Case Report

Nutcracker Syndrome: An Overlooked Cause of Hematuria

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Nutcracker syndrome is caused by compression of the left renal vein between the aorta and the superior mesenteric artery, where it courses in the fork formed at the bifurcation of these arteries. The phenomenon results in left renal venous hypertension, which leads to left renal vein and left gonadal vein varices and unilateral hematuria. The main presenting symptom is hematuria, with or without left flank pain. The disorder is easily missed by routine diagnostic methods. Its incidence is likely underestimated. We report on a 25-year-old woman who experienced intermittent gross hematuria and left flank pain. The diagnosis of nutcracker syndrome was missed initially. Abdominal computed tomography, angiography, venography, and magnetic resonance angiography, which were later performed, showed that the left renal vein was compressed between the aorta and the superior mesenteric artery. The pressure gradient between the left renal vein and the inferior vena cava was 6.8 cm H2O. A diagnosis of nutcracker syndrome was established. She refused surgery and was lost to follow-up. The diagnosis and treatment of nutcracker syndrome are discussed. Magnetic resonance angiography is a safe and reliable tool for diagnosing this disorder. (Chang Gung Med J 2002;25:700-5)

Key words: nutcracker syndrome, left renal venous hypertension, hematuria.

A 25-year-old woman was admitted to our hospital with intermittent gross hematuria and intermittent left flank pain in December 1994. There was no history of upper respiratory infection prior to episodes of hematuria before the first admission. Her medical history was otherwise unremarkable. She weighed 43 kg and was 156 cm tall. Her blood pressure was 110/70 mmHg, pulse rate 80/min, respiratory rate 16/min, and body temperature 36.5°C. Her physical examination was normal. There was no tenderness at the costovertebral angle or lower leg edema. At that time, urinalysis showed numerous

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red blood cells, protein of 100 mg/dl, and 3-5 white blood cells per high-power field. Blood chemistry tests showed BUN of 16 mg/dl and creatinine of 0.7 mg/dl. Hemoglobin was 12.0 gm/dl, and the white blood cell count was 7900/mm³. Antinuclear antibody was negative; C3 was 105 mg/dl (normal 71.87-122.03); and C4 was 13.5 mg/dl (normal 9.92-29.96 mg/dl). IgA was 174 mg (normal 71.20-434.12 mg/dl); IgG was 1570 mg/dl (normal 835-1716 mg/dl); and IgM was 265 mg/dl (normal 24.29-198.69 mg/dl). Prothrombin time, partial thromboplastin time, and bleeding time were normal. Urine cytology and cultures for tuberculosis bacilli were negative. Renal ultrasonography showed normal renal size and outline with no anatomical defect. The length of the left kidney was 10.6 cm, and that of the right was 10.9 cm. The result of intravenous pyelography was normal. Cystoscopy showed bleeding from the bilateral ureteral orifices. Retrograde pyelography revealed filling defects in the left renal pelvis and upper ureter, which were possibly caused by blood clots. On light microscopy, the histology of the renal biopsy was normal. Immunofluorescence showed trace depositions of IgA, IgM, and C1q in the mesangial areas. Hence, IgA nephropathy was suspected. The patient was discharged without medication. However, intermittent gross hematuria persisted and some episodes were preceded by upper respiratory tract infection. So, she was again admitted in September 1998. Renal ultrasonography showed dilatation of the left renal vein in the hilar

![Fig. 1](image1.png) **Fig. 1** CT scan showing dilatation of the distal left renal vein (LRV) with narrowing between the superior mesenteric artery (SMA) and the aorta (Ao) (arrow).

![Fig. 2](image2.png) **Fig. 2** (A) Simultaneous angiography and venography showing narrowing of the proximal left renal vein (1) with filling defects due to compression by the superior mesenteric artery (2) and aorta (3). (C) Venography showing reflux of the left renal venous flow into the left ovarian vein (arrow).
area. The length of the left kidney was 11.3 cm and that of the right 10.3 cm. Blood biochemistry and complete blood count were normal. Antistreptolysin O antibody was 155 IU/ml (normal < 200 IU/ml), and IgA was 187.0 mg/dl. Abdominal computed tomography (CT) scan revealed that the left renal vein was compressed between the aorta and the superior mesenteric artery (Fig. 1). Angiography and venography also showed filling defects in the left renal vein because of compression by the aorta and the left renal vein (Fig. 2A, B). In addition, there were prominent periretoral and peripelvic venous collaterals and reversal of the left renal venous blood flow into the left ovarian vein (Fig. 2C). The pressure gradient between the left renal vein and inferior vena cava was 6.8 cm H₂O. Magnetic resonance angiography (MRA) showed compression of the left mid-renal vein between the superior mesenteric artery and the aorta. The distal third of the left renal vein was dilated, whereas the proximal third was relatively small in caliber (Fig. 3A, B). Cystoscopy was again performed, and it showed bleeding from the left ureteral orifice. Therefore, nutcracker syndrome was our impression. Surgery was suggested. However, the patient refused, and was lost to follow-up thereafter.

