

## CO-EXISTENCE OF SUPERIOR MESENTERIC ARTERY SYNDROME WITH NUTCRACKER SYNDROME – A DIAGNOSTIC DILEMMA

Vijaya Kamble <sup>1</sup>, Arpana Agrawal <sup>2</sup>, Saurav Bhagat <sup>3\*</sup>,  
Vishal Gupta <sup>4</sup> and Sarthak Jain <sup>5</sup>

<sup>1,4</sup> Professor, Department of Radiology, School of Medical Sciences and Research, Greater Noida, Uttar Pradesh, India.

<sup>2,5</sup> Post Graduate Resident, Department of Radiology, School of Medical Sciences and Research, Greater Noida, Uttar Pradesh, India.

<sup>3</sup> Assistant Professor, Department of Radiology, School of Medical Sciences and Research, Greater Noida, Uttar Pradesh, India.

Email: <sup>1</sup>vijaya.kamble@sharda.ac.in, <sup>2</sup>dr.arpana28@gmail.com,

<sup>3</sup>dr.sauravbhagat@gmail.com (\*Corresponding Author),

<sup>4</sup>vishal.gupta@sharda.ac.in, <sup>5</sup>sarthak.jain@sharda.ac.in

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### Abstract

The third section of the duodenum is compressed between the aorta and the superior mesenteric artery (SMA), which is also referred to as Wilkie syndrome or superior mesenteric artery syndrome. This is a very uncommon condition, with a frequency ranging from 0.13% to 0.3% and a preference for young women who are generally slim [1]. Weight loss, which reduces the fat plane that is located between the duodenum and the small bowel aorta [2], is the most common cause of this condition. There is a correlation between the clinical presentation and radiologic alterations, such as duodenal dilatation with the presence of an acute angle of takeoff of the SMA [3] which is used for diagnosis. Nutcracker syndrome (NCS) is characterised by its compression of the left renal vein (LRV) between the superior vena cava (SMA) and the aorta in an anterior manner, or, less frequently, between the aorta and the vertebral column in a posterior manner. **Case report:** 15-year-old boy that presented with pain abdomen since 6 days, associated with nausea and vomiting since 6 days. Physical examination and labs were unremarkable. CT revealed two renal veins, anterior and posterior to aorta with their compression confirming diagnosis of nutcracker syndrome. Additionally, the aortomesenteric distance, it was found to be 7 mm with aortomesenteric angle of 15 degrees, confirming the diagnosis of a co-existent SMA syndrome. **Discussion:** The mechanism of SMA is duodenal blockage, which results in postprandial abdominal pain that is alleviated by vomiting and an inability to accept oral intake. Two of the congenital anatomical alterations that have been proposed as potential causes to the development of the syndrome are a low takeoff of the SMA and a high insertion of the ligament of Treitz. It has been observed that the usual aorto-mesenteric angle is between 28 and 65 degrees, and the normal aorto-mesenteric distance is between 10 and 34 millimetres. The issue of diagnosing SMA syndrome has always been there due to the absence of a single test that is specifically designed for this condition. Similarly, for nutcracker syndrome, hallmark of NCS is the aggravation of symptoms with position, as standing upright causes the viscera to drop down thus pulling the SMA and aggravating the compression [17]. The diagnosis can be made by Doppler ultrasound, CT scan, or MRI where a dilated vein with a delayed washout and pelvic varicosities. Majority of cases are cured by the management, which focuses on nutritional support and weight regain strategies. **Conclusion:** SMA syndrome with nutcracker syndrome- in the presence of two left renal vein is a very rare occurrence, and makes the diagnosis challenging. Hence, a high incidence of suspicion is necessary to diagnose multiple anomalies on imaging.

### INTRODUCTION

The third section of the duodenum is compressed between the aorta and the superior mesenteric artery (SMA), which is also referred to as Wilkie syndrome. This condition is also known as superior mesenteric artery syndrome. This is a very uncommon condition, with a frequency ranging from 0.13% to 0.3% and a preference for young women who are generally slim [1].

Abdominal discomfort and vomiting are typical symptoms, with the latter experiencing a greater degree of severity after a meal. Weight loss, which reduces the fat plane that is located between the duodenum and the small bowel aorta [2], is the most common cause of this condition.

There is a correlation between the clinical presentation and radiologic alterations, such as duodenal dilatation with the presence of an acute angle of takeoff of the SMA [3]. This correlation is used to make the diagnosis. The majority of cases are cured by the management, which focuses on nutritional support and weight regain strategies.

Nutcracker syndrome (NCS) is characterised by its compression of the left renal vein (LRV) between the superior vena cava (SMA) and the aorta in an anterior manner, or, less frequently, between the aorta and the vertebral column in a posterior manner. Additionally, it is an extremely uncommon entity. It is typically characterised by pain in the flank, which may or may not be accompanied by hematuria, proteinuria, or renal insufficiency. There is typically a connection between the causes and anatomical variations [5].

