CASE REPORT

SUPERIOR MESENTERIC ARTERY SYNDROME COUPLED WITH RENAL NUTCRACKER SYNDROME

Sana Sharafat Ali, Shazad Hussain Waqar, Sajid Ali Shah, Isbah Rashid, Fatima Shahzad

Department of General Surgery, PIMS, Islamabad-Pakistan

An acute vascular angle between the superior mesenteric artery and the Aorta may compress the third part of the duodenum (SMA Syndrome) and the Left Renal Vein (Renal Nutcracker Syndrome). Usually, patients are young females with a lean stature. Symptoms of outlet obstruction and pelvic congestion syndrome may be present. Our patient presented with bilious vomiting and unintentional weight loss for the last 6–8 months. On further inquiry, she had dysmenorrhea and left flank pain associated with microscopic haematuria. Duodenojejunostomy with Left Renal Vein (LRV) re-positioning was performed. She recovered uneventfully and has improved intake with mild or no pain troubling her at 6 weeks of surgery.

Keywords: Dysmenorrhea; Haematuria; Renal Nutcracker Syndrome; Superior Mesenteric Artery Syndrome; Wilkies Syndrome; Vomiting

Citation: Ali SS, Waqar SH, Shah SA, Rashid I, Shahzad F. Superior Mesenteric Artery Syndrome coupled with Renal Nutcracker Syndrome. J Ayub Med Col Abbottabad 2024;36(1):221–5.

DOI: 10.55519/JAMC-01-12540

INTRODUCTION

The Superior Mesenteric Artery (SMA) originates from the anterior aspect of the abdominal aorta (AA), at the level of L1 and runs caudally to give rise to the Aortomesenteric angle (AMA). This creates a space which is occupied by the third part of the duodenum anteriorly and the LRV posteriorly (L3) and normally measures 45° (38–65°).[1] In cases when the SMA originates at a more acute angle (6–25°), the aortomesenteric distance reduces to 2-8mm (Normal range 10–28 mm).[1] The SMA may compress on the duodenum leading to features of outlet obstruction and is associated with weight loss. This is called the SMA Syndrome.[1]

The left renal vein may be compressed in this angle and lead to left flank pain, haematuria, orthostatic proteinuria (presence of protein in the urine while in an upright position but not when lying down) and features of pelvic congestion syndrome including dysmenorrhea, lower abdominal pain, dyspareunia, varicocele. This is called the Renal Nutcracker syndrome.[2] These two syndromes may rarely coexist.[3,4] SMA Syndrome is an unusual cause of vomiting and weight loss and is thus, difficult to diagnose. The Nutcracker syndrome is also an unusual cause of haematuria and other symptoms of the syndrome. The patient's history of multiple visits to health care professionals from different specialities is due to their young age and persistent spectrum of widespread symptoms. Both are regarded as diagnostic dilemmas and diagnoses of exclusion.

This more acute aortomesenteric angle is more common in weak debilitated body types and young females. It may be due to a recent rapid weight loss as it is linked with the loss of the fat pad between the SMA and aorta.[1] SMA syndrome in females with anorexia nervosa may lead to a vicious cycle of vomiting, decreased oral intake and weight loss leading to a more acute aortomesenteric angle. Psychiatric evaluation and nutritional counselling sessions remain an important part of the management of these patients. The cause may be like in our patient, from a congenitally acute angle which shows up with time, or from pathologies or surgeries of the spine, aorta or gut that may alter this angle as a whole.

Once the diagnosis is established by radiologic studies, first-line treatment is usually conservative with jejunal or parenteral nutrition to regain weight and subsequently increase the aortomesenteric fatty tissue. In case of failure of treatment, surgery is often required in both cases. For SMA syndrome, the standard surgical management remains a Laparoscopic Duodenojejunostomy.[5] Strong's procedure (duodenal mobilization and derotation) or Gastrojejunostomy may also be chosen.[5–7] In the case of the Nutcracker syndrome, transposition of the left renal vein remains the standard management plan.[8–10] We present a case of a young female who has bilious vomiting and unintentional weight loss for the last 6–8 months. Further, she had dysmenorrhea and left flank pain associated with microscopic haematuria.

