Nutcracker syndrome (NCS) was first described by Grand in 1944 as compression of the left renal vein with the superior artery (SMA) between the abdominal aorta. Although there are limited number of clinical studies on NCS, the most common symptoms and signs are left side pain, hematuria and proteinuria due to venous hypertension of the left kidney. Dyspareunia, dysuria, dysmenorrhea, scrotal and varicose vein formation in the lower extremity, abdominal pain are other rare symptoms and signs. Rarely, NCS has been reported to cause arterial hypertension. We found NCS in two young patients who were investigated for secondary reasons of arterial hypertension, and we decided to present them in this study.

Keywords: Blood pressure measurement; hypertension; nutcracker syndrome.

Can Nutcracker Syndrome Cause Secondary Hypertension? Two Case Reports

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Abstract

Nutcracker syndrome (NCS) was first described by Grand in 1944 as compression of the left renal vein with the superior mesenteric artery (SMA) between the abdominal aorta. Although there are a limited number of clinical studies on NCS, the most common symptoms and signs are left side pain, hematuria and proteinuria due to venous hypertension of the left kidney. Dyspareunia, dysuria, dysmenorrhea, scrotal and varicose vein formation in the lower extremity, abdominal pain are other rare symptoms and signs. Rarely, NCS has been reported to cause arterial hypertension. We found NCS in two young patients who were investigated for secondary reasons of arterial hypertension, and we decided to present them in this study.

Keywords: Blood pressure measurement; hypertension; nutcracker syndrome.
Case Reports

Case 1 – The 26-year-old female patient, who was a nurse, stated that her hypertensive blood pressure was first detected at the age of 13, especially when she stood up for a long period of time and as she got tired, her blood pressure rose up to 160/100 mmHg and a feeling of badness and tachycardia came with it. On physical examination, blood pressure was 135/85 mmHg from both arms, and the pulse was 80/min. The patient, who planned pregnancy in the near future, inquired about whether there was a condition that could prevent pregnancy.

Case 2 – A 16-year-old male patient was admitted to our clinic complaining of a headache. He claims that he has been suffering from headaches when he got tired ever since he was a child. Blood pressure measured from both upper extremities and both arms was 180/110 mmHg, and the pulse was 100/min.

No other features were found in the background and family history of both cases. There was no herbal medicine, licorice, continuous drug use, diarrhea and flushing attacks in their cases. In both cases, the blood pressures measured from the lower extremities were high and their pulses were clear. There was no additional pathological finding on their physical examinations.

Routine biochemical and hematological examinations, thyroid function tests, sedimentation, CRP, C3, C4 and venous blood gas were normal in both cases. There was no reproduction in urine cultures. ARB was negative in the urine. ANA, anti-dsDNA, catecholamine metabolites that were examined twice in urine, 5-HIAA, serum chromogranin A, inpatient plasma aldosterone concentration/plasma renin activity rate, dexamethasone suppression test result, electrocardiography, telegraphy, echocardiography were normal. Proteinuria was negative and 15-16 erythrocytes were detected in spot urine tests. Grade-1 hypertensive changes were detected in Case-1, and these were considered normal in Case-2. In the outpatient 24-hour blood pressure follow-up, it was found that the patients had hypertensive attacks that occurred during the daytime with long periods of standing up. Abdominal USG showed normal kidney sizes and normal parenchymal thicknesses. Renal artery Doppler USG was reported as normal for Case-1 and as an accessory renal artery/circumstantial renal artery for Case-2. For the cases where no microalbumin was detected in their urines, the amount of microalbumin and protein was examined standing and lying down in their 24 hour-urine.

Urinalysis and PAC/PRA results of the cases are shown in Table 1. Cerebral MR examination of case-2 is normal. In CT angiography, the left renal vein was squeezed between

| Table 1. Urine analysis and PAC/PRA levels of Case-1 and Case-2 |
|-----------------|-----|------|-------|-----|
|                | Case-1  | Case-1  | Case-2  | Case-2  |
|                | (outpatient) | (inpatient) | (outpatient) | (inpatient) |
| Albumin excretion rate (mg/g creatinine, spot urine) | 10.58 | - | 5 | - |
| Microalbumin (mg/day) | 0.27 | - | 5.33 | |
| Protein (g/day) | 0.350 | 0.200 | 0.300 | 0.07 |
| Urine sediment | 15-16 erythrocyte | - | 7-8 erythrocyte | - |
| Plasma aldosterone concentration (ng/dl) | 13 | - | 9 | |
| Plasma renin activity (ng/ml/hour) | 0.92 | 0.75 | - | 12 |
the abdominal aorta and SMA in both cases, and the left renal vein was dilated. Furthermore, it was detected that in case-1, the polar artery that emerged from the right main renal artery and entered the upper pole parenchyma of the right kidney, and in case-2, the bilateral double renal artery was detected (Figs. 1, 2). Metaiodobenzylguanidine (mIBG) scintigraphy was obtained by taking the endocrine into view for Case-1, who had hypertensive attacks and planned pregnancy. No pathological involvement was detected. No pathology was detected in the gynecological and urological evaluations of the cases. A control on blood pressure was achieved with alpha-methyldopa 500mg/day for Case-1, who was planning pregnancy and losartan 100mg/day for Case-2.

Discussion

NCS may cause various symptoms as a result of compression of the left renal vein between the aorta and SMA, often causes hematuria, proteinuria and left flank pain due to venous hypertension of the left kidney. More rarely, due to venous congestion syndrome, NCS may even cause tenderness on the left side of the vagina, dyspareunia, dysuria, dysmenorrhea, scrotal varicose vein formation, the formation of varicose veins in the gluteal and lower extremities, abdominal pain, gastrointestinal symptoms. There are also groups of patients who are symptomatic, detected by chance during the radiological examination. The diagnosis of NCS can be made by renal artery Doppler US, CT or MR angiography following a careful physical examination[1–4]. In both of our cases, a definitive diagnosis was made with a CT angiogram. There are very few studies showing that NCS may coexist with arterial hypertension. Peripheral PAC and PRA levels have been shown to be high in hypertensive Japanese women with young NCS. In patients who have increased the renocaval pressure gradient along with PRA levels by performing left renal venography, it was seen that these levels decreased after normotension by endovascular stenting [5]. On the other hand, there are doctors who think that NCS is not a cause of hypertension, and that coexistence with NCS is hypertension. The literature demonstrates its association with IgA nephropathy and familial Mediterranean fever (FMF) [6–8]. In both of our cases evaluated for secondary hypertension, we could not detect any pathology other than NCS that would explain hypertension. In our cases with microscopic hematuria and moderate orthostatic proteinuria, the inpatient PAC/PRA ratio and levels were not high. In outpatient blood pressure follow-ups, we found that the patients had elevated blood pressure, especially when they were standing up for a long period during the day. Therefore, we concluded that PAC and PRA may be rising, especially during hypertensive attacks and that arterial hypertension may be due to NCS. There are various opinions and methods concerning possible treatment. Open surgery, open or laparoscopic extravascular stent placement, endovascular stent placement or gonado-cabal bypass important methods for a possible treatment. Medical nephropexy is no longer used today. Left renal vein transposition, autotransplantation of the left kidney are other methods of treatment. SMA transposition is not recommended for its potential complications [1, 3, 9]. For our cases, vascular surgical opinion was considered, and it was decided to follow up with medical treatment.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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Conflict of Interest: None declared.


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References


