Case report

Nutcracker syndrome complicated with intestinal malrotation

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SUMMARY

Nutcracker syndrome (NCS) is a pathological condition in which the left renal vein (LRV) is compressed between the superior mesenteric artery (SMA) and aorta. NCS can predispose patients to the onset of chronic kidney disease because of persistent increase in LRV pressure. Although NCS in children is often idiopathic, it can also be caused by underlying pathologies such as retroperitoneal tumours. To the best of our knowledge, there have been no reports regarding paediatric cases of NCS complicated with intestinal malrotation. Here, we report the case of a 12-year-old girl with intestinal malrotation complicated with NCS whose haematuria resolved after surgical intervention for intestinal malrotation. The present case findings indicate that intestinal malrotation with concomitant weight loss is a potential underlying aetiology in NCS. Thus, when NCS is especially diagnosed with gastrointestinal symptoms, intestinal malrotation should be considered as an underlying aetiology.

BACKGROUND

Haematuria in children is caused by various pathological conditions, including the nutcracker syndrome (NCS), which involves compression of the left renal vein (LRV) in the angle between the superior mesenteric artery (SMA) and aorta.¹ The increased renal vein pressure proximal to the hilum causes symptoms such as both micro- and macrohaematuria, as well as pelvic and flank pain.² Although NCS is often an idiopathic condition, previous reports have shown that it can also be caused by other pathologies and conditions (eg, pancreatic neoplasms, para-aortic lymphadenopathies, retroperitoneal tumour, and reduced retroperitoneal and mesenteric fat), which potentially narrow the angle between the SMA and aorta.²⁻⁵ Regarding anatomical aspects that predispose patients to the onset of NCS, a likely aetiology includes intestinal malrotation, which is a congenital anomaly that results from abnormal rotation and fixation of the midgut during development.⁶ The resulting weight loss and abnormal arrangement of the intestine, primarily duodenal misalignment on the right side of the abdomen, such that the duodenum does not pass through the aortomesenteric angle, the angle between the SMA and aorta, can predispose patients to the onset of NCS. To the best of our knowledge, however, there have been no reports regarding paediatric cases of preoperatively diagnosed NCS associated with intestinal malrotation.

In contrast, there has been an adult case of intraoperatively and incidentally identified asymptomatic NCS complicated with intestinal malrotation.⁷ The present report describes the first paediatric case of NCS, complicated with (and possibly induced by) intestinal malrotation, wherein haematuria improved after surgical intervention for intestinal malrotation.

CASE PRESENTATION

A 12-year-old girl presented to the emergency department with a 5-day history of abdominal pain and vomiting. Her body weight, height and body mass index were 41.5 kg (−0.29 SD), 149.8 cm (−0.61 SD) and 18.4 (slightly underweight). She had no significant medical history during infancy that included failure to thrive or multiple vomiting episodes. Her social and family history was not significant either. On physical examination, her vital signs were stable. The abdomen was flat and soft, with tenderness in the right-sided abdominal region; there were no signs of peritonitis. Other physical examination findings were normal. Laboratory examination was unremarkable, in which haemoglobin level was 137 g/L and estimated glomerular filtration rate was 127.5 mL/min/1.73 m². No proteinuria but microscopic haematuria with isomorphic red blood cells was present. Imaging studies were not conclusive; abdominal X-ray did not show any dilated bowel or niveau, and abdominal echography, including Doppler analysis, revealed no signs of ascites, inflamed bowels, apparent abnormal vascular arrangement or evidence of congenital anomalies of the kidney and urinary tract. Because of these findings and the patient's relatively good general appearance, further investigations, such as a CT scan, were not performed at the first visit. However, because her symptoms exacerbated for 2 weeks after the first visit, including increasing frequency of vomiting (ultimately occurring at every meal), weight loss of 1 kg, and worsening of haematuria, plain CT imaging was performed.

INVESTIGATIONS

An abdominal plain CT imaging revealed the absence of the ascending colon at the right retroperitoneal area, absence of the duodenum passing through the aortomesenteric portion and presence of most parts of the small intestine at right side of the abdomen, which were indications of intestinal malrotation. These findings were supported by

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Figure 1 Contrast-enhanced CT images. (A) Coronal, (B) axial and (C) sagittal images of severely compressed left renal vein (asterisk) between the superior mesenteric artery (arrow) and aorta (arrowhead). Note that the duodenum does not pass through the aortomesenteric angle and the small intestine, in most parts, is present at right side of the abdomen.

additional contrast-enhanced CT as shown in figure 1. In this patient, CT angiography was not performed, considering the high specificity of contrast-enhanced CT findings and to avoid the excess radiation exposure. Finally, the diagnosis of intestinal malrotation, without any sign of midgut volvulus, was confirmed by an upper gastrointestinal series. Because of the intestinal misalignment, the duodenum did not pass through the angle between the SMA and aorta as mentioned above; this could help narrow the angle to 11° such that LRV was compressed. Based on the imaging characteristics and additional tests to exclude other aetiologies of haematuria, such as nephritis, the patient's haematuria was finally diagnosed as a symptom of NCS.