Duodenojejunostomy with LRV re-positioning was performed. She recovered uneventfully and has improved intake with mild or no pain troubling her at 6 weeks of surgery.

CASE REPORT

A 16-year-old female with albinism and a slender physique had a long-term history of bilious vomiting which aggravated for the past few months. Vomiting occurred more after an hour and a half post-prandial. Her symptoms were associated with severe nausea, postprandial epigastric pain and early satiety. She felt better when fasting or overnight. There was a history of unintentional weight loss. She presented to the department of gynaecology of the hospital with complaints of dysmenorrhea and otherwise lower abdominal and back pain for the last few years. The symptoms have become more severe for the last two months. She also occasionally had left flank pain.

On examination, a lean lady with a BMI of 15.1 Kg/m^2, had a distended abdomen and a positive succession splash with no focal point of tenderness. Her bowel sounds were high-pitched. Digital rectal examination was normal. Baseline routine investigations showed a hypochloremic, hypokalemic metabolic alkalosis and hyponatremia. A urine routine examination revealed microscopic haematuria. She was managed via a nasogastric tube for decompression and intravenous fluid and electrolyte replacement.

Monitoring of her vitals, intake & output and her biochemistry was carried out regularly and management was accordingly tailored. Upper Gastrointestinal Endoscopy showed a dilated stomach full of bilious fluid. There was mild gastropathy with a few petechial haemorrhages. There was a normal-looking mucosa with an acute angle at the level of the second to the third part of the duodenum. There was no ulcer, varices, stricture or an intraluminal mass. CT scan Abdomen and Pelvis with intravenous contrast reported a narrow aorto-mesenteric angle of 5° and a short aorto-mesenteric distance of 5.5mm only. (Figure-1) There was a markedly dilated stomach and proximal duodenum, with an abrupt cut-off at the point where SMA crossed over the third part of the duodenum.

The third part of the duodenum was being compressed between the SMA and the Aorta. (Fig 2) The downstream small bowel was normal in calibre. The LRV coursed between SMA and the Aorta with its significant compression caused by the aortomesenteric angle. The LRV as well as the left ovarian vein demonstrated mild dilatation with a calibre of 8.6mm and 5.2mm respectively. (Figure-3) Bilateral parametrial vascular congestion was appreciated. (Figure-4) So, a diagnosis of SMA Syndrome with Renal Nutcracker Syndrome was made.

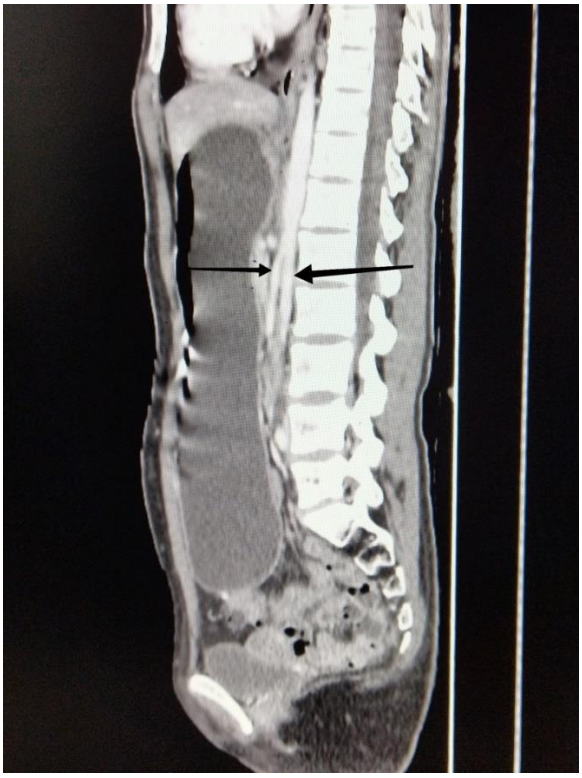

Figure-1: Sagittal view of CT scan of Abdomen and Pelvis showing an acute Aorto-mesenteric angle of 5° and a reduced Aorto-mesenteric distance of 5.5mm. SMA is marked with a short arrow, Aorta is marked with a long arrow.

