Nutcracker syndrome as an incidental finding on computed tomography scan after blunt abdominal trauma

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RÉSUMÉ

Prérequis : Le nutcracker syndrome résulte de la compression de la veine rénale gauche entre l'aorte et l'artère mésentérique supérieure. Il peut rester asymptomatique pendant de longues périodes et est de découverte fortuite.
But : Rapporter un nouveau cas, diagnostiqué par examen radiologique après un traumatisme abdominal. Le diagnostic et le traitement du Nutcracker syndrome sont discutés.
Observation : Un homme de 32 ans, a été victime d'un accident de la circulation causant une fracture de la jambe et un traumatisme abdominal fermé. Dans les suites de la chirurgie orthopédique, il a présenté une hématurie isolée. La tomodensitométrie abdominale a montré une veine rénale gauche comprimée entre l'aorte et l'artère mésentérique supérieure. Le diagnostic de nutcracker syndrome a été retenu. La surveillance a été indiquée avec une excellente évolution.
Conclusion: Le nutcracker syndrome est une entité rare. Il peut être asymptomatique. Son diagnostic est le privilège de l'imagerie médicale. Son traitement est controversé et doit être discutée au cas par cas.

Mots-clés

Accidents, traumatisme, fracture, veine rénale, hématurie, nutcracker syndrome, CTscan, surveillance

SUMMARY

Background: The nutcracker syndrome refers to compression of the left renal vein between the aorta and the superior mesenteric artery. It may be asymptomatic for long period and constitute casual findings.
Aim: To report a new case diagnosed by radiological exploration after an abdominal trauma. The diagnosis and treatment of nutcracker syndrome are discussed.
Case: A 32-year-old man was a victim of traffic accident causing leg and abdominal trauma. Three days after orthopedic surgery, he presented an isolated gross hematuria. Abdominal computed tomography showed that the left renal vein was compressed between the aorta and the superior mesenteric artery. The diagnosis of nutcracker syndrome was established. Surveillance was indicated with excellent evolution.
Conclusion: The nutcracker syndrome is a rare entity. It may be asymptomatic. Its diagnosis is the privilege of medical imaging. Its treatment is controversial and should be discussed case by case.

Key-words

Accidents, traumatism, fracture, renal vein, hematuria, nutcracker syndrome, CT scan, surveillance

Nutcracker syndrome (NCS) is caused by compression of the left renal vein (LRV) between the aorta and the superior mesenteric artery (SMA) where it passes in the fork formed at the bifurcation of these arteries (1). One of the most common symptoms of this entity is spontaneous hematuria but, it may be totally asymptomatic, presenting as an incidental radiographic finding. The incidental diagnosis of a NCS, after a closed abdominal trauma, has not been reported before. Herein, we report a case.

CASE REPORT

A 32 year old male without medical or surgical antecedents of interest, victim of a road traffic accident causing leg and abdominal trauma, was admitted to the emergency service. He suffered of continuous, non irradiating pain in the left half of the abdomen and the left thigh.

Physical examination showed the patient to be conscious and presenting moderate paleness of the skin and mucosal membranes. Abdominal palpation was normal so was neurological exam. His thigh was swollen and he was unable to move it. Blood pressure: 80/40 mmHg. Heart rate: 80 beats/min. The initial blood tests showed hemoglobin 12.2 g/dl, hematocrit 37.3%, 22,200 leukocytes/mm^3, neutrophils 82%, and creatinine 0.6 mg/dl. The coagulation test results were normal. The radiograph concluded to a femur middle diaphyseal complex fracture type C1.1 according to the AO/OTA Classification. Abdominal ultrasound was normal. Under general anesthesia and radioscopic control, he underwent closed reduction and internal fixation of the femur fracture with intramedullary femoral nailing. Three days post-operatively, he presented a painless gross hematuria. There were no associated symptoms; no lower urinary tract symptoms, no fever, no gastro-intestinal symptoms. His physical examination was normal. His blood pressure was 110/70 mmHg, pulse rate 80/min, respiratory rate 16/min, and body temperature 37.3°C. Laboratories included a complete blood count which showed a normal white blood cell count and an haemoglobin of 12g/dl. The electrolytes and renal function tests were normal. The urine culture was negative. When we asked him again, he reported gross hematuria long time ago. Renal ultrasonography showed normal renal size and outline with no anatomical defect. CT scan concluded to a compression of the left renal vein between the aorta and the superior mesenteric artery, with multiple collateral pathways and varicosities in the renal hilum and around the kidney and confirmed the diagnosis of nutcracker syndrome (Figure 1). There was no evidence of any intrarenal pathology. Cystoscopy showed little bleeding from the left ureteral orifices. The initial option chosen by the medical team was surveillance, mostly because the patient was not bothered enough by his symptoms to warrant an intervention. The patient was discharged without any specific medication. Four months after, radiological control showed the consolidation of femoral fracture without any complications. Functional result is excellent; the motion of hip and knee was complete. With 12 months follow-up, the patient is doing well, without recurrence of lumbar pain or hematuria. The patient remained asymptomatic and had returned to his usual life. Urinalysis is normal.