It is possible to arrive at a diagnosis by analysing the link between the radiologic findings and the clinical presentation, either with or without the requirement of doing an invasive pressure measurement of the LRV [6]. In mild cases of non-cancerous sclerosis (NCS), the therapy of the condition might range from cautious waiting to a variety of reconstructive surgical procedures [7]. Although the literature provides a detailed description of both things, each of them is extremely uncommon.

Through this case report, we describe the diagnostic challenge we encountered in the diagnosis of a young male patient.

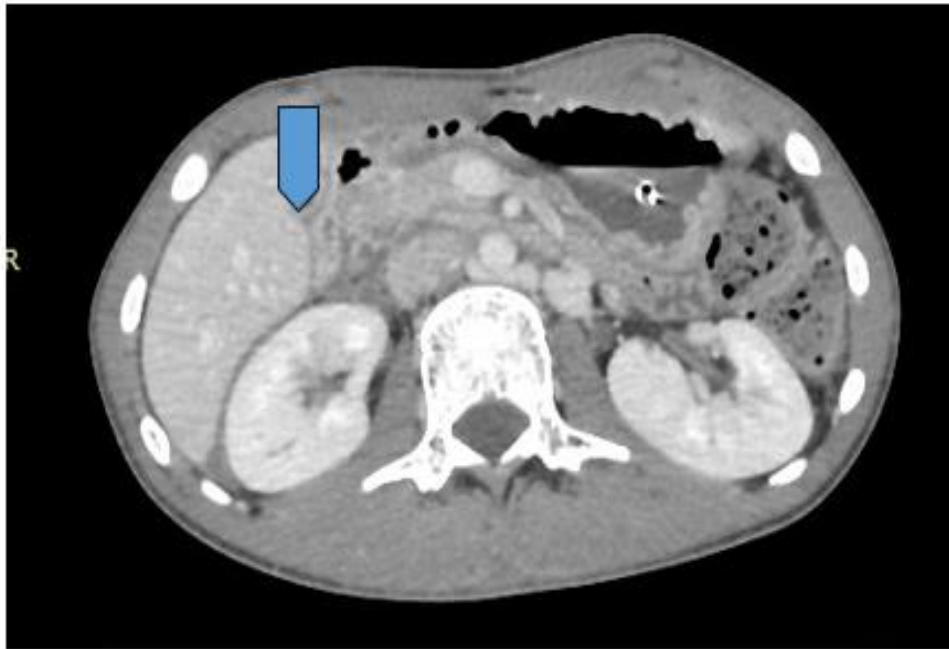
## **CASE DISCUSSION**

This is the case of a 15-year-old boy that presented with pain abdomen since 6 days, associated with nausea and vomiting since 6 days. The pain abdomen was colicky in nature, around the umbilical region and aggravated on eating food, with no relieving factors. The vomitus was bilious and non-projectile. Following this, he developed fever, weight loss and abdominal distension, for 3 days. He had regular bowel and bladder habits with no other symptoms.

On physical examination, there was no localized tenderness and the abdomen was soft. There was no bulging and there were no palpable masses with no evidence of hepatosplenomegaly. He was evaluated for possible causes of pain abdomen through a complete blood count, Renal function test, C-reactive protein, Liver function test; all within normal limits.

The ultrasound abdomen and pelvis was inconclusive, and hence a computed tomography was performed. Incidentally, it was found out that there are two left renal veins, with one of them anterior to the aorta and the other one being posterior to the aorta.

Both the left renal veins were found to be compressed; antero-aortic left renal vein between SMA and aorta while postero-aortic left renal vein between aorta and vertebral body. Hence a diagnosis of nutcracker syndrome- was made.



**Figure1: The axial section of CECT Whole abdomen demonstrating compression (marked by arrow head) of duodenum between aorta and Superior Mesenteric Artery**



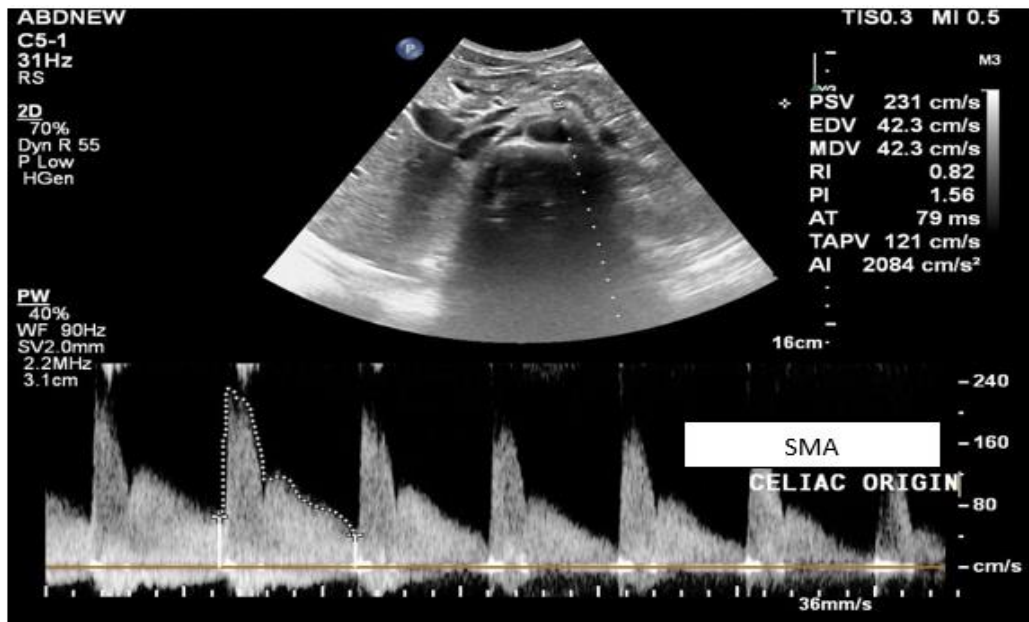
**Figure 2: The axial section of CECT Whole abdomen demonstrating compression (marked by arrow)of anterior renal vein between aorta and Superior Mesenteric Artery**



**Figure 3: The sagittal section of CECT Whole abdomen demonstrating aortomesenteric distance which is reduced & measures ~ 7 mm**



**Figure 4 :The sagittal section of CECT Whole abdomen demonstrating Aortomesenteric angle which is reduced & measures~ 20.7 degrees**



**Figure 5: The ultrasound doppler of Superior mesenteric artery demonstrates raised Peak Systolic Velocity at the compressed segment**

Additionally, when we measured the aortomesenteric distance, it was found to be 7 mm with aortomesenteric angle of 20.7 degrees, confirming the diagnosis of a co-existent SMA syndrome.

## DISCUSSION

There have been reports of an incidence ranging from 0.013 percent to 0.3 percent [1]. SMA syndrome is a rare condition. In most cases, the patient is an adolescent male who is considered to be skinny; nonetheless, this condition can manifest in people of any age or gender. The mechanism is duodenal blockage, which results in postprandial abdominal pain that is alleviated by vomiting and an inability to accept oral intake. In light of the fact that the duodenum is situated between the aorta and the SMA, one of the most common causes of the condition is weight loss. This diminishes the fat plane that is located between the two vessels, which in turn leads to compression, which in turn causes additional weight loss, so creating a vicious cycle. Two of the congenital anatomical alterations that have been proposed as potential causes or contributors to the development of the syndrome are a low takeoff of the SMA and a high insertion of the ligament of Treitz [2]. Both of these variations are characteristics of the disease. There are some authors who propose that conditions like burns, bariatric procedures, and cancers are the causes of the syndrome.

Similarly, for nutcracker syndrome, hallmark of NCS is the aggravation of symptoms with position, as standing upright causes the viscera to drop down thus pulling the SMA and aggravating the compression [17]. The diagnosis can be made by Doppler ultrasound, CT scan, or MRI where a dilated vein with a delayed washout and pelvic varicosities in the right clinical setting are highly suggestive. LRV phlebography is an invasive but conclusive method for confirming the elevated pressure in the LRV [6]. The issue of diagnosing SMA syndrome has always been there due to the absence of a single test that is specifically designed for this condition. It is possible for laboratory readings to exhibit nonspecific alterations concerning electrolyte imbalances and



weight loss, or it is even possible for laboratory values to be normal. A CT scan or an MRI may reveal abnormalities such as a dilated duodenum proximal to the SMA takeoff, a short aorto-mesenteric distance, or a narrow aorto-mesenteric takeoff angle. These abnormalities may be present in addition to other abnormalities. It has been observed that the usual aorto-mesenteric angle is between 28 and 65 degrees, and the normal aorto-mesenteric distance is between 10 and 34 millimetres [8]. An SMA syndrome patient will typically have a shorter distance and a narrower angle than the average patient. An average angle of 13.5 degrees and an average distance of 4.4 millimetres were discovered by a study that utilised multidetector computed tomography (MDCT) to analyse patients who were diagnosed with superior mesenteric artery syndrome (SMA). There is no definitive cutoff for diagnosis that can be determined only on the basis of imaging; nevertheless, some authors have proposed that in the appropriate clinical scenario, patients with angles that are fewer than 22 degrees and distances that are less than 8 mm are strongly suggestive of the diagnosis [3]. Magnetic resonance enterography (MRE), which is a mix of magnetic resonance imaging with dynamic PO contrast, was used by Guiseppe et al. to diagnose two patients. This was a novel technique that was recommended by the researchers. MRE has the advantage of displaying the anatomic variances while simultaneously revealing the mechanical repercussions that emerge from them, such as delayed transit and blockage [9].

## CONCLUSION

SMA syndrome with nutcracker syndrome- in the presence of two left renal vein is a very rare occurrence, and makes the diagnosis challenging. Hence, a high incidence of suspicion is necessary to diagnose multiple anomalies on imaging.

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