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Case Report

The Elusive Diagnosis of Superior Mesenteric Artery Syndrome Coexisting with Nutcracker Phenomenon

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Abstract

Superior Mesenteric Artery (SMA) Syndrome is a rare disease characterized by compression of the distal duodenal segment between the SMA and abdominal aorta (AA). Nutcracker Phenomenon is another condition that occurs when the left renal vein is compressed between the same blood vessels. These 2 rare entities are embodied in an 18-year-old Filipina who presented with a 7-week duration of recurrent abdominal pain and a 5-week duration of intractable vomiting. These symptoms were preceded by hand weakness, finger joint pains, and lower back pain. Due to substantial fluid losses from recurrent vomiting, she was admitted and managed as a case of hypovolemic shock. Further investigation revealed SMA Syndrome with Nutcracker Phenomenon on abdominal CT scan and angiogram. We take the opportunity to present this rare coexistence and discuss its pathophysiology, epidemiology, clinical course, management, and prognosis.

Keywords: Superior Mesenteric Artery; Nutcracker Phenomenon; Vomiting; Abdominal Pain

Case Report

A previously healthy 18- year-old, female, residing in a congested urban city, was brought to the emergency department due recurrent abdominal pain and intractable vomiting.

History started two months prior to admission when the patient experienced hand weakness described as heavy, exemplified by difficulty holding a cup. Later, this was replaced by pinching finger joint pains, non-edematous and non- erythematous in appearance, associated with severe gnawing lower back pain. Initial consult revealed normal complete blood count (CBC), blood urea nitrogen (BUN), and creatinine levels with negative rheumatoid factor but with high uric acid. Pyuria and albuminuria were detected in a slightly turbid urinalysis specimen. She was managed as a case of presumptive urinary tract infection and was prescribed ciprofloxacin, etoricoxib, mecobalamin, and multivitamins. She developed puffy eyelids on the third day of treatment, thus discontinued intake of the aforementioned drugs. All symptoms resolved temporarily.

Seven weeks prior to admission, she started having severe, intermittent left periumbilical pain. Each episode lasted for 15 to 20 minutes. There was no distinct timing but it was associated with early satiety. No aggravating or alleviating factors were identified. Laboratory tests were performed revealing normal levels of BUN, total protein, albumin, and albumin/protein ratio but with hyponatremia, hypoalbuminemia, and low creatinine levels. CBC showed leukopenia. Midstream clean-catch urine detected pyuria. No medications were given. Another consult was done a few days later where a repeat urinalysis revealed proteinuria. The patient was prescribed ciprofloxacin and omeprazole but was noncomplaint.

Five weeks prior to admission, there was the persistence of recurring left periumbilical pain now accompanied by post-prandial, non-bilious, non-projectile vomiting, occurring 3 to 4 episodes per week with 3 to 4 bouts per episode, amounting to 300 to 400 cc per bout, and returning to baseline health in between episodes. She developed poor appetite, consuming only spoonfuls

of general liquid. Abdominal pain was less severe but more generalized. Finger joint pains also recurred. Consult was done and whole abdominal computed tomography (CT) scan with contrast was performed which was indicative of superior mesenteric artery (SMA) syndrome, further detailed as compression of the distal duodenum between the SMA and aorta. The aortomesenteric (AOM) angle was narrowed at 18° and the AOM distance was shortened to 2.6 mm. No mass and signs of obstruction seen. Subcutaneous edema on posterior and lateral pelvic regions and subcentimeter lymph nodes on both inguinal regions were also seen. Special home maneuvers were advised such as assuming a left lateral decubitus position after eating. Small frequent feeding was encouraged. Vomiting resolved temporarily but intermittent abdominal pain, early satiety, and finger joint pains persisted. The patient had lost a considerable amount of weight. Clothes fit more loose than before. Progression of persistent vomiting for 3 consecutive days, having two to three episodes per day, amounting to approximately 200 cc per bout, eventually prompted admission. The patient did not present with fever, cough, nasal catarrh, diarrhea, dysuria, polyuria, oliguria, hematuria, and constipation. All three pregnancy tests performed in this course of illness were negative.

