Wretched Retching in a 16-Year-Old Girl: A Case Report of Nutcracker and Superior Mesenteric Artery Syndromes

Carrillo MM*, Loi RQ, Kroger K, Legacy MJ and Kumar B

1Department of Pediatrics, Children’s Hospital of Michigan, Detroit, Michigan, USA
2Pediatric Hospitalist, Children’s Hospital of Michigan, Central Michigan University, Michigan, USA

Abstract

Both Nutcracker Syndrome (NCS) and Superior Mesenteric Artery Syndrome (SMAS) are, at their core, compression syndromes. NCS is characterized by an impedance of flow from the Left Renal Vein (LRV) to the Inferior Vena Cava (IVC) by extrinsic compression of the LRV, most commonly by the Superior Mesenteric Artery (SMA) and the Abdominal Aorta (AA), also known as Anterior Nutcracker Syndrome. This syndrome is sometimes asymptomatic; when symptoms do arise, they can include hematuria, left flank pain, pelvic congestion syndrome, or varicocele. Imaging methods used to diagnose NCS include Doppler Ultrasound (US), Computed Tomography Angiography (CTA), Magnetic Resonance Angiography (MRA), and retrograde venography.

Keywords: Nutcracker syndrome; Superior mesenteric artery syndrome; Dysmenorrhea; Anxiety

Introduction

Both Nutcracker Syndrome (NCS) and Superior Mesenteric Artery Syndrome (SMAS) are, at their core, compression syndromes. NCS is characterized by an impedance of flow from the Left Renal Vein (LRV) to the Inferior Vena Cava (IVC) by extrinsic compression of the LRV, most commonly by the Superior Mesenteric Artery (SMA) and the Abdominal Aorta (AA), also known as Anterior Nutcracker Syndrome [1,2]. SMAS is characterized by the compression of the third part of the duodenum by the SMA and the AA [3]. The incidences of these syndromes individually and combined are unknown [2,4,5]. There are approximately 450 cases and 100 cases reported in literature since each syndrome’s first description in 1842 and 1950, respectively [2,6]. We are presenting the case of a 16-year-old girl with chronic abdominal pain and vomiting with a history of abdominal migraines and newly diagnosed NCS and SMAS.

Case Presentation

A 16 year old African American female presented to the ED with a two day history of vomiting characterized as nonbloody, non-bilious, and with a frequency of over ten times per day. Her past medical history included cyclical vomiting for 9 years with multiple hospitalizations, abdominal migraines, dysmenorrhea, anxiety, depression, previous suicide attempt, and chronic marijuana use. She had developed abdominal pain 5 days prior to admission and on the day of admission, could no longer tolerate fluids. She had been admitted twice for similar symptoms in the past month. She denied any URI symptoms, fevers, constipation, or diarrhea. She denied any recent changes in her diet or exposure to sick contacts at home or at school, or any current stressors or changes in her lifestyle. She had been compliant with her medications including OCP and amitriptyline. She attempted rectal prochlorperazine to help with her nausea on the day of presentation to the ED, but it did not help. Of note, she was on her menstrual cycle at the time. She stated that the last time she smoked marijuana was a week and a half prior to admission. Upon presentation, vitals were temperature 37.2 degrees Celsius, blood pressure 130/95 mmHg, pulse 88/min, respiration rate 20/min, and oxygen saturation 100% on room air. Her weight was 51.5 kg. She appeared well developed, thin, and in mild distress. Her abdomen was soft, non-distended, and non-tender to palpation, and non-guarding with normal bowel sounds and no organomegaly. She was cooperative but only provided short answers with flat affect. The remainder of her physical exam was unremarkable. Urine pregnancy test was negative. Urine drug screen positive for marijuana. Serum drug screen was negative. BMP significant for hyponatremia (130), hypochloremia (95), and hypokalemia (2.9). EKG showed normal sinus rhythm. CBC showed mild leukocytosis at 12.5 × 10^9/L that was neutrophil predominant. UA revealed no bacteriuria, but demonstrated mild proteinuria (1+) and hematuria (10-20 RBC). She was admitted to the inpatient pediatric unit and administered ondansetron for...
nausea, MIVF, and pantoprazole. Her home dosage of amitriptyline was continued. Electrolytes were monitored and replaced accordingly. Review of her chart revealed dramatic weight loss of greater than 20 kg over 6 months and a negative EGD study. She stated the weight loss was unintentional. Nutrition was consulted due to insufficient intake. Nutritional supplements were attempted, but she was unable to tolerate them. On Day 5 of admission, CT Abdomen/Pelvis was obtained and showed narrowing of the 3rd portion of the duodenum as it passed between the SMA and AA indicative of Superior Mesenteric Artery Syndrome (SMAS) which may have been the likely cause of her postprandial epigastric pain, nausea, vomiting, and weight loss (Figure 1A to 1D). Left renal vein stenosis, also demonstrated on CT Abdomen/Pelvis, correlated with urinalysis (hematuria/proteinuria) for underlying Nutcracker Syndrome (NCT). SMAS was confirmed by UGI with oral barium contrast and small bowel follow through. Surgery was consulted and recommended Naso Jejunal (NJ) tube placement for feeding. GI was consulted and agreed with this management plan (Figure 2A and 2B). Nephrology was consulted and did not feel her presenting symptoms were related to any renal findings and agreed to follow up outpatient. NJ Feeds were gradually escalated and electrolytes monitored for refeeding syndrome and replaced accordingly. She was then discharged home with NJ tube feeds. Patient’s NJ tube came out after about a month and she did not want it replaced. Her weight was 62 kg during a follow up visit 3 months later and patient reported being compliant with her daily medications: Amitriptyline 25 mg, cyproheptadine 8 mg, omeprazole 40 mg at that time (Figure 3A to 3C).

**Discussion**

NCS is a condition where the LRV is compressed by the AA,
...resulting in decreased outflow to the IVC. This compression can be described as anterior (between the AA and the SMA) or posterior (between the AA and the vertebral column) [2]. This syndrome is sometimes asymptomatic; when symptoms do arise, they can include hematuria, left flank pain, pelvic congestion syndrome, or varicocele [1]. Imaging methods used to diagnose NCS include Doppler Ultrasound (US), Computed Tomography Angiography (CTA), Magnetic Resonance Angiography (MRA), and retrograde venography [1]. For patients with intermittent hematuria, insignificant flank pain, and normal hemoglobin; conservative management with close observation is the best option, especially in young patients less than 18-year-old such as our patient [2]. If conservative management fails, both stenting and open surgical interventions may relieve symptoms; however, selection criteria regarding those interventions are not well-defined [7]. Our patient was found to be doing well with conservative management at the 3 month clinic follow up visit. Nephrology agreed to continue to follow our patient in the outpatient setting, but she is expected to have a good prognosis. SMAS is a condition where the third part of the duodenum is compressed by the SMA. Normally, the duodenum will pass between the SMA and the aorta with the angle between the two vessels ranging from 38° to 56°; the angle is reduced to a range of 6° to 25° in patients with SMAS [3]. This compression can be either congenital or acquired. Congenital causes include malrotation, adhesions, or anatomic variations of the aortic or mesenteric vasculature; acquired causes include significant weight loss, postoperative anatomic changes (for example scoliosis repair), or local pathology (such as a neoplasm) [8]. Patients often present with symptoms such as nausea, intractable vomiting, postprandial epigastric pain, bloating, gastrointestinal reflux, early satiety, and food intolerance [6]. This often leads to a fear of eating which can result in anorexia, weight loss, dehydration, and electrolyte abnormalities [9]. Computed tomography angiography is considered as the gold standard for diagnosis, for it can clearly demonstrate the aortomesenteric angle and distance, and gastric and proximal duodenal dilatation [8]. Current therapy requires long term management, medications, costly parenteral nutrition and rigorous follow-up [3]. In the case of failure of medical treatment, the best surgical options include gastrojejunostomy or duodenojejunostomy, by laparotomy or laparoscopic means [8,10]. One other surgical option is division of the ligament of Treitz with duodenal mobilization [7]. In our patient, it was difficult to distinguish the etiology of her SMAS. Given her history of cyclic vomiting syndrome secondary to marijuana use as well as abdominal migraines, it is possible this led to her significant weight loss, resulting in the SMA compression. However, it could not be ruled out that she had a primary SMAS that led to her vomiting and weight loss. This was thought to be less likely given her normal growth pattern until 16 years of age. Although reports and definitive diagnosis of SMAS as well as NCS in pediatric patients are rare, this case demonstrates that chronic conditions should raise clinical suspicion for these causes [1,3,5,8].

References