TREATMENT

A laparoscopic Ladd's procedure as the surgical intervention for intestinal malrotation, without any direct intervention for NCS, was performed.

OUTCOME AND FOLLOW-UP

After surgical intervention for intestinal malrotation, her gastrointestinal symptoms improved; her haematuria disappeared 3 weeks after intervention for intestinal malrotation along with subsequent weight gain. About a year after surgical intervention, she has not developed haematuria and is still under our follow-up.

DISCUSSION

NCS is one of the many causative aetiologies related to the presence of haematuria in children. Its exact prevalence is not known but is suspected to comprise approximately 40% of unexplained cases of haematuria.¹ The pathology of NCS is defined as the compression of LRV between the angle of SMA and aorta, which leads to increased LRV pressure, venous reflux and subsequent rupture of the septum adjacent to the collecting duct.⁸ Definitive diagnostic criteria are not currently available, and the diagnosis is supported by one or more imaging methods, including ultrasound, CT or MRI.¹ Proposed CT-based diagnostic criteria are as follows: (1) narrowing of LRV at the aortomesenteric angle ($<32^\circ$); (2) severe narrowing of LRV at the aortomesenteric portion; (3) LRV diameter ratio (hilar-to-aortomesenteric) ≥ 4.9 and (4) angle between SMA and aorta $<41^\circ$.⁹ Although NCS is often idiopathic, notably, some pathologies and conditions can predispose patients to the onset of NCS. Pancreatic neoplasm, para-aortic lymphadenopathy, retroperitoneal tumour, abdominal aortic aneurysm, and reduced retroperitoneal and mesenteric fat comprise portions of these pathologies and conditions¹²; all of them may narrow the aortomesenteric angle, thus causing

compression of the LRV. Although NCS can spontaneously resolve, it could require therapeutic approaches for the respective underlying aetiologies for these pathologies and conditions. Considering the unfavourable complications of NCS, such as chronic kidney disease due to the persistent increase in LRV pressure or LRV thrombosis, it is crucial to recognise the underlying aetiologies to optimise the therapeutic approaches. In addition, considering the severity of these predisposing aetiologies, it is important that they are examined when NCS is diagnosed so as to prevent their unfavourable complications from arising.

Regarding potential narrowing of the aortomesenteric angle, intestinal malrotation can predispose patients to the onset of NCS, but there have been no reported cases involving the association of NCS and intestinal malrotation in children. However, there was a case of a 28-year-old urologically asymptomatic man incidentally diagnosed with NCS during surgical intervention for intestinal malrotation.⁹

Intestinal malrotation is a congenital anomaly that results from abnormal rotation and fixation of the midgut during fetal development.⁶ Its incidence has not been clearly defined because most patients remain asymptomatic throughout their lives. Most patients, approximately 75%–85%, are diagnosed during infancy; the remaining patients are either diagnosed after infancy or during adulthood.¹⁰ Because of the abnormal intestinal arrangement, the duodenum does not pass between the SMA and aorta, which potentially narrows the angle between these structures. In addition, affected patients typically exhibit abdominal pain and vomiting, which might be caused by the compression of the duodenum by Ladd bands, subsequently leading to weight loss or inadequate weight gain. The combination of the narrowed aortomesenteric angle and concomitant weight loss can predispose patients to the onset of NCS; thus, intestinal malrotation can be an aetiological factor in the onset of NCS.

In the present case, NCS was preoperatively diagnosed based on the incidental finding of haematuria during diagnosis of intestinal malrotation. This is the first paediatric case in which these two pathologies were associated. CT imaging revealed absence of the duodenum in the angle between SMA and aorta, which could allow the angle to narrow to 11° , such that LRV was compressed. After surgical intervention, the patient's haematuria improved without any urological intervention mainly because of weight gain and possibly due to a slight SMA shift that was partly confirmed by post-operative CT imaging. This could help secure the aortomesenteric space.

In conclusion, intestinal malrotation can potentially predispose patients to the onset of NCS. Thus, when NCS is diagnosed, intestinal malrotation should be considered to optimise the therapeutic approach and prevent unfavourable complications.

Learning points

- ▶ Nutcracker syndrome (NCS) is often idiopathic but can also be caused by some pathologies and conditions which potentially narrow the aortomesenteric angle.
- ▶ Intestinal malrotation due to abnormal arrangement of the duodenum and concomitant weight loss can be a potential aetiology of the NCS.
- ▶ When NCS is especially diagnosed with gastrointestinal symptoms, intestinal malrotation should be considered as an underlying aetiology.

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