**DISCUSSION**

Compression of the left renal vein was first described in 1950. In 1972, De Schepper described compression of the left renal vein between the aorta and the superior mesenteric artery as nutcracker syndrome. Nutcracker syndrome occurs most frequently in young women. It has been associated with unilateral hematuria, gonadal vein syndrome, and varicocele. Various degrees of proteinuria are present. Unilateral hematuria is due to abnormal communication between the submucosal venous plexus and the calyceal system presumably induced by renal venous hypertension. The gonadal vein syndrome is characterized by abdominal and flank pain exacerbated by sitting, standing, or walking. Zerhouni et al. reported the nutcracker phenomenon in 3 patients investigated for varicocele.

The pathophysiology of nutcracker syndrome is not well known. It was proposed that posterior renal ptosis with stretching of the left renal vein may be a factor. In recent studies, abnormal branching of the superior mesenteric artery from the aorta was identified as its cause.

Nutcracker syndrome cannot be diagnosed with routine diagnostic methods. Therefore, it is easily misdiagnosed or undiagnosed. Intravenous pyelog-
raphy may show notching by varicosities, cystoscopy may reveal bleeding from the left ureteral orifice, and a CT scan may show compression of the left renal vein between the aorta and superior mesenteric artery and the coexistence of abnormal venous collaterals.\(^6\) Angiographic CT, magnetic resonance imaging (MRI), and MRA can also be used as diagnostic tools.\(^8\) In this case, we demonstrated the findings on angiography, CT, and MRA. MRA is less invasive than angiography and can be used initially for detecting this rare cause of hematuria. Ultrasonography and Doppler ultrasonography are alternative noninvasive methods for diagnosis.\(^1\) Kim et al. reported that the ratio of the diameter of the left renal vein between the hilar portion and the aortomesenteric portion is greater than 5 in patients with nutcracker syndrome. In addition, the ratio of the peak velocity of the left renal vein between the aortomesenteric portion and the hilar portion is also greater than 5 in patients with this syndrome.\(^2\) Takebayashi et al. reported that the sensitivity and specificity of color Doppler sonography for diagnosing the nutcracker syndrome were 78% and 100%, respectively.\(^4\) Venography combined with venous pressure measurement is recognized as the procedure of choice for diagnosis.\(^6\) Venography may show narrowing of the renal vein where it crosses the aorta beneath the superior mesenteric artery, dilatation of the distal left renal vein, and opacification of tributaries of the left renal vein (the gonadal, ascending lumbar, adrenal, ureteral, and capsular veins). The pressure gradient may range from 4.9 to 14.0 cm H\(_2\)O in patients with nutcracker syndrome, whereas normal values range from 1.3 to 10 cm H\(_2\)O.\(^6\) In our patient, the pressure gradient between the left renal vein and inferior vena cava was 6.8 cm H\(_2\)O.

In our patient, immunofluorescence on renal biopsy showed trace depositions of IgA, IgM, and C1q in the mesangial areas, and the result of the first cystoscopy showed bleeding from the bilateral ureteral orifices. Therefore, the possibility of nutcracker syndrome combined with IgA nephropathy cannot be excluded. Ozono et al. reported 2 patients with nutcracker syndrome associated with IgA nephropathy.\(^3\)

Conservative treatment has been suggested for mild hematuria.\(^9\) Surgery is indicated for severe persistent or recurrent gross hematuria causing anemia and bothersome abdominal or flank pain.\(^5\) Medial nephropexy with excision of the renal varicosities, left renal vein bypass, and transposition of the left renal vein have been described.\(^2\) Autotransplantation is an alternative treatment that provides the kidney with better protection from ischemia.\(^1\) An intravascular stent, which can be placed with minimal invasiveness, offered physiological relief in a recently reported case.\(^10\)

In conclusion, nutcracker syndrome is a rare cause of hematuria and is easily missed by routine diagnostic methods. Hence, patients with unknown causes of unilateral hematuria and flank pain should undergo further studies such as ultrasonography, CT, MRI, MRA, angiography, or venography to clarify this possibility. MRA is a safe and reliable tool for diagnosing this disorder.

REFERENCES

胡桃銹症候群：一個被忽視造成血尿的原因

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胡桃銹症候群是因爲左腎靜脈在行經主動脈和上腔靜脈時，被主動脈和上腸腺膜動脈壓迫所造成。此現象造成左腎靜脈高壓，進而造成左腎靜脈和左側性腺靜脈曲張及單側血尿。主要的症狀爲血尿和左側腰痛。由於此疾病不能被常規的方法診斷，它的發生率常被低估。

我們報告一位25歲女性，因血尿及左側腰痛而求診。胡桃銹症候群剛開始並沒有被正確診斷。之後藉由腹部電腦斷層掃描，血管攝影，靜脈攝影及核磁共振血管攝影而顯現左腎靜脈被主動脈和上腸腺膜動脈壓迫。左腎靜脈和下腔靜脈的壓力差為6.8公分水柱。因此診斷出胡桃銹症候群。此病患拒絕接受手術治療，我們討論如何診斷和治療此疾病。核磁共振血管攝影是一種診斷胡桃銹症候群的安全且可靠的工具。(民康醫誌 2002,25:700-5)

關鍵字：胡桃銹症候群，左腎靜脈高壓，血尿。

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