A Unique Case of Wilkie Syndrome Reported in a Middle-Aged Female with Posterior Nutcracker Syndrome

Roxana Elena Mirică

roxmirica@yahoo.com

Carol Davila University of Medicine and Pharmacy

Bogdan Apan

Social Insurance Medicine Office

Case Report

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Abstract

Nutcracker and Wilkie syndromes are rarely encountered, often being diagnosed incidentally during imaging investigations for other conditions. In this paper, we present the case of a 36-year-old patient with quasi-permanent symptoms characterized by epigastralgia, loss of appetite, early satiety, left lumbar colic pain, and dysuria. The clinical examination revealed a supple abdomen, sensitivity to palpation in the epigastrium and hypogastrium, frequent urination, and severe protein-caloric malnutrition body mass index (BMI = 15 kg/m²). Laboratory tests indicated a persistent microscopic hematuria without proteinuria and repeated urinary infections. Abdominal-pelvic ultrasound with Doppler showed a dilated left renal vein (LRV) up to 10 mm left of the paraaortic (Nutcracker syndrome) and duodenal obstruction with distension at this level, also confirmed by gastroduodenoscopy (EGD) (Wilkie syndrome). Abdominal-pelvic angioCT results indicated a malformation of the left renal vein dilated by compression in the aorto-mesenteric clamp and communicating with an aberrant left paravertebral and paraspinal network extending to L1 and L5 and a thrombosis of the left ovarian vein. The patient benefited from conservative treatment.

INTRODUCTION

Superior mesenteric artery (SMA) syndrome (Wilkie syndrome, Cast syndrome, aorto-mesenteric duodenal compression syndrome or chronic duodenal ileus) represents a rare cause of obstruction of the upper digestive tract (1). Wilkie syndrome was first described in 1861 by an anatomopathologist named Baron Carl von Rokitansky and in 1927, English surgeon Sir David Percival Dalbreck Wilkie provided clinical, pathophysiological details and therapeutic suggestions after analyzing a batch of patients (2, 3). The incidence rate is up to 1% in the adult population and occurs predominantly in females, with the onset happening in childhood (over the age of 10) (1). Wilkie syndrome can be of congenital cause (anatomical shortening of the Treitz ligament with suspension of the duodenum in an abnormal position) or acquired, which is the most common form (by the significant reduction of perivascular adipose tissue in the aorto-mesenteric region as a consequence of some medical, psychological, or surgical conditions) (4, 5). A genetic predisposition seems evident in two cases one of which involves identical twins (6) and another that features an intrauterine diagnosis (7). The triggering factors that cause the syndrome to manifest in adults are neoplasia, malabsorption syndromes, cachexia, AIDS, hypercatabolic states (trauma) and burns, significant weight loss, surgical correction of scoliosis, paraplegia, and anorexia nervosa (5, 8). The main physiopathological mechanism involved in Wilkie syndrome is the reduction of both the angle and the distance between the SMA and the abdominal aorta, with values between 6° and 22°, respectively, or between 2 and 8 mm. Normally, the angle between the arteries is 28–65°, and the distance between them is 10–34 mm (9) the angles are correlated with BMI (10).

Nutcracker syndrome is another rare disorder characterized by compression of the LRV between the SMA and the abdominal aorta, leading to an increase in renal pressure and the pelvic venous system; this is the anterior syndrome, which is the most frequently encountered form. The posterior syndrome (rare form) is represented by the compression of the LRV between the abdominal aorta and the lumbar
vertebra (11, 12) and the inverted syndrome occurs when there is pressure on the right renal vein (13).

Nutcracker syndrome was first described in 1937 by the pathologist Grant (14), and in 1974, a team of specialists led by Dr. Schepper continued the research (13). The prevalence of Nutcracker syndrome is higher in women between the ages of 20 and 30 years and in those with a low BMI (15).

We present the clinical case of a 36-year-old patient, who, about 10 years ago, was diagnosed with posterior Nutcracker syndrome (rare form) associated with Wilkie syndrome.

**CASE REPORT**

We present the case of a 36-year-old patient with a family history of Sjogren’s syndrome (present in the mother) and a significant personal history of ichthyosis vulgaris from childhood. The patient was diagnosed with micropolycystic ovaries in 2014, with antiphospholipid syndrome in the context of the detection of an ovarian vein thrombosis and deficiency of protein C and S and of coagulation factors II, and with suspicion of thrombophilia in 2016 and lupus-like syndrome in 2019. On multiple occasions, the patient was admitted to specialized clinical hospitals during a period of intermittent symptomatology that lasted several years and was characterized by postprandial epigastric pain, early satiety, loss of appetite, left lumbar and hypogastric colic pains, and post micturition dysuria. The objective examination revealed an aspect of severe protein-caloric malnutrition (BMI = 15.4 kg/m²), supple abdomen, sensitivity to palpation in the epigastrium and hypogastrium and quasi-permanent pollakiuria. The laboratory tests indicated a persistent microscopic hematuria (onset from the age of 5) without proteinuria and repeated urinary infections with a positive urine culture of mixed etiology for which antibiotic therapy was instituted. The complete blood count, the liver, kidney and thyroid function tests and the ionogram showed normal values. From an imaging perspective, the first abdominal-pelvic angio-CT was performed on the patient in 2014 and revealed an ectatic LRV, with the left paraaortic less than 1 cm from the aorta. Being drained through a large venous vessel that has a diameter of 1 cm and stemmed from behind the LRV, an aberrant left paravertebral and left paraspinous venous plexus extended between the L1 and L5 vertebrae. Posterosuperior drainage occurred into the left iliac vein in the plane of the L5 vertebral body, posteriorly in the LRV less than 10 mm from the aorta in the L1–L2 disc plane (posterior Nutcracker syndrome). At the same time, a thrombosis of the left ovarian vein was highlighted. One year later, an EGD was performed that revealed penetration with difficulty at the D3 level under the conditions of an extrinsic traction at this level (Wilkie syndrome) following a significant weight loss. The last abdominal-pelvic ultrasound evaluation with Doppler showed liquid distension of the D3 duodenum with a diameter of 1.7 cm (Wilkie syndrome). Concurrently, the LRV was reevaluated, highlighting an important difference in caliber between 7.7 mm prepensa (with continuous turbulent flow) and 1.7 mm post pensa (flow at the limit of detection). The distance between the aorta and the SMA in the region of the renal compression was 2.3 mm and was 2.8 mm in the region of the duodenal obstruction. Abdominal-pelvic angio-CT reevaluation (Figs. 1 and 2) highlights the emergence of the SMA under an angle of 21°; between the proximal segment and the aorta, there is a maximum space of 3 mm and the distal can be up to 8 mm.
SMA syndrome is a rare entity, occurring most frequently in women between the ages of 30 and 40 years (16).

In the present case, when the acquired form of Wilkie syndrome was identified, the patient presented a mild symptomatology (postprandial epigastralgia and early satiety) compared to the clinical picture that is characteristic of a severe obstruction, which may involve severe nausea accompanied by recurrent vomiting (5). The triggering factor was a malabsorption syndrome due to multiple food intolerances and allergies known in the personal history of the patient. The patient’s significant weight loss (BMI = 15 kg/m2) led to the reduction of perivascular adipose tissue in the aorto-mesenteric region, representing the main cause of the SMA syndrome.

The necessary investigations for the diagnosis of Wilkie syndrome are abdominal ultrasound, EDS, angio-CT, and MRI imaging (1).

Imaging diagnostic criteria are as follows:

1) Obstruction of the duodenum at the D3 level

2) The aorto-mesenteric angle ≤ 25° and the distance ≤ 8 mm

3) Suspension of the duodenum by shortening of the Treitz ligament or anomalies of the SMA (17)

In the presented case, the abdominal ultrasonography was highly suggestive, identifying and measuring the angle (< 21°) and the aorto-mesenteric distance (maximum 3 mm in the proximal region and up to 8 mm in the distal region), and the EGD showed extrinsic traction at the D3 level.

Therapeutic options for Wilkie syndrome include conservative treatment consisting of gastric decompression with a nasogastric tube, correction of hydroelectrolytic disorders, and enteral or total parenteral nutritional support (5). Surgical treatment is represented by gastrojejunostomy, duodenojejunostomy (5), Roux Y reconstruction or infrarenal transposition of the SMA (16). In the present case, the patient did not require all these therapies, being hemodynamically and electrolytically balanced. At the same time, correcting the malabsorption syndrome resulted in the advantage of weight gain.

Description of Nutcracker syndrome

Nutcracker syndrome is diagnosed incidentally during imaging investigations (18). The triggering factors are anatomical variants of the left kidney, hyperlordosis, and significant weight loss (19). In our case, the patient presents a particular, very rare anatomical form, namely, the posterior type characterized by compression of the LRV between the aorta and the lumbar vertebra (20) (less than 10 mm from the aorta; normal values are between 10 and 28 mm) (14), and elements were highlighted in angio-CT abdominal-pelvic with contrast substance and confirmed ultrasonographically. Venography remains the gold standard for confirming the diagnosis of Nutcracker syndrome (14), but the patient could not benefit from...
this investigation due to the fact that the femoral vein approach was not successful after several attempts and, additionally, due to the suspicion of thrombophilia and antiphospholipid syndrome that the patient presented.

The clinical picture of Nutcracker syndrome is characterized by left lumbar and left flank pain, intermittent macro/microscopic hematuria and pelvic congestion syndrome (dysmenorrhea, dyspareunia, dysuria) with only orthostatic proteinuria (14) being absent in our patient’s case.

From a therapeutic point of view, for severe cases, surgical and endovascular interventions are recommended, such as the transposition of the LRV or the implantation of a self-expanding intravascular metallic stent in the stenotic tract of the LRV(16) in order to revascularize and decompress it (18), transposition of the SMA, renal autotransplantation or nephrectomy (16).

In mild or moderate cases, the recommendation is to monitor the entire progression of Nutcracker syndrome in patients. During her years of living with the disorder, the patient was hospitalized in several medical institutions, but a therapeutic scheme could not be established to control the existing symptoms. Therefore, various therapies are still being implemented, although without improving the patient’s condition in the long term.

**Conclusions**

1. Both Nutcracker syndrome and Wilkie syndrome represent rare pathologies that could easily be overlooked in the absence of appropriate laboratory analyses and imaging investigations, which, in the present case, include abdominal ultrasonography with Doppler signal and abdominal-pelvic angioCT to help diagnose the condition with certainty.

2. The peculiarity of the current case is that the patient presents a very rare form of Nutcracker syndrome (posterior type) associated with another rare syndrome, Wilkie. The simultaneous occurrence of the two syndromes is not frequent in the specialized literature.

3. Future clinical studies of Nutcracker and Wilkie syndromes are necessary for faster diagnosis and targeted treatments to improve the quality of life of patients who suffer from these conditions.

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Conflict of interest

The authors declare that they have no conflict of interest.

Availability of data and materials

Not applicable.

Consent to publish

Informed consent to publish was taken from the patient to publish the case report.

References


Figures
Figure 1

Abdominal-pelvic angioCT
Figure 2

Abdominal-pelvic angioCT