Nutcracker Syndrome Diagnosed with 3-Dimensional Computed Tomography Angiography

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We report a case of nutcracker syndrome diagnosed with 3-dimensional computed tomography angiography (3-D CTA). Nutcracker syndrome had been confirmed by conventional venography until recent years. Nowadays, with the development of imaging techniques, color Doppler sonogram and 3-D CTA are replacing venography for the diagnosis of nutcracker syndrome. The patient, a 20-year-old male, had abrupt gross hematuria and left abdominal pain 6 months previously and intermittent microscopic hematuria thereafter. Including renal biopsy, the results of conventional hematuria study showed no abnormalities. 3-D CTA showed left renal vein compression between the abdominal aorta and superior mesenteric artery and collateral veins. The angle and distance between the superior mesenteric artery and aorta at the level of the left renal vein were 35° and 3.0 mm, respectively. We diagnosed nutcracker syndrome and later confirmed the diagnosis with venography. **(Korean J Urol 2009;50:711-713)**

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Nutcracker syndrome was discovered and named by De Schepper[1] in 1972 and occurs when the left renal vein is pressed and occluded by entrapment between the abdominal aorta and superior mesenteric artery (SMA). There are common cases in which symptoms are not observed, but sudden macroscopic hematuria and left abdominal pain may be observed. In particular, varicocele and ovarian vein syndrome may occur in men and women, respectively. Symptoms vary with age, renal venous pressure, and collateral vascular development. It is still not certain how high pressure of the renal vein can induce hematuria.

Nutcracker syndrome has been diagnosed by performing venography and measuring the pressure difference between the left renal vein and the inferior vena cava, but debate exists on how to determine the cut-off value of pressure difference. More recently, computed tomography (CT) scanning, color Doppler sonography, and Doppler MRA have been substituted for invasive venography.[2] In particular, 3-dimensional computed tomography angiography 3 (3-D CTA3) has been widely used.[3]

For treatments, left renal vein transplantation and endovascular stent insertion are usually performed. Recently, noninvasive stent insertion has been chosen as the primary treatment by many doctors.[3] In special cases, SMA transplantation may be performed.

Here we report the usefulness of 3-D CTA in diagnosing nutcracker syndrome in a 20-year-old man who complained of hematuria.

CASE REPORT

A 20-year-old man visited our hospital in January 2008 for intermittent macroscopic hematuria. The patient had undergone hematuria tests, including abdominal sonography and CT, in another hospital in November 2007 as the result of sudden macroscopic hematuria and left abdominal pain, but the results of all tests were negative. No abnormalities were observed in the patient's history and physical examinations, and his blood pressure was 115/80 mmHg. Urinalysis showed many erythrocytes and few leukocytes. The results of a complete blood count, LFT, bleeding tendency, complements, and I g study

Fig. 1. Coronal CT scan: the distance between the superior mesenteric artery and the aorta at the level of the renal vein is only 3 mm.

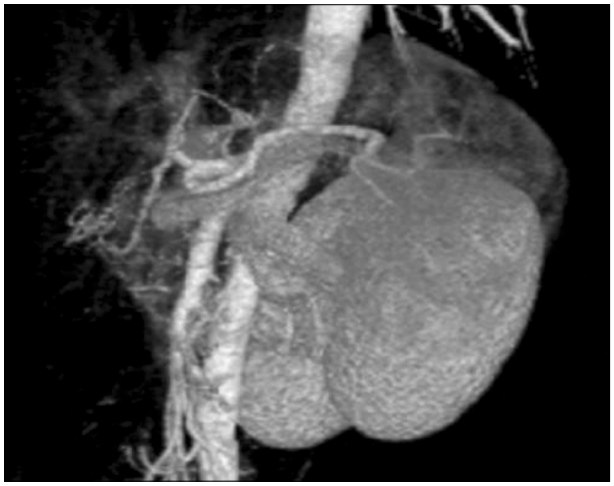

Fig. 2. 3-Dimensional computed tomography angiography (3-D CTA): the left renal vein is compressed by the aorta and the superior mesenteric artery and the angle between these 2 arteries is narrowed to 35°.

were not significant. IVP showed no abnormalities. No ureteral hematuria or mucosal bleeding were observed during cystoscopy. Renal biopsy under electron microscopy showed no abnormalities. During the CT performed in this hospital, findings suggestive of nutcracker syndrome were observed: the aorta was just 3.0 mm distant from the SMA at the level of the left renal vein (Fig. 1). 3-D CTA revealed that the left renal vein was pressed between the SMA and the abdominal aorta and also the outstanding collateral veins. The angle between the SMA and the abdominal aorta was just 35 degrees (Fig. 2). Nutcracker syndrome was confirmed by venography. Because the patient is scheduled to undergo treatment after his military service, he is currently being observed. Macroscopic hematuria has not yet been observed.

DISCUSSION

Nutcracker syndrome frequently occurs in schoolchildren, or in young and healthy adults, as shown in the present case. Nutcracker syndrome is assumed to occur very rarely, but in reality it may be more common if it is easier to diagnose.[4]

Etiologically, it is known that the SMA or its abnormal branch puts pressure on the left renal vein. Also, the syndrome has been known to occur in cases where the left renal vein is tensely pressed by the abdominal aorta due to the influence of nephroptosis, where the left renal vein originates from a point that is too high from the inferior vena cava and as a result the SMA puts pressure on the left renal vein, or where an abnormal branch of the renal artery puts pressure on the left renal vein.[5]

Symptoms of nutcracker syndrome are related to the high pressure of the left renal vein, such as unilateral hematuria, gonadal vein syndrome, and varicocele. It is presumed that pain and hematuria are connected with the high pressure of the left renal vein and the ureteropelvic varix. The high pressure of the left renal vein congests intrarenal vessels to rupture the weak veins of the calyceal fornix and the collecting system. In rare cases, anemia, proteinuria, and varicose veins occur.[6] In the present case, gonadal varix was not observed. In addition to macroscopic hematuria with sudden abdominal pain, painless gross hematuria was observed several times.