CTscan findings are the same as the first one (same collateralization and same aorto-mesenteric angle).

Figure 1 : Computerized tomography shows left renal vein compressed between abdominal aorta (black asterix) and superior mesenteric artery (yellow arrowhead).

DISCUSSION

Computed tomography scanning (CTscan) is actually the exam of choice in the evaluation of abdominal trauma. It allows a rapid identification of intra-abdominal injuries with a high level of specificity and sensitivity and can assist the trauma surgeon in determining operative versus non-operative management. Also, it is an excellent modality for detecting other pathologic conditions that are unrelated to the trauma and may be clinically significant (2). Our patient was admitted mainly for femur middle shaft fracture. CTScan discovered incidentally a NCS totally asymptomatic up to now. NCS is a rare condition characterized by the entrapment of the LRV between the SMA and the aorta (3). It was first described in 1950 by El Sadr and Mina (4). In 1972, de Schepper gave the name “nutcracker syndrome” to the disease (5) as the LRV between the aorta and the SMA resembled a nut between the jaws of a nutcracker.

NCS is a rare condition with only few reported cases in adult (6). Its incidence is likely underestimated. It is more prevalent in the female aged between 20 to 40 years old (6).

The pathophysiology of NCS is not well known. It was proposed that posterior renal ptosis with stretching of the LRV may be a factor (7). In recent studies, abnormal branching of the SMA from the aorta was identified as its cause (8, 9). NCS is classified as either anterior or posterior. The anterior NCS, which is the most common, is defined by the compression of the LRV between the aorta and the SMA. The posterior NCS is defined by the retro-aortic course of the LRV causing a

compression of the LRV between the aorta and the vertebral column (10). The compression causes LRV hypertension, and creates a higher venous pressure gradient, leading to congestion of the left kidney. This may result in rupture of the thin walled vein into the renal calyceal fornix with the clinical presentation of intermittent gross or microscopic haematuria as in our case. Moreover, the compression leads occasionally to renal hilar varices formation (11, 12) and collateral venous circulation formation such as a prominent left ovarian vein or testicular vein causing vulvar varices in females or varicoele in males (7). Common manifestations for NCS include left flank pain, usually associated with hematuria, either microscopic or macroscopic (1, 13). Other possible symptoms include left sided varicocele (14, 15) pelvic congestion syndrome (16), chronic fatigue syndrome (17), orthostatic proteinuria (18), gastrointestinal symptoms (19) and arterial hypertension (20). NCS cannot be diagnosed with routine diagnostic methods. Therefore, it is easily misdiagnosed or undiagnosed (12). Work up for NCS includes a series of tests such as Doppler ultrasound (DUS), CT scan, magnetic resonance imaging, angiography and retrograde phlebography (21, 22). DUS is the initial diagnostic test. Sensitivity is 78% and specificity up to 100% (17). CT scan shows compression of the left renal vein between the aorta and superior mesenteric artery and the coexistence of abnormal venous collaterals (23).

The most reliable non-invasive method for diagnosis of the NCS is CT angiography (24).

Treatment of this syndrome is still controversial (25). Various ways has been reported. Treatment depends on the severity of the symptoms. Surveillance constitutes a valuable choice in selected cases with mild symptoms, such as microscopic hematuria and short periods of painless gross hematuria (1) as in our case. A close follow up is required in this situation. Hemostatic drugs have little effect in the NCS (11).

On the other hand, significant hematuria, severe flank pain, renal functional impairment, and inefficacy or aggravation of conservative treatment after 24 months of follow-up might require surgery (8, 9, 26-28). It includes especially intravascular and extravascular stenting (9, 29). Medial nephropexy with excision of the renal varicosities, left renal vein bypass, transposition of the left renal vein and autotransplantation have been described (7, 8, 27, 30, 31).

CONCLUSION

The NCS is a very uncommon condition. It may be totally asymptomatic and discovered as a transient incidental finding after blunt abdominal trauma. It represents a challenge for urologists in terms of accurate diagnosis and proper management.

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