She was infected with varicella, mumps, and measles in childhood but has never been hospitalized nor had any surgeries. She has no known food nor drug allergies. Birth and maternal, obstetrics- gynecologic, and family history were noncontributory. She belonged to a low- socioeconomic status family. Other than that, no psychosocial issues were identified. Patient admits that she is comfortable with her body image and has not made any attempts to lose weight. Meals are often shared with family for which she deems enjoyable. She reportedly received vaccines at the rural health unit until 1 year of age.

At the emergency department, the patient appeared cachectic, awake, and conversant but weak looking and assessed to be in hypovolemic shock with a palpatory systolic blood pressure of 120 mmHg, tachycardia at 120 beats per minute (bpm), and tachypnea at 30 cycles per minute (cpm). Physical examination revealed sunken eyes, pale palpebral conjunctiva, dry lips, and scaphoid abdomen with normoactive bowel sounds. The abdomen was soft but with direct tenderness on the epigastric area. No abdominal masses nor organomegaly were palpated. Palmar and plantar areas were pale. Extremities were equally cold with fair pulses and more than 3 seconds capillary refill time. No edema nor rashes were appreciated. Motor strength scored 3 out of 5 on all extremities. Reflexes were 2+ and symmetric at the biceps, triceps, knees, and ankles. Shock was reversed until hemodynamically stable. Anemia, hyponatremia, hypokalemia, and hypoalbuminemia were corrected accordingly. Urinalysis showed pyuria again, thus was treated as urinary tract infection with intravenous cefuroxime for 7 days.

Chest and abdominal X-rays were normal. Creatinine, BUN, ALT, AST, serum amylase and lipase, and thyroid hormones were within normal levels. Systemic lupus erythematosus (SLE) was considered due to the adolescent female demographic, joint pains, weight loss, proteinuria, albuminuria, leukopenia, and anemia. The antinuclear antibody (ANA) test was positive but the anti-double stranded DNA test was negative. The two-dimensional (2-D) echo was normal. Overall, rendering this differential diagnosis less likely.

Findings from the triphasic whole abdominal (WA) CT scan were consistent to SMA syndrome with nutcracker syndrome, showing a decrease in angulation of SMA and AA with an AOM angle of about 17° and a distance of about 0.3cm, causing a compression of the distal duodenal segment and left renal vein (Figure 1, 2, 3). Bilateral renal doppler ultrasound (UTZ) showed finings known for nutcracker phenomenon with no demonstrable collateral vessels (Figure 4).

Figure 1: Sagittal view WA CT scan with contrast. Aortomesenteric angle 17 ° and aortomesenteric distance 0.3cm.

Figure 2: Axial view WA CT scan showing decompression of duodenum.

Figure 3: Axial view WA CT scan depicting compression of left renal vein.

Figure 4: Renal Doppler UTZ.

CT angiogram of aorta and branches revealed narrowed AOM angle of 10° and distance of 0.3 to 0.4 cm with the left renal vein and 4th portion of the duodenum compressed. Suspicious nodular density in the right retrovesical area was noted (Figure 5).

Figure 5: CT angiogram.

Emesis and abdominal pain were controlled through gastric decompression. Proton pump inhibitors were given. The nutritional build-up was initiated via total parenteral nutrition (TPN) gradually shifting to NGT feedings until full oral feeding was achieved. A target caloric intake for ideal body mass index for age served as guide. Supplemental milk and f75 per meal aided this weight gain. Small, frequent feedings diet regimen was advised. Applying maneuvers that aid in the passage of food such as assuming the knee flexed towards the chest and left lateral decubitus positions were instructed. The patient was discharged with a 5% weight gain after 3 weeks in the hospital with subsequent resolution of the accompanying symptoms (early satiety and finger joint pains).

