

## Case Report

# Superior Mesenteric Artery and Nutcracker Syndromes in a Healthy 14-Year-Old Girl Requiring Surgical Intervention after Failed Conservative Management

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### ABSTRACT

This case report presents the diagnosis of superior mesenteric artery and nutcracker syndromes in a previously fit and well 14-year-old girl. Although these two entities usually occur in isolation, despite their related aetiology, our patient was a rare example of their occurrence together. In this case the duodenal compression of superior mesenteric artery syndrome caused intractable vomiting leading to weight loss, and her nutcracker syndrome caused severe left-sided abdominal pain and microscopic haematuria without renal compromise. Management of the superior mesenteric artery syndrome can be conservative by increasing the weight of the child which leads to improvement of retroperitoneal fat and hence the angle of the artery. The weight can be improved either by enteral feeds or parenteral nutrition. This conservative management initially helped but not in the long-term as the child started losing weight again. The next step in management is surgery (duodenojejunostomy – if the conservative management fails), which the child went through, remarkably improving their symptoms.

### BACKGROUND

The superior mesenteric artery (SMA) is the second major vessel to arise from the anterior surface of the abdominal aorta. The duodenum and left renal vein pass between the aorta and the SMA. SMA syndrome or Wilkie's syndrome is the external compression of the third part of the duodenum between the aorta and the superior mesenteric artery.<sup>1</sup> It is an uncommon cause of duodenal obstruction most commonly effecting females aged 10-40 with an approximate prevalence of 0.013-0.3% in the general population.<sup>1,2</sup>

Nutcracker syndrome (NCS) is the compression of the left renal vein (LRV) in between the superior mesenteric artery and the aorta. This can lead to abdominal pain, macroscopic or microscopic haematuria, renal failure and dilatation of collateral vessels.<sup>3</sup>

Although SMA syndrome and NCS have related aetiologies the occurrence of both together is highly unusual.<sup>4</sup> Both can be precipitated by weight loss *via* reduction in retroperitoneal fat, or the result of surgical intervention; including correction of sco-

liosis and anything which affects the anatomical relationship of the vessels involved. Therefore, both conditions are more common when there is an excessively acute aortomesenteric angle or a shortened aortomesenteric distance.<sup>1,3,5</sup>

### CASE REPORT

A previously fit and well 14-year-old female, presented to the paediatric emergency department with generalised abdominal pain and symptoms of constipation, there were no indications for admission and she was discharged with laxatives. Several months later she represented on two successive days with similar symptoms, however, this time there was associated fresh blood per rectum. Full blood count (FBC), urea and electrolytes (U&E), liver function tests (LFTs), C-reactive protein (CRP) and coagulation were normal. Abdominal X-ray revealed only non-specific bowel gas and faecal loading. She was discharged with laxatives and an outpatient appointment for oesophago-gastro-duodenoscopy (OGD) and colonoscopy was arranged. It was noted that she had a family history of inflammatory bowel disease.

Oesophago-gastro-duodenoscopy (OGD) and colonoscopy showed mild proctitis and a rectal ulcer but was otherwise normal. Histology exhibited non-specific mild chronic inflammation of gastric mucosa and changes in keeping with the solitary rectal ulcer. Prior to follow-up she presented with progressive vomiting and weight loss and was admitted. Her initial weight had been 51.8 kg (75<sup>th</sup> centile), height 165.5 cm (91<sup>st</sup> centile), body mass index (BMI) 18.9; over a one-month period she dropped to 46.8 kg (50<sup>th</sup> centile) BMI 17.1. Abdominal ultrasound (USS) revealed no pathology and a barium meal and follow through was arranged for the following morning.

The barium meal revealed mild to moderate dilatation of the proximal half of the third part of the duodenum with a diverticulum arising from the area of dilatation, likely secondary to increased intraluminal pressure. There was a short segment of narrowing in the mid segment overlying the spine. The distal duodenal segment was of normal calibre and there was a normally placed duodenojejunal junction. The remainder of the scan was normal (Figure 1). These findings raised the suspicion of SMA syndrome so a computed tomography angiogram (CTA) was arranged.