Urinalysis, IVP, urine cytology, cystoscopy, ultrasonography, and biopsy are usually performed to diagnose the cause of hematuria. Nevertheless, it is not easy to diagnose nutcracker syndrome. When hematuria occurs on the left side without other causes, nutcracker syndrome should be considered. The standard diagnostic method is to check left ureteral hematuria with a cystoscope and to measure the pressure difference between the left renal vein and the inferior vena cava with venography. There have been dissenting opinions, but ordinarily in nutcracker syndrome, the left renal vein and the inferior vena cava show a pressure difference of at least 4 mmHg.[2] Venography enables one to observe the distally dilated renal vein and collateral veins between the SMA and the abdominal aorta. However, because of its invasiveness and the occasional failure, it is not easy to perform venography to measure intravenous pressure.

Recently, with the advancement of imaging techniques, it has

become possible to diagnose nutcracker syndrome without performing venography and measuring intravenous pressure. 3-D CTA is noninvasive and shows 3-D images. Because it shows the abnormal pathological findings of the SMA as well as the pressure of the left renal vein more accurately than renal vein angiography, it is more useful for diagnosing nutcracker syndrome.[3,7] Additionally, it enables observation of the distally dilated renal vein and collateral veins. 3-D CTA shows clear images of the renal vein, the SMA, and the abdominal aorta and enables measurement of the angle and distance between the SMA and the abdominal aorta. As a result, nutcracker syndrome can be diagnosed without difficulty. Normally, the angle and the distance between the SMA and the abdominal aorta are 9010 degrees and 121.8 mm, respectively.[7] In nutcracker syndrome, the angle and the distance are 39.34.3 degrees and 3.10.2 mm, respectively. The present case was consistent with the report of Fu et al.[7] The angle and the distance were compatible enough to diagnose nutcracker syndrome.

Nutcracker syndrome should be treated differently according to severity. In cases in which specific symptoms are not observed or microscopic hematuria occurs, conservative treatment is suitable. In cases with severe hematuria or severe pain, aggressive treatment is necessary. Renal vessel reanastomosis, renal autotransplantation, nephropexy, or renalectomy may be performed according to the circumstances. However, reanastomosis is regarded as the chief treatment. Through reanastomosis, the left renal vein, originated from the inferior vena cava, is shifted downward by about 5 cm so that it cannot be pressed.[5] Lately, stent insertion has been performed because it is less invasive. It has been reported to have high efficacy and low complications, but there is controversy about thrombi, stent occlusion, and stent movement.[3] The Korean Urological Association has reported 3 cases of nutcracker syndrome since 1960, which were all diagnosed by venography. The treatments done were hypogastric aortic reanastomosis,[8] observation,[9] and stent insertion,[10] respectively.

It is presumed that 3-D CTA is reliable and independent enough to diagnose nutcracker syndrome, because it offers accurate 3-D images of the compression of the left renal vein and enables measurement of the angle and distance between the SMA and the abdominal aorta. 3-D CTA should be applied to more cases to accurately evaluate its efficacy in diagnosis.

REFERENCES

1. de Schepper A. “Nutcracker” phenomenon of the renal vein and venous pathology of the left kidney. J Belge Radiol 1972;55:507-11
2. Fitoz S, Yalcinkaya F. Compression of left inferior vena cava: a form of nutcracker syndrome. J Clin Ultrasound 2008; 36:101-4
3. Zhang HS, Li M, Jin W, San P, Xu P, Pan S. The left renal entrapment syndrome: diagnosis and treatment. Ann Vasc Surg 2007;21:198-203
4. Wendel RG, Crawford ED, Hehman KN. The “nutcracker” phenomenon: an unusual cause for renal varicosities with hematuria. J Urol 1980;123:761-3
5. Rudloff U, Holmes RJ, Prem JT, Faust GR, Moldwin R, Siegel D. Mesoaortic compression of the left renal vein (nutcracker syndrome): case reports and review of the literature. Ann Vasc Surg 2006;20:120-9
6. Little AF, Lavoipierre AM. Unusual clinical manifestations of the Nutcracker Syndrome. Australas Radiol 2002;46:197-200
7. Fu WJ, Hong BF, Gao JP, Xiao YY, Yang Y, Cai W, et al. Nutcracker phenomenon: a new diagnostic method of multislice computed tomography angiography. Int J Urol 2006;13: 870-3
8. Lee SE, Cho SW, Yang SC. A case of nutcracker syndrome surgically corrected by extraperitoneal flank approach. Korean J Urol 1996;37:1027-30
9. Cho SY, Shin MS, Rhee HW, Seo JM. A case of the Nutcracker syndrome. Korean J Urol 1999;40:120-1
10. Yim SH, Koh JS, Kim HW, Yang CH, Jung JH, Lee JY. Expandable metallic stent placement for Nutcracker syndrome. Korean J Urol 2004;45:390-2