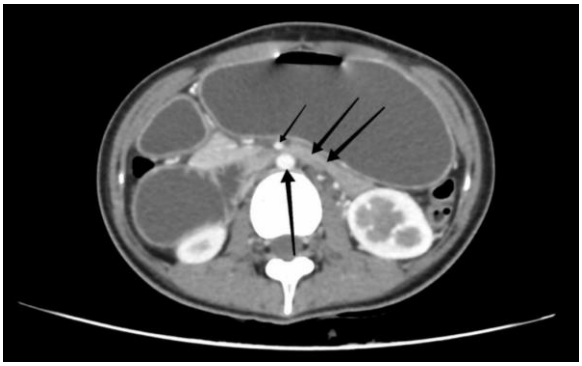

Figure-2: Axial view of CT scan Abdomen and Pelvis showing A short Aorto-mesenteric distance with SMA (short arrow) compressing the duodenum (two arrows). Dilated stomach and proximal duodenum are seen. Aorta is marked with a long arrow.

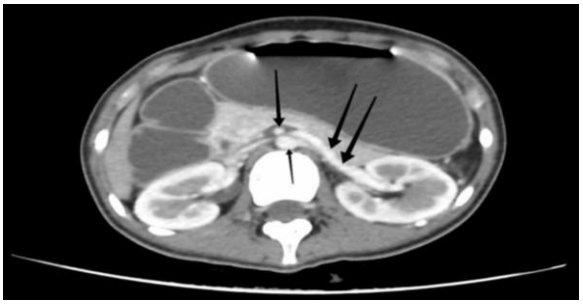

Figure-3: Axial view of CT scan Abdomen and Pelvis showing A short Aorto-mesenteric distance with SMA (long arrow) compressing the left renal vein which runs a tortuous and dilated course from the hilum of the left kidney (two arrows). Aorta is marked with a short arrow.

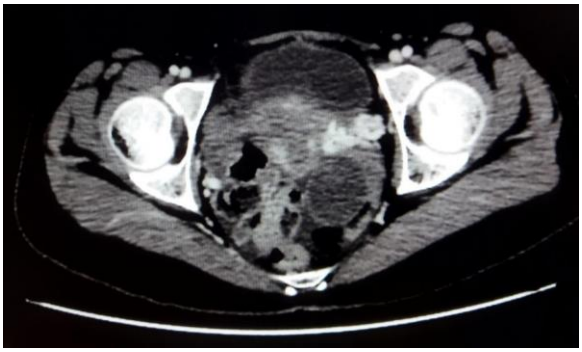

Figure-4: Axial view of CT scan pelvis showing bilateral parametrial vascular congestion.

