Nutcracker phenomenon: An unusual presentation of acute aortic dissection

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ABSTRACT
We report a case of acute aortic dissection leading to compression of the left renal vein (LRV), thereby resulting in the nutcracker phenomenon. A 49-year-old previously healthy woman presented with intermittent gross hematuria and mild left flank pain of five days’ duration. Laboratory examinations were within normal limits, except for the elevated C-reactive protein. Cystoscopy revealed bleeding from the left ureteral orifice. Contrast-enhanced computed tomography demonstrated acute Type B aortic dissection and compression of LRV between the enlarged aorta and superior mesenteric artery with an associated dilatation of the left gonadal vein as a collateral circulation.

Key words: Aortic dissection, hematuria, nutcracker phenomenon

INTRODUCTION
The nutcracker phenomenon (NCP) is characterized by impeded outflow from the left renal vein (LRV) into the inferior vena cava due to extrinsic LRV compression. LRV compression results in LRV hypertension which can cause hematuria originating from the left collecting system. Most typical nutcracker morphological features imply compression of LRV between the aorta and superior mesenteric artery (SMA). LRV compression can be produced by several secondary causes. However, the association between acute aortic dissection and NCP has not been reported previously. Here we report a case of acute aortic dissection leading to LRV compression, thereby resulting in NCP.

CASE REPORT
A 49-year-old previously healthy woman with no history of trauma presented with intermittent gross hematuria and mild left flank pain of five days’ duration. The patient described that her intermittent pain was aggravated by postural change and it gradually waned. The patient had no apparent respiratory, gastrointestinal or voiding symptoms. Her past medical history including hypertension, atherosclerosis and congenital diseases was non-contributory. She was afebrile and her physical examination findings were almost normal. Her blood pressure was 182/66mmHg. Urinalysis demonstrated 20–29 red blood cells per high-power field, 10–19 hyaline casts per field of vision and a sterile culture. Urine cytology was negative for malignancy. Laboratory examinations were within normal limits, except for the elevated C-reactive protein levels of 4.64mg/dL (normal, <0.30mg/dL). Ultrasonography and excretory urography did not reveal any definite structural abnormality in the urinary tract.

When the patient visited our outpatient clinic the next day, urinalysis showed dark urine containing numerous red blood cells without dysmorphic changes. Cystoscopy revealed bleeding from the left ureteral orifice. Contrast-enhanced computed tomography (CT) demonstrated aortic dissection originating just distal to the left subclavian artery and continuing into the bilateral common iliac arteries with a re-entry site in the abdominal aorta, compatible with acute Type B aortic dissection. No apparent branch vessel ischemia was observed. The patient was transferred to the intensive care unit in order to provide medical treatment, and prompt attention was paid to stabilizing the patient by reducing blood pressure.

On the retrospective review of CT scans, the arterial phase
demonstrated the left renal artery originating from the true lumen of the aorta and no apparent renal ischemia [Figure 1a]. The venous phase showed compression of LRV between the enlarged aorta and SMA with distal dilatation [Figure 1b]. A prominent left gonadal vein, implicating formation of collateral circulation, was also observed. Three-dimensional CT imaging demonstrated the compressed LRV and development of collaterals [Figure 2]. After the possibility of more common renal conditions such as tumors, urolithiasis and other medical renal diseases was excluded, we made a final diagnosis of NCP due to acute aortic dissection, explaining the gross hematuria. Invasive venography to measure the pressure gradient between LRV and the inferior vena cava was not performed.

During the following month, her clinical symptoms subsided and urinalysis abnormalities normalized. However, one month after the diagnosis, CT findings of LRV compression and left gonadal vein dilatation remained unchanged.

DISCUSSION

Acute aortic dissection is one of the most dramatic cardiovascular emergencies. Although this condition is not an infrequent clinical entity, its outcome is frequently grave. Classically, aortic dissection presents as sudden, severe chest, back or abdominal pain characterized as ripping or tearing. Symptoms in patients with aortic dissection are more variable than those recognized previously, and classic findings are often absent.[1] About 10% of aortic dissections are painless and may present with symptoms secondary to the complications of dissection.

Renal ischemia, a major complication of aortic dissection, arises from extrinsic compression of the true lumen by the false channel or by an initial flap compressing the orifice of the renal artery.[2] Approximately half of the patients with renal infarction have hematuria. In our case, however, no elevation in the levels of lactate dehydrogenase, an indicative serum marker of renal infarction, was observed, and no apparent radiological findings of renal ischemia were found.

NCP should be considered during differential diagnosis in patients with unexpected hematuria originating from the left collecting system. Although the precise mechanism of hematuria due to this phenomenon remains unclear, LRV compression presumably results in LRV hypertension, leading to the development of collaterals with intrarenal and perirenal varicosities, which can cause hematuria if the thin-walled septum separating the veins from the collecting system ruptures.[3]

Although the precise etiological factors of NCP are unknown, several possible causes have been proposed. These include posterior renal ptosis and an abnormally high course of LRV. These anatomical variations result in stretching of LRV and poor venous drainage. An abnormal angle of the origin and course of the proximal part of SMA may be responsible for this phenomenon. LRV compression may be produced by secondary causes such as pancreatic neoplasms, para-aortic lymphadenopathy, retroperitoneal tumors, overarched testicular artery or strangulating fibrolymphatic tissue between SMA and the aorta.[4] Moreover, increase of the aortic diameter can lead to LRV compression, thereby resulting in NCP. This includes posterior NCP due to an abdominal aortic aneurysm reported by Puig et al.[5] In the present case, LRV anatomically got stretched across the enlarged aorta and pinched with the comparatively narrower SMA–aorta angle. To our knowledge, this is the first reported case of NCP due to acute aortic dissection.

LRV flow patterns associated with NCP depend on the degree and stage of NCP. Persistence of LRV hypertension probably causes the development of collateral veins. In patients with collateral veins, the presence of both a distended LRV and LRV hypertension indicates that

![Figure 1: (a) Arterial contrast-enhanced CT demonstrates acute aortic dissection, without renal ischemia. True (*) and false lumens, contrast opacified. (b) Venous CT demonstrates LRV compression between the enlarged aorta and SMA (arrow) with distal dilatation. Hepatic cysts are also shown.](image-url)
NCP is noncompensatory. Well-developed collateral veins dissipate the high pressure gradient and diminish the blood flow volume of LRV resulting in the absence of distended LRV and LRV hypertension. We believe that follow-up of the present case with acute-onset NCP is necessary to understand the natural history of NCP.

In summary, the clinical symptoms in patients with acute aortic dissection may be highly variable, mimicking more common conditions. In acute aortic dissection, increased aortic diameter may contribute to LRV compression, resulting in NCP.

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