

A rare cause of recurrent abdominal pain; the coexistence of Wilkie's syndrome and nutcracker syndrome

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ABSTRACT

Nutcracker syndrome is a syndrome that has clinical symptoms such as hematuria, orthostatic proteinuria, pelvic congestion, left-sided varicocele, hypertension, and flank pain. These symptoms occur because of the compression of the left renal vein between the aorta and the superior mesenteric artery. In Wilkie's syndrome, the third part of the duodenum is compressed between the superior mesenteric artery and the abdominal aorta, causing various gastrointestinal symptoms. The coexistence of these two syndromes is a rare condition and is included as case reports in the literature.

This article presents the clinical and radiological results of a 17-year-old male patient who had recurrent abdominal pain due to Wilkie's syndrome, which was accompanied by nutcracker syndrome that caused proteinuria, and for this reason, the patient was referred to the Pediatric Rheumatology outpatient clinic with a preliminary diagnosis of familial Mediterranean fever.

Key words: superior mesenteric artery syndrome, abdominal pain, proteinuria, renal nutcracker syndrome.

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INTRODUCTION

Nutcracker syndrome (NS) is a clinical condition that occurs when the left renal vein (LRV) is compressed between the superior mesenteric artery (SMA) and abdominal aorta (AA). Compression of the LRV in NS leads to the development of venous varices surrounding the renal pelvis, ureter, and gonadal vein, and causes a pressure increase in the LRV. As a result of the increased pressure, clinical complaints and findings such as hematuria, orthostatic proteinuria, pelvic congestion, left-sided varicocele, hypertension, and flank pain may occur.^{1,2}

The SMA syndrome known as Wilkie's syndrome (WS) is defined as the compression of the third segment of the duodenum between the AA and SMA which gives nonspecific signs and symptoms.³ Sometimes this condition may accompany the anterior NS. These patients have complaints of aggravating intermittent abdominal discomfort depending on the severity of the obstruction in the duodenum.⁴

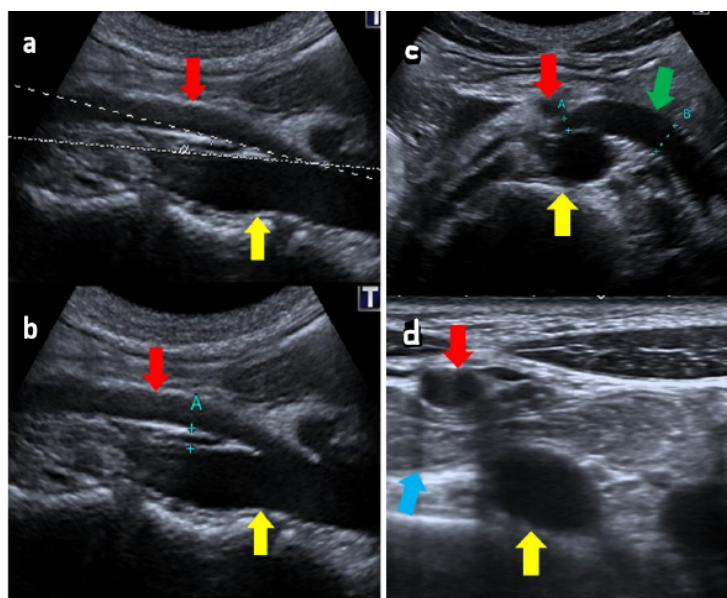
In this article, we investigated the recurrent abdominal pain and proteinuria in which these two syndromes are seen together, and with the preliminary diagnosis of familial Mediterranean fever (FMF), a rare case diagnosed by color

Doppler ultrasonography (CDU) and contrast enhanced computed tomography (CECT) is presented.

CASE REPORT

For 2 years, a 17-year-old male patient had been experiencing excruciating abdominal pain and accompanying nausea and/or vomiting attacks, especially after meals. The occurrence of these symptoms was occasionally once a week, sometimes once every 2 weeks, and lasted for approximately 1 day but was not accompanied by fever. His pediatrician suspected FMF due to recurrent abdominal pain and the existence of proteinuria and then, with the result of the gene analysis, decided to refer to a pediatric rheumatologist. In addition to these symptoms, the patient also had regurgitation, loss of appetite, loss of 5 kg in the last year, and occasionally had severe flank pain. The patient did not have a rheumatological disease and FMF in his family history; his physical examination was height: 177 cm (50-75p), weight: 61.5 kg (25p). His vitals signs, abdominal examination and other system examinations were normal. In the laboratory examination, WBC: 7300/ μ L, Hb: 14.9 g/dL, Plt: 334,000/ μ L, neutrophil 5300/ μ L, biochemical analysis were normal, C-reactive