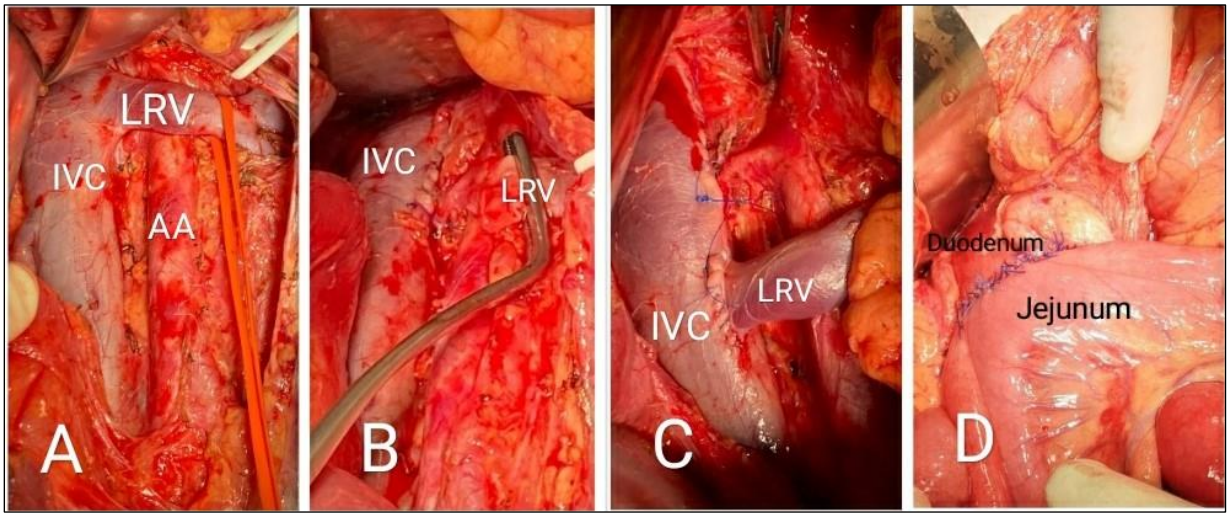

Figure-5: Preoperative pictures. (A) Patient's anatomy with a dilated Left Renal Vein (LRV) draining into the Inferior Vena cava (IVC) after crossing in front of the Abdominal Aorta (AAA), Superior Mesenteric Artery (SMA) has been retracted. (B) LRV excised from the IVC. (C) LRV re-anastomosed about 3cm distal to its original location on the IVC, away from the SMA. (D) The Stitch line of the Duodenojejunostomy.

An informed consent and counselling session was done regarding information about the prerequisites, the pros and cons, the nature of the surgery, the possible complications and the delay in recovery along with the chance of failure of complete resolution of symptoms. The patient also had her sessions of discussion with the psychiatrist and the nutritionist, who aimed to manage her perioperatively in the context of the syndromes. Her BMI was 25.5, albumin 2.9 with low vitamin D levels but calcium and iron levels were normal. Nutritional support was given for two weeks before surgery through both TPN and PPN. The patient visited multiple times to gastroenterologists and medical specialists before she presented to us. She didn't have depression or any eating disorder. The gastroenterologist had already got her psychiatrist consultation done.

An open midline approach was adopted for Laparotomy to perform a duodenojejunostomy with transposition of the LRV. Firstly, small and large guts were packed away to gain access to the retroperitoneum. Aorta, SMA, LRV and left Adrenal vein and Inferior Vena Cava (IVC) were delineated. The obstruction to drain for the LRV was evident by its dilated tortuous tense course. Injection heparin 5000IU was administered intravenously by the anaesthetist before partially clamping the IVC around the entry site of the LRV. A side-biting Satinsky clamp was used. LRV was excised with a small rim of the vena cava and IVC was continuously repaired with Prolene 5/0. The left adrenal vein and left lumbar vein were ligated to mobilize the LRV. LRV was then re-anastomosed with the Left lateral IVC 3cm caudally, using Prolene 5/0 parachuting continuously. This end-to-side anastomosis was free of rotation and was

tension-free. The jejunal loop was taken about 25 cm from the Duodenojejunal junction.

A Duodenojejunostomy, a standard bypass procedure for SMA syndrome was then performed with GIA 60 as a side-to-side anastomosis, to the right of the SMA. Neither the duodenum was mobilized nor the ligament of trietz was taken down. Jejunum taken 25 cm from the DJ and antecolic duodenojejunostomy made. The anastomosis was oversewed with Vicryl-2/0 interrupted stitches. (Figure-5) A pelvic drain was placed as there was intestinal anastomosis. The patient's post-operative recovery was uneventful. At six weeks of follow up she has started eating quite normally and is effectively not troubled by vomiting and pain that was earlier disturbing her daily life. She had gained about 6 Kg weight and her malnutrition started settling.

DISCUSSION

Both the SMA Syndrome and the Renal Nutcracker syndrome are well-recognized entities but are rarely seen together in a patient. Rotikansky described duodenal compression during an autopsy in 1842, and Wilkie used the term 'Chronic Duodenal Ileus' and first described SMA Syndrome in detail (1921), thus also known as Wilkie Syndrome.[1] Prevalence of SMA Syndrome has been 0.0024–0.3%. 75% of the diagnosed cases are females aged 10–30 years.[1] Patients earlier may present with epigastric pain and early satiety, and later with severe nausea, postprandial abdominal pain, bilious vomiting along weight loss. The prevalence of nutcracker syndrome is not exactly known due to its variety of overlapping symptoms but its diagnosis is being improved with improving technology and more cases are coming to light. Commonly diagnosed patients are females in the second or third decades of life.[2,8–10]