The CTA was performed along with oral contrast to better visualise the bowel and vasculature together. It showed the third part of the duodenum compressed between the aorta and superior mesenteric artery, confirming SMA syndrome. The aortomesenteric angle was reduced and measured 14.5 degrees (normal 28-65 degrees) (Figure 2) with a shortened aortomesenteric distance of 3.2 mm (normal 10-34 mm).<sup>6</sup> There was also compression of the left renal vein between the aorta and the superior mesenteric artery with proximal dilatation (Figure 3). There was a normal appearance of both renal arteries and kidneys. The patient's left-sided abdominal pain had been thought to be constipation related, however the scan showed that this pain was likely to be caused by NCS.<sup>3,5</sup>

Initially conservative management was trialled because weight gain has been shown to help alter the aortomesenteric an-

gle by increasing retroperitoneal fat, thus reducing symptoms.<sup>1-3,5</sup> Nasogastric (NG) feeds were commenced but were converted to nasojejunal (NJ) feeds due to vomiting and these were better tolerated. Her feeds were gradually increased and she was discharged.

**Figure 2.** Sagittal Plane Computed Tomography Angiogram. A – Superior Mesenteric Angle, Reduced at 14.5 degrees. B – Superior Mesenteric Artery. C – Aorta



**Figure 3.** Axial Plane Computed Tomography Angiogram Revealing Compression of the Left Renal Vein (C) With Associated Dilatation as it Passes between the Superior Mesenteric Artery (A) and The Aorta (B)



Vomiting returned, and as her weight faltered, dropping from 51.4 kg to 46.8 kg (BMI 17), her pain became more prominent, especially on her left side, in keeping with NCS. This worsening was likely due to reduction in retroperitoneal fat leading to a further reduction in aortomesenteric angle.<sup>3,5</sup> She required regular oral opiate analgesia as well as antiemetics. Further supporting the diagnosis of NCS she had several episodes of microscopic haematuria, but there was no decrease in renal function. The decision was made by the multidisciplinary team to perform a gastrostomy to enable her to have feeds *via* gastro-jejunal extension and to decompress her stomach during periods of worsening distention. After

initial success, this too failed and she was commenced on total parenteral nutrition (TPN). She gradually began to gain weight and tolerated better amounts of enteral feeds especially when positioned on her left side.

After repeated admissions with vomiting and pain, and with little improvement seen on scans, she was listed for surgery. Duodenojejunostomy is the recognised surgical treatment for SMA syndrome and involves the anastomosis of the jejunum with the proximal duodenum so as to bypass the area of compression; other surgical options include Strong's procedure whereby the ligament of Treitz is divided to allow the duodenum to move caudally, escaping compression.<sup>7,8</sup> The patient underwent the procedure laparoscopically. Her TPN was weaned and she was started on low volumes of continuous feeds *via* her gastrostomy's jejunal extension before reintroducing a soft oral diet. She tolerated this well and was able to be weaned off TPN within two-weeks. She was discharged shortly after, to be followed-up regularly by a wide range of specialities including dietetics, gastroenterology, pain, psychology and surgical. At discharge she was taking laxatives; mesalazine enemas for inflammation in the rectum; hyoscine butyl bromide, gabapentin and oral opiates as required, in weaning doses for analgesia, as well as a lidocaine patch; and cyclizine as an oral antiemetic. Her condition has greatly improved since her successful surgery and she is gaining weight.

## DISCUSSION AND CONCLUSION

The normal aortomesenteric distance is 10-34 mm and the aortomesenteric angle is normally between 28-65 degrees.<sup>6</sup> Reduction of these parameters increases the likelihood of compression between the aorta and superior mesenteric artery of both the third part of the duodenum and the left renal vein, with symptoms exacerbated by weight loss or surgical interventions. This case highlights that SMA syndrome can be a cause of small bowel obstruction and that compression of the left renal vein can cause abdominal pain, as well as renal compromise, although our patient had no renal involvement. Conservative management of SMA syndrome is often successful but definitive surgical management is sometimes required in the form of duodenojejunostomy which has been shown to be a safe and effective treatment.<sup>1,2,7,8</sup> Although there are reports of SMA syndrome being diagnosed on plain film and abdominal USS, it was not evident in this case and barium meal followed by CTA with oral contrast was required to reveal the diagnosis, which is the recognised gold standard.<sup>1,6</sup> SMA and nutcracker syndromes can have a huge impact on quality of life in the form of regular hospital admissions, pain, vomiting and weight loss. NCS can resolve spontaneously or cause significant morbidity in others, requiring vascular surgical intervention.<sup>3,5</sup> SMA syndrome should be considered as a cause of upper gastrointestinal (GI) obstruction and when diagnosed, NCS should be looked for too. Most reported cases have been managed conservatively, but when conservative

management fails, the decision to proceed to surgery should be taken.

## CONSENT

The authors have received written informed consent from the patient.

## CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

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