FIGURE 1. In gray scale ultrasonography (US), the angle between the abdominal aorta and the superior mesenteric artery (SMA) was measured 8 degrees (a) and the distance was 5 mm (b) (yellow arrow: abdominal aorta, red arrow: SMA). (c, d): Gray scale ultrasonography (US) showed that the left renal vein was compressed and proximal to dilated (c) and distally the third part of the duodenum was similarly compressed (d) (yellow arrow: abdominal aorta, red arrow: SMA, left renal vein: green arrow, duodenum: blue arrow)

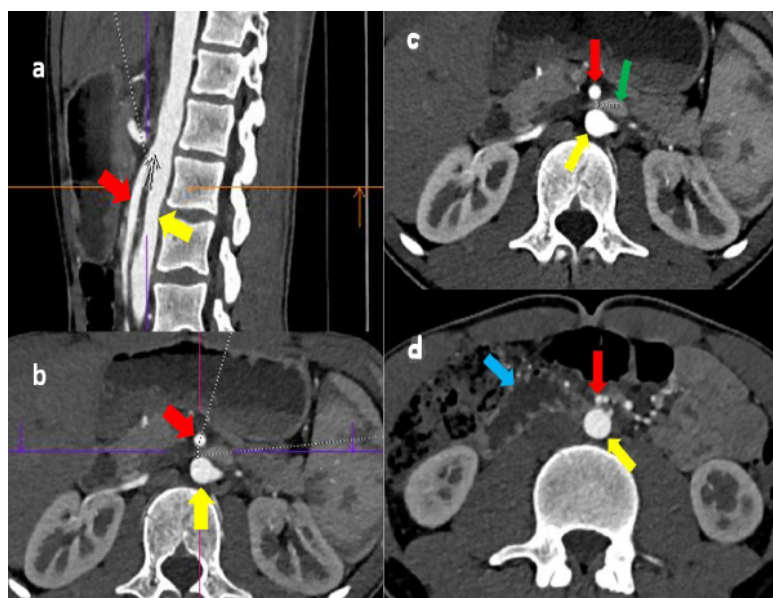


protein 1.31 mg/L, sedimentation 4 mm/hour, and fibrinogen was detected as 223 mg/dL. Urine density analysis was detected as 1018, pH: 7, protein +++, and no hematuria was observed. The patient's previous blood test results were also examined from the computer, and it was observed that the inflammatory parameters suggestive of FMF never increased. Urinalysis of the day and night, abdominal ultrasonography, and CDU were planned for the patient with proteinuria. The protein level of the 12-hour urine examination at night was 5.76 mg/m²/hour, and during the daytime the 12-hour urine examination was 35.2 mg/m²/hr. On the US, the angle between the AA and the SMA was measured as 8 degrees and the distance was 5 mm. At this level, the compression and the proximal dilatation of the LRV were observed (suggestive of NS). In addition to this, it was observed that the third part of the duodenum was also under pressure in the distal part (suggestive of WS) (*Figure 1*). Thus, similar results were found in the contrast enhanced abdominal CT (CECT) which was performed to clarify the diagnosis. The diagnosis was confirmed by the computed tomography angiography (*Figure 2*). The relevant departments were consulted for surgical procedures for the patient with significant proteinuria and gastrointestinal symptoms.

DISCUSSION

In the WS, there is external pressure at the tip of the duodenum.³ WS has been shown to occur mainly in women and people with skinny body types, and two-thirds of the cases are between the ages of 10 and 39. In healthy individuals, mesenteric adipose tissue and lymphatic tissues in the para-aortic region and around the SMA protect the duodenum from compression.⁵ Therefore, conditions such as acute weight loss and scoliosis surgery that reduce the aortomesenteric angle may also cause this syndrome.⁶ Duodenal obstruction in this syndrome can be intermittent, partial or complete, acute or chronic. Duodenal obstruction causes backward duodenal stasis, obstruction of the proximal duodenum, and enlargement of the stomach, and it can also cause bile reflux that backs up towards the stomach.⁷ In acute cases, there are symptoms of acute upper gastrointestinal obstruction such as nausea, vomiting, and abdominal pain which is accompanied by the enlargement of the stomach and duodenum,^{8,9} while chronic WS manifests itself in the form of long-lasting, intermittent attacks, abdominal pain and discomfort in the abdomen, and generally shows as nausea and bilious vomiting after meals. Weight loss is also observed in such patients under these