Nasojejunal feeding or feeding via a laparoscopically placed jejunostomy tube is the initial conservative management for SMA.[1] Electrolytes and fluid must be replaced. These should be coupled with help from a psychiatrist and a nutritionist to achieve optimal caloric replacement. The SMA is never surgically altered as a key principle in this management.[5] Surgical management in SMA syndrome is a Laparoscopic Duodenojejunostomy with a success rate of >90% or Strong's Procedure (Duodenal mobilization and derotation).[5] The open bypass procedures are Duodenojejunostomy, Gastrojejunostomy and duodenojejunostomy after the division of the fourth part of the duodenum.[5,6] For the Renal Nutcracker Syndrome, LRV Transposition is the standard procedure of choice, with other prominent options being endovascular and extravascular stents for the LRV, both procedures with their reservations.[2,8–11] Various treatment options have been tried for these patients over time aimed at reducing renal vein hypertension and pelvic venous reflux.[9]

In both of these syndromes, the patient may present acutely or more insidiously with progressive nonspecific symptoms. The presentation is linked to the degree of compression at the AMA.[1,2] Patients from either of these varieties would have had multiple visits to healthcare professionals from various departments, both elective and in the emergency. A clinician should have a high index of suspicion to look for the spectrum of symptoms occurring as a set in either of these. Both syndromes are often overlooked and may result in severe outcomes detrimental to the patient's health and life.[1,2] SMA syndrome may be confused with other causes of bowel obstruction, duodenal dysmotility or gastroparesis. A delay in management may lead to malnutrition, electrolyte imbalance, gastric perforation, pneumatosis and hypovolemia. A mortality rate of 33% has been reported.[1] Nutcracker syndrome was first described by Grant in 1937 equating LVR in the AMA with "a nut in the jaws of a nutcracker."[2] It is confused clinically, with other causes of haematuria and flank pain prioritized. There is special significance in patient education, practising the art of medicine and deciding on surgery when it is required when dealing with these diagnostic dilemmas.

Our patient presented similarly to various clinicians before she finally landed and was correctly diagnosed and treated. Surgery should indeed be reserved for patients with failure of medical management and should only be contemplated when necessary.

Conflicts of interest: The authors declare no conflicts of interest.

Consent: The patient and her parents have signed a consent for publication of the case report, which may be provided if asked for.

REFERENCES

1. Diab S, Hayek S. Combined Superior Mesenteric Artery Syndrome and Nutcracker Syndrome in a Young Patient: A Case Report and Review of the Literature. Am J Case Rep 2020;21:e922619.
2. Brogna B, Rocca AL, Giovanetti V, Ventola M, Bignardi E, Musto LA. An interesting presentation of a rare association of the Wilkie and Nutcracker syndromes. Radiol Case Rep 2023;18(8):2677–80.
3. Pacheco TBS, Chacon ACM, Brite J, Sohail AH, Gangwani MK, Malgor RD, *et al*. Nutcracker phenomenon secondary to superior mesenteric artery syndrome. J Surg Case Rep 2023;2023(1):rjac622.
4. Shi Y, Shi G, Li Z, Chen Y, Tang S, Huang W. Superior mesenteric artery syndrome coexists with Nutcracker syndrome in a female: a case report. BMC Gastroenterol 2019;19(1):15.
5. Yao SY, Mikami R, Mikami S. Minimally invasive surgery for superior mesenteric artery syndrome: a case report. World J Gastroenterol 2015;21(45):12970.

6. Pillay Y. Superior mesenteric artery syndrome: a case report of two surgical options, duodenal derotation and duodenojejunostomy. Case Rep Vasc Med 2016;2016:8301025.
7. Merrett ND, Wilson RB, Cosman P, Biankin AV. Superior mesenteric artery syndrome: diagnosis and treatment strategies. J Gastrointest Surg 2009;13(2):287–97.
8. Denchev B, Domuschieva E, Jelev G, Govedarski V, Zahariev T. Surgical treatment of a patient with nutcracker syndrome via transposition of the left renal vein. EJVES Short Rep 2018;41:10–2.
9. Kurklinsky AK, Rooke TW. Nutcracker phenomenon and nutcracker syndrome. Mayo Clin Proc 2010;85(6):552–9.
10. Wang L, Yi L, