Case report of gross hematuria in the nutcracker syndrome resolved by renocaval reimplantation

Prikaz izlečenja bolesnika sa obilnom hematurijom kod sindroma nutcracker primenom renokavalne reimplantacije

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Abstract

Introduction. Nutcracker syndrome is defined as a set of signs and symptoms secondary to compression of the left renal vein (LRV) in the acute anatomic angle between the aorta and its superior mesenteric branch. Case report. A 38-year-old woman with asymptomatic and “idiopathic” gross hematuria came to the Clinic for Vascular and Endovascular Surgery, Clinical Center of Serbia in Belgrade. Hematuria was documented by cystoscopy and was found to be unilateral, located to the left urethral orifice. The contrast-enhanced multidetector computed tomography (MDCT) scan showed a stenotic LRV due to the extrinsic compression in the angle formed by the ventral aorta and superior mesenteric artery (MSA), with a jet of contrast through the lumen. Considering the negative investigations for more common causes of hematuria, its incapacitating nature, and above mentioned imaging findings suggestive of the nutcracker syndrome, an indication for the open surgical correction of the LRV entrapment was established. The patient underwent reimplantation of the LRV into the more distal inferior vena cava (IVC), to relocate it out of the constrictive aortomesenteric space. Intraoperative findings were notable for blood flow turbulence in the LRV and hypertrophy of its tributaries, which were ligated. We presented the first published case in the Serbian literature on nutcracker syndrome with hematuria resolved by renocaval reimplantation. Conclusion. This case report demonstrates that renocaval reimplantation, as the open surgery technique, could be the adequate method for resolving gross hematuria in patients with nutcracker syndrome.

Key words: renal nutcracker syndrome; diagnosis; vascular surgical procedures; renal veins; replantation; treatment outcome; serbia.
Introduction

The nutcracker syndrome is defined as a set of signs and symptoms secondary to compression of the left renal vein (LRV) in the acute anatomic angle between the aorta and its superior mesenteric branch. Extrinsic compression of the left kidney main venous drainage leads to local venous hypertension, with the development of collaterals, pelvic varicoses, and gonadal tributary reflux, resulting in hematuria, left flank pain and symptoms of pelvic congestion.

We presented the first published case in the Serbian literature of the nutcracker syndrome with hematuria resolved by renocaval reimplantation.

Case Report

A 38-year-old women of two was referred to our clinic because of the chronic “idiopathic” gross hematuria, which occurred intermittently for the last ten years, but recently became persistent and demanded blood transfusions for the correction of anemia, prompting more extensive diagnostic investigation. Besides hematuria, the patient complained of no other symptoms, and her past medical, surgical and family history was noncontributory. Except for anemia (laboratory findings: Er 3.2 × 10¹²/L; Hb 104 g/L; Ht 0,32l/L; MCV 80 fl; MCH26 pg; MCHC 300 g/L) and asthenic constitution, physical examination was unremarkable, with soft and no tender abdomen, without any signs of peripheral vascular lesions, and novulvar varicosities. The patient underwent repetitive urine cultures and excretory urography with negative results. Hematuria was documented by cystoscopy and was found to be unilateral, located to the left urethral orifice. Urine cytology was negative for malignancy. After we excluded urological, nephrology, gynecological and oncological reasons for hematuria, contrast-enhanced multidetector computed tomography (MDCT) revealed a hyperacute aortomesenteric angle with impingement of the left renal vein, findings that were corroborated by the contrast inferior cava venography with selective catheterization of the left renal vein. Dilated and tortuous adrenal and gonadal venous tributaries were also noted.

The contrast-enhanced MDCT scan showed a stenotic LRV due to the extrinsic compression in the angle formed by the ventral aorta and superior mesenteric artery (MSA), with a jet of contrast through the lumen (Figures 1 and 2). Color Doppler ultrasonography and selective retrograde left renal venography showed dilatation of the distal part of the LRV and gonadal vein with pelvic varicosities (Figure 3).

Considering the negative investigations for more common causes of hematuria, its incapacitating nature, and abovementioned imaging findings suggestive of the nutcracker syndrome, an indication for the open surgical correction of

Fig. 1 – Compression of the left renal vein between the aorta and the superior mesenteric artery, multidetector computed tomography (MDCT) longitudinal section

Fig. 2 – Compression of the left renal vein between the aorta and the superior mesenteric artery, multidetector computed tomography (MDCT) transverse section

Fig. 3 – Selective retrograde left renal venography showing dilatation of the distal part of the left renal vein (LRV) with pelvic varicosities

the LRV entrapment was established. Under general anesthesia, using midline laparotomy approach, the patient underwent reimplantation of the LRV into the more distal inferior vena cava (IVC), to relocate it out of the constrictive aortomesenteric space. Intraoperative ultrasound findings were notable for blood flow turbulence in the LRV and hypertrophy of its tributaries, which were ligated.

Discussion

The true incidence of the symptomatic nutcracker syndrome in a general population is unknown. It seems that this syndrome is most common in women in the third and fourth decades of life. In their pathologic study, Macmahon and Latorraca elucidated an association between the nutcracker syndrome and hematuria.

According to the current international experience, there are several ways of solving this problem – by nephrectomy, nephropexy, renocaval reimplantation of the LRV or LRV stenting. We opted for the reno caval reimplantation as the most commonly used method, and because of our experience with open surgical procedures and lack of endovascular material at that moment. The LRV was excised from the entry site at the IVC and reanastomosed with the IVC 4 cm below the original anatomic site. There were no intraoperative or early postoperative complications (< 30 days). During the postoperative follow-up of one and a half year macrohematuria despaired. According to the previously published reports, in most similarly treated cases of the nutcracker syndrome hematuria resolved in up to 5 months of follow-up.

Conclusion

This case report demonstrates that renocaval reimplantation, as the open surgery technique, could be the adequate method for resolving gross hematuria in patients with nutcracker syndrome.

REFERENCES


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