BMC Nephrology

CASE REPORT

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# A unique case report of wilkie syndrome in a middle-aged female with posterior nutcracker syndrome

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**Abstract**

Nutcracker syndrome and Wilkie syndrome are rare and often diagnosed incidentally during imaging investigations for other conditions and, on occasion, together. In this paper, we present the case of a 36-year-old patient with quasi-permanent symptoms including epigastralgia, loss of appetite, early satiety, left lumbar colic pain, normal stool and dysuria. The clinical examination revealed a non-distended abdomen, sensitivity to palpation in the epigastrium and hypogastrium regions, frequent urination in small amounts, and a body mass index (BMI) of 15 kg/m2, indicating severe protein calorie malnutrition. Laboratory tests indicated persistent microscopic hematuria without proteinuria with repeated urinary infections. Abdominal-pelvic ultrasound with Doppler revealed a left renal vein dilated up to 10 mm in the left paraaortic region (Nutcracker syndrome) and distal duodenal obstruction with distension in the same region, which was also confirmed by gastroduodenoscopy (Wilkie syndrome). Abdominal–pelvic computed tomography angiography revealed a malformed and dilated left renal vein that was compressed as a result of aorto-mesenteric obstruction and communicating with an aberrant left paravertebral and paraspinal network extending to L1 and L5 and thrombosis of the left ovarian vein. The patient benefitted from conservative treatment, which intended to correct the malabsorption syndrome with the help of a nutritionist, who suggested a personalized diet to help gain weight. As a result, the patient was able to avoid surgical treatment. The case is peculiar in that the patient presented with a very rare form of Nutcracker syndrome (posterior type) associated with another rare syndrome, Wilkie syndrome.

**Keywords** Nutcracker syndrome, Superior mesenteric artery (SMA) syndrome, Duodenal dilation, Dilated left renal vein, Doppler abdominal ultrasound, CT angiography abdomen and pelvis

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## Introduction

Superior mesenteric artery (SMA) syndrome (Wilkie syndrome, Cast syndrome, aorto-mesenteric duodenal compression syndrome) is a rare cause of obstruction of the upper digestive tract [1] characterized by compression of the duodenum between the aorta and superior mesenteric artery, whereas Nutcracker syndrome involves compression of the left renal vein between the aorta and superior mesenteric artery or between the aorta and the lumbar vertebra. Wilkie syndrome was first described in 1861 by an anatomopathologist named Baron Carl von Rokitansky, and in 1927, the English surgeon Sir David Percival Dalbreck Wilkie provided clinical and pathophysiological details and therapeutic suggestions after analysing data from a group of patients [2, 3]. Affecting only 0.2%to 0.78% of adults, most of whom are female, the first symptoms of the disease present 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). Certain patients with SMA syndrome have genetic predisposition, such as in a case report involving identical twins [4] and another case featuring an intrauterine diagnosis [5]. Wilkie syndrome can also be acquired and is the most common form (due to the significant reduction in perivascular adipose tissue in the aortomesenteric region as a consequence of medical, psychological or surgical conditions) [6, 7]. The factors that cause the syndrome to manifest in adults are neoplasia, malabsorption syndrome, cachexia, AIDS, hypercatabolic states (trauma) and burns; significant weight loss, including bariatric surgery [8] and surgical correction of scoliosis, paraplegia, and anorexia nervosa [7, 9]. The incidence of superior mesenteric artery syndrome has increased in children and young patients who are placed in a body cast after corrective spinal surgery. This procedure lengthens the spine [10], distorting the anatomy of the superior mesenteric artery [11] by displacing its origin and reducing the aorto-mesenteric angle [12].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. Typically, the angle between the arteries is 28–65°, and the distance between them is 10–34 mm [13]; these angles are correlated with BMI [14].

Nutcracker syndrome is another rare disorder characterized by compression of the left renal vein(LRV) between the SMA and the abdominal aorta, leading to an increase in renal pressure and the pelvic venous system (compression occurs in front of the aorta); this is the anterior syndrome, which is the most common. Posterior syndrome (rare form) is characterized by compression of the left renal vein (LRV) between the abdominal aorta and the lumbar vertebra (compression occurs behind the aorta) [15, 16], and inverted syndrome occurs when there is pressure on the right renal vein [17]. Nutcracker syndrome was first described in 1937 by a pathologist named Grant [18, 19], and in 1974, a team of specialists led by Dr. Schepper continued the research [18]. The prevalence of Nutcracker syndrome is higher in women between the ages of 20 and 30 years and in those with a low BMI [20].

We present the clinical case of a 36-year-old patient who, approximately 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, antiphospholipid syndrome on the basis of the presence of ovarian vein thrombosis and deficiency in proteins C and S and in coagulation factor II, suspicion of thrombophilia and a food pluriallergic terrain, owing to the presence of a single allele (DQA1*0501) in the gluten intolerance genetic test. Her serology was negative for celiac disease even though her duodenal mucosa biopsy revealed borderline results (Marsh score 3a– a minor degree of gluten enteropathy). She has experienced nausea, bloating and weight loss but no gastrointestinal transit changes and has been monitored since 2016. She presented with lupus-like syndrome in 2019.On multiple occasions, the patient was admitted to specialized clinical hospitals for intermittent periods of symptoms that lasted several years and was diagnosed with postprandial epigastric pain, early satiety, loss of appetite, left lumbar and hypogastric colic pains, and post micturition dysuria. The objective examination revealed severe protein-caloric malnutrition (BMI = 15.4 $kg/m^2$), a non-distended abdomen, sensitivity to palpation in the epigastrium and hypogastrium regions and quasi-permanent frequent urination. The laboratory tests indicated persistent microscopic hematuria (onset from the age of 5) without proteinuria. However, she experienced repeated urinary infections for which her urine culture was positive and revealed mixed etiology and was treated with antibiotics. Her complete blood count; liver, kidney and thyroid function tests; and electrolytes were normal. She underwent her first abdominal–pelvic angio-CT in 2014, which revealed an ectatic LRV in the left paraaortic region less than 1 cm from the aorta. A large vein with a diameter of 1 cm was observed behind the left renal vein and draining into an aberrant left paravertebral and left paraspinous venous plexus extending between the L1 and L5 vertebrae. The left paravertebral venous plexus drains into two vessels: the posterosuperior region in

the left iliac vein, at the level of the L5 vertebral body and posteriorly in the LRV in the region of the L1–L2 disc plane (posterior Nutcracker syndrome). At the same time, thrombosis of the left ovarian vein was highlighted. One year later, an EGD was performed, which revealed penetration with difficulty at the D3 level under conditions of extrinsic traction at this level (Wilkie syndrome); moreover, the patient experienced significant weight loss. The last abdominal–pelvic ultrasound with Doppler revealed liquid distension of the D3 duodenum with a diameter of 1.7 cm (Wilkie syndrome). Concurrently, the LRV was reevaluated by aortomesenteric compression, highlighting an important difference in caliber between 7.7 mm in proximal stenoses (with continuous turbulent flow) and 1.7 mm at the distal stenoses (flow at the limit of detection). The distance between the aorta and the SMA 2.3 mm in the region of renal compression and 2.8 mm in the region of duodenal obstruction. In 2020, an abdominal-pelvic angio-CT (Figs. 1 and 2) revealed the emergence of the SMA at an angle of 21° and the distance between the proximal segment of the superior mesenteric artery and the aorta (at the passage of the LVR); there was a maximum space of 3 mm, and the distal region can reach 8 mm. In January 2024,the patient underwent new