Discussion and Conclusion

Superior Mesenteric Artery (SMA) Syndrome results when the distal portion of the duodenum is compressed between the superior mesenteric artery and abdominal aorta (AA), leading to intestinal obstruction [1]. This syndrome was first reported by a Bohemian pathologist in 1861 as an autopsy finding [2,3]. Sixty years later, an

English surgeon named David Wilkie did an extensive study on 75 patients in 1927 and provided a more comprehensive description of this entity [2,3]. In general population, its prevalence rate ranges from 0.013 to 0.3% with higher incidence in female gender and adolescent age [2-4,6]. Its radiologic feature involves the measurement of aortomesenteric (AOM) angle and AOM distance. AOM angle is the angle formed between the proximal part of the SMA and the AA. It ranges from 38 to 65° in normal individuals but it is decreased at 10° in this patient as detected in CT angiogram [4]. AOM distance is the point from where the SMA branches out of the AA to the point where it meets the duodenum. It normally ranges from 10 to 34 mm but it is shortened to 3 to 4mm in this patient [5]. This anatomical abnormality occurs as a consequence to a reduced mesenteric fat pad, or fatty tissue buffer, that narrows the AO angle and shortens the AO distance, Thereby impinging the duodenum between the SMA and AA [4,6,7]. Any condition that disrupts this buffer could trigger the syndrome. Such conditions include rapid weight loss, immobilization or prolonged bed rest, post abdominal surgery, and exaggerated lumbar lordosis [1,8,9].

Nutcracker Phenomenon (NP), on the other hand, is a blood vessel entrapment of the left renal vein (LRV). The LRV compression is also due to the loss of the mesenteric fat pad buffer between the SMA and AA, resulting to narrowed AOM angle and shortened AOM distance. Due to variability of symptoms, its prevalence is unknown [5,7,10-12]. NP is differentiated from Nutcracker Syndrome in the absence of hematuria, proteinuria, and left flank pain [5,10]. Our patient was only reported to have NP.

For over a decade, only 3 cases of SMA Syndrome have been encountered in our pediatric tertiary hospital. None co- existed with Nutcracker Phenomenon [13]. The global prevalence of this syndrome coexistence is still lacking.

There has been no established criteria for diagnosing but from the few collected case reports of SMA syndrome coexisting with Nutcracker Phenomenon, patients have been properly diagnosed as soon as 1 month to as long as 3 years into the illness duration [7,10-12]. Manifestations include vague gastrointestinal symptoms namely nausea, abdominal bloating, early satiety, vomiting, left flank pain, and direct epigastric tenderness [7,10-12]. Some cases were not preceded by weight loss [12]. Data gathered posit that these rare vascular compression entities should be suspected in the context of gastrointestinal obstruction cases. However, it is worth noting, that the differential diagnosis for patients presenting with abdominal pain and vomiting is elaborate.

This patient's illness course started with hand weakness and finger joint pains followed by gastrointestinal symptoms as manifested by abdominal pain, early satiety and eventually intractable vomiting. Blood works have shown hypoalbuminemia, hyponatremia, hypokalemia, anemia, high uric acid, and low creatinine. Urinalysis detected pyuria, proteinuria, and albuminuria. After more than a year from diagnosis and hospital discharge, there has been no recurrence of the same symptoms based on the follow-up consult.

SMA syndrome coexisting with Nutcracker Phenomenon lacks consensus for criteria, subsequently posing diagnostic challenges. History and physical examination supported by imaging scans have shown integral role in management. Upper gastrointestinal series, computed tomography (CT), magnetic resonance (MR) angiography, and ultrasound are able to exhibit similar radiographic features and are used to confirm diagnosis. Prognosis is favorable but could be life threatening if the manifestation is severe and the treatment is inappropriate. Initial management is conservative, which includes gastric decompression and nutritional support. Lateral and prone positioning can be applied to shift the duodenum and left renal vein away from the obstructing structures and allow resumption of oral intake. Surgical correction to bypass obstruction can be done if medical management happens to be unsuccessful [4,5,7-9,11,12].

This syndrome is difficult to diagnose but not impossible to detect promptly. Sharing this disease course imparts awareness of such condition and promotes early detection, in hopes to ultimately reduce the accompanying morbidity rate.

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