FIGURE 2. Figure 2 a, b. Angle (a) and distance (b) measurements between the abdominal aorta and the superior mesenteric artery (SMA) on contrast-enhanced abdominal CT. (Yellow arrow: abdominal aorta, red arrow: SMA). (c, d): Contrast-enhanced abdominal CT shows that the left renal vein is compressed and proximal to dilated (c) and distally the third part of the duodenum is similarly compressed (yellow arrow: abdominal aorta, red arrow: SMA, left renal vein: green arrow, duodenum: blue arrow)



circumstances.⁹ In our case, the patient had a 2-year history of attacks of vomiting and/or nausea that lasted for approximately one day, because it resembled an FMF attack, the physician mistakenly classified these symptoms as FMF, which is a common cause of abdominal pain, and thereupon, the patient was referred to the Pediatric Rheumatology outpatient clinic. However, FMF is an autoinflammatory disease and it is not possible to think of an autoinflammatory disease without inflammation. Also, the proteinuria in FMF is secondary to amyloidosis, which occurs over a longer period of time.

In NS, an increase in intraluminal pressure occurs as a result of compression of the LRV at the meso-aortic angle. The etiology includes renal ptosis, abnormally high outflow of the LRV, abnormally narrow-angle in the aortic outlet of SMA, pancreatic masses, lymphadenomegaly, and lack of retroperitoneal adipose tissue.¹⁰ Although the disease can occur at any period of time since childhood, it is most commonly seen in healthy and thin women in the third and fourth decades of life.¹¹

Unlike WS, NS usually does not have gastrointestinal symptoms. The most common clinical finding is hematuria, which is attributed to ruptures in the wall of the thin-walled veins at the level of the renal calyces due to increased pressure. Again, depending on the increase in pressure, it has been reported that the development of orthostatic proteinuria is caused by the protein leakage from the calyceal system, and it is generally observed at a rate of 0.6-10.7%. However, in the 112-case NS series of Orcyzk et al., it was reported that hematuria (78.57%), left flank pain (38.39%), varicocele (35.71%), proteinuria (30.36%), and anemia (13.39%) were observed.¹²

CDU is the first non-invasive radiological method in the diagnosis of both compression syndromes. The sensitivity of CDU in NS is 78%, and the specificity is nearly 100%. A peak systolic flow ratio of more than 4.2 and an anterior-posterior diameter ratio of more than 4.0 obtained in the renal vein before and after stenosis is an important indicator. CT and magnetic resonance angiography (MRA) are other non-invasive radiological methods that can show external compression of the duodenum and proximal dilatation of the compression in WS, on the other hand, it can show the compression of the LRV at the aortomesenteric angle and collateral

veins in NS. Measurement of SMA angle is a very important criterion for the diagnosis of NS. Taktak et al. describes the changes of SMA angle in supine and upright positions and upright SMA angle measurement is more efficient than supine SMA angle measurement on clinico-radiological correlation. They don't find any significant correlation with SMA angle, gender and BMI.¹³ In a study that was performed with contrast-enhanced CT in healthy adolescents, the SMA angle, the duodenal, and LRV distance values were measured as 41.1 degrees, 11.2 mm and 8.2 mm for girls, and 42.7 degrees, 12.6 mm and 9.2 mm for boys, and no difference was found according to gender. In this study, in both genders, there was a positive correlation between body mass index with superior mesenteric artery angle, duodenal and left renal vein distances.¹⁴ In our case, the angle and distance values measured in CDU and CECT were lower than the healthy population and were significant in terms of WS and NS.

In cases with mild symptoms in NS, follow-up and conservative treatment are sufficient, while surgical treatment is recommended in cases with hematuria, severe proteinuria, and severe flank pain that leads to anemia.¹⁵

Although the coexistence of these two syndromes in adolescents is quite rare, it has been reported in the literature as case reports that the WS may accompany especially the anterior NS.^{4,7}

In conclusion, pediatricians should suspect WS in the absence of signs of inflammation, especially in recurrent episodes of abdominal pain or vomiting after meals, and the association of NS should be kept in mind if this situation is accompanied by proteinuria. We think this case study is instructive because it highlights the need for physicians to be skeptical, paying attention to every detail of symptoms, as missed diagnoses are inevitable. ■

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