by the modified Helsinki Declaration.

DISCUSSION

The causes of hematuria in children under oncologic treatment are diverse, and initially, the study should focus on general causes.

The most frequent etiologies are the first to investigate. However, when the initial findings do not explain it, especially when the symptom is progressive, it is crucial to be insistent in identifying a cause.

Since this is an immunocompromised host, we looked for infrequent infections such as the BK virus and urinary adenovirus.

Hemorrhagic cystitis due to cyclophosphamide is described in association with higher doses than those used in the metronomic scheme. In our case, when using it for a prolonged duration and not for a posterior-type complication that could associate this case with metastatic spinal involvement.

At puberty, the rapid growth and development of the vertebral bodies can produce a narrowing of the aortomesenteric angle that predisposes to this syndrome [10,11]. The same happens when lumbar hyperlordosis develops or if significant weight loss occurs due to decreased retroperitoneal adipose tissue.

Published data encourage a conservative treatment approach in patients younger than 18 years, as growing individuals may experience resolution of symptoms from increased fibrous tissue at the superior mesenteric artery origin[5]. In addition, weight gain leading to an increase in retroperitoneal adipose tissue has shown a reduction in compression[12].

The modification of the orthopedic enhancement adjusted to the pondostatural growth seems to have attenuated the anatomical compression. Although the literature describes spontaneous symptomatic resolution in most cases[13], the direct association between this intervention and the decrease in bleeding is striking.

We emphasize the importance of decision-making when opting for non-conservative treatment; venous stents, as analyzed in this case, seemed to offer a feasible solution. Intervention is only indicated in severe lesions when disabling symptoms do not respond to conservative management 9. This device is associated with relatively frequent complications such as migration and requires platelet antiplatelet therapy or anticoagulation[14].

When weighing the costs and benefits of each therapeutic alternative, it is essential to consider each specific case to offer the best treatment that suits the particularities of each individual.

CONCLUSION

The diagnosis of nutcracker syndrome often delays due to the lack of consensus on the diagnostic criteria to be applied and the number of other more frequent etiologies. It is essential to know this entity to achieve a timely diagnosis.

Conflict of interest: the authors declare no conflict of interest.

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