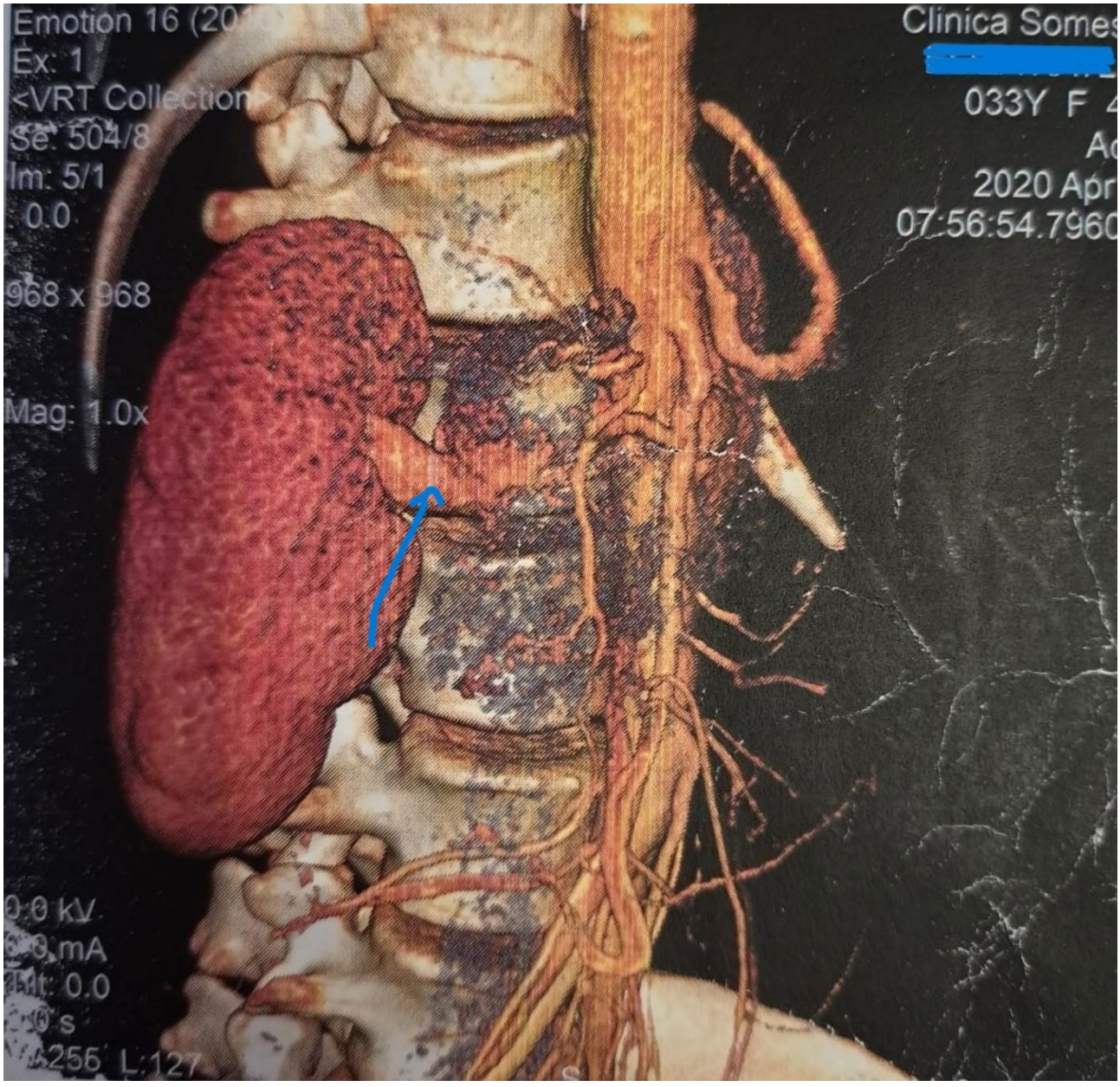


**Fig. 1** Abdominal-pelvic angioCT– posterior region - dilated left renal vein

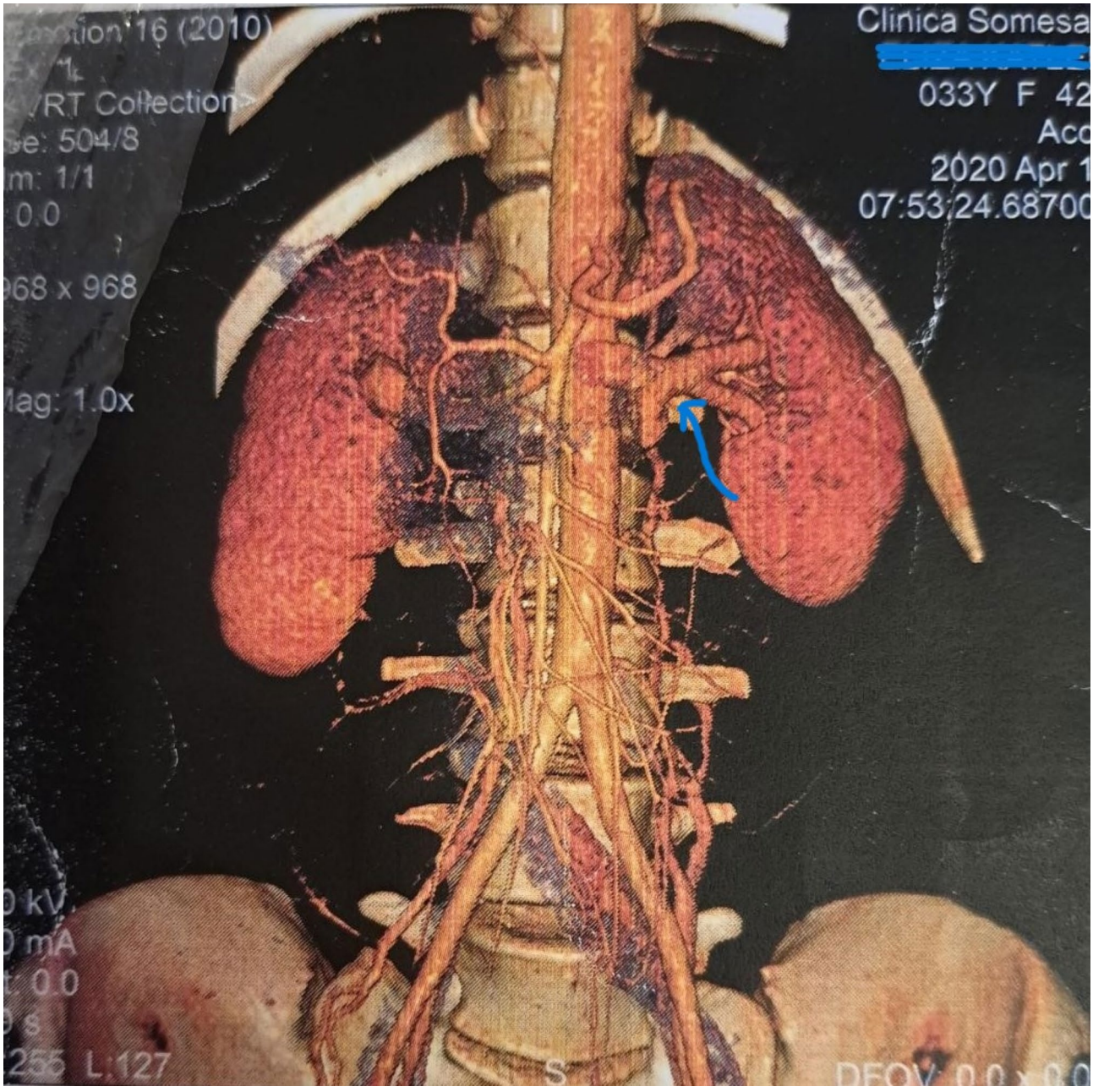


**Fig. 2** Abdominal-pelvic angioCT - the anterior incidence with the visualization of both kidneys, with dilation of the left renal vein

abdominal–pelvic angio-CT, which revealed an aorto-mesenteric angle of 10° and important compression of the LRV by aorto-mesenteric obstruction, with a significant difference in caliber between 9 mm in the proximal region and 2 mm in the distal region. Additionally, the D3 level was compressed by aorto-mesenteric obstruction in the region of the L2-L3 vertebrae, and the distance between the aorta and superior mesenteric artery was approximately 5 mm, without stasis in the proximal duodenal region. Although the latter imaging result highlighted a worsening of the aorto-mesenteric angle, the patient's weight gain of 13 kg led to an improvement in symptoms. Currently, her state of health is good, as she can perform daily activities.

## Discussion

SMA syndrome is rare and occurs most frequently in women between the ages of 30 and 40 years [21].

In the present case, when the acquired form of Wilkie syndrome was identified, the patient presented mild symptomatology (postprandial epigastralgia and early satiety) compared with the clinical picture that is

characteristic of severe duodenal obstruction, which may involve severe nausea accompanied by recurrent vomiting [7]. The contributing factor was malabsorption syndrome due to multiple food intolerances and allergies known in the patient's personal history. The patient's significant weight loss (BMI = 15 kg/m$^2$) probably led to a reduction in perivascular adipose tissue in the aorto-mesenteric region, representing the main cause of SMA syndrome.

The protocols used for the diagnosis of Wilkie syndrome include abdominal ultrasound, gastroduodenoscopy (EGD), angio-CT, and MRI [1].

The imaging diagnostic criteria are as follows:

1) Obstruction of the duodenum at D3.

2) Aorto-mesenteric angle ≤ 25° and distance ≤ 8 mm.

3) Suspension of the duodenum by shortening of the Treitz ligament or anomalies of the SMA [22].

In the present case, 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 EGD revealed extrinsic compression at the D3 level.

Therapeutic options for Wilkie syndrome include conservative attempts at treatment consisting of gastric decompression with a nasogastric tube, correction of hydroelectrolytic disorders, and enteral or total parenteral nutritional support [7]. Surgical treatment includes gastrojejunostomy, Roux-en-Y duodenojejunostomy [7] or infrarenal transposition of the SMA [21]. Because the patient was hemodynamically stable and electrolytically balanced, we pursued a non operative approach in which the malabsorption syndrome was corrected, resulting in weight gain. Because the patient has food allergies and gluten sensitivity (she has not met all the criteria for the diagnosis of celiac disease), the nutritionist designed a customized diet that involves the exclusion of inflammatory foods and those that harm the patient (primarily lactose and gluten). Additionally, the nutritionist constructed an eating and exercise schedule with regular, frequent, high-protein, high-calorie meals and supervised minimally physical exercise to improve the muscles. Notably, the patient improved in terms of weight, which improved her symptoms and overall health.

Nutcracker syndrome is diagnosed incidentally during imaging investigations [23].The contributing factors are anatomical variants of the left kidney, hyperlordosis, and significant weight loss [24]. In our case, the patient presented a particular, very rare anatomical form, namely, the posterior type, characterized by compression of the LRV between the aorta and the lumbar vertebra [25] (less than 10 mm from the aorta; normal values between 10 and 28 mm) [19]. These elements were highlighted on angio-CT of the abdomen–pelvis with contrast substances and were confirmed ultrasonographically. Venography used to be the gold standard for confirming the diagnosis of Nutcracker syndrome [19], but the patient could not benefit from this investigation because the femoral vein approach was not successful after several attempts and, additionally, because of 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 [19], the latter being absent in our patient's case.

From a therapeutic point of view, for severe cases, surgical and endovascular interventions are recommended, such as transposition of the LRV or implantation of a self-expanding intravascular metallic stent in the stenotic tract of the LRV [21], to revascularize and decompress it [23]. Other recommendations include transposition of the SMA, renal autotransplantation or nephrectomy [21]. In our case, the option of endovascular stenting was offered to the patient, but she opted not to proceed with it, considering that placement of a stent does not represent a permanent solution and that there is a risk of stent migration. The patient was evaluated by the vascular surgeon, who recommended monitoring by the nephrologist and maintaining a normal weight so that the angle did not narrow even more and the symptoms that could lead to the need for surgical intervention did not appear. In view of normal renal function, no further intervention was necessary for treating her disorder, and a wait-and-see strategy was recommended.

In mild or moderate cases, monitoring the entire progression of Nutcracker syndrome in patients is recommended. During her years of living with the disorder, the patient was hospitalized at several medical institutions, but a therapeutic scheme could not be established to control the existing symptoms. Therefore, various therapies are still being implemented, although they have not led to long-term improvements in the patient's condition. Because the patient managed to gain weight, she no longer experienced left lumbar pain, and the dyspeptic syndrome disappeared. Laboratory tests revealed persistent hematuria, which has fluctuated over the past few months, with rare instances of exacerbation. The other tests yielded normal values. Fortunately, the patient follows all the doctors' recommendations and returns for follow-up appointments.

## Conclusions

1. Both Nutcracker syndrome and Wilkie syndrome are rare pathologies that can easily be overlooked in the absence of appropriate laboratory analyses

and imaging investigations. In the present case, abdominal ultrasonography with Doppler signals and abdominal-pelvic angio-CT were used to establish a definitive diagnosis.
2. The simultaneous occurrence of these two syndromes, especially the posterior type of Nutcracker syndrome, is rare.
3. Future clinical studies of Nutcracker syndrome and Wilkie syndrome are necessary for precise diagnosis and targeted treatments to improve the quality of life of patients who suffer from these conditions.

**Acknowledgements**
Not applicable.

**Author contributions**
Roxana Mirica wrote the main manuscript text and Bogdan Apan communicate with the patient and prepared the Figs. 1 and 2. All authors reviewed the manuscript.

**Funding**
No funding was received.

**Data availability**
No datasets were generated or analysed during the current study.

## Declarations

**Competing interests**
The authors declare no competing interests.

**Conflict of interest**
The authors declare that they have no conflict of interest.

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

Received: 10 March 2024 / Accepted: 4 February 2025
Published online: 11 February 2025

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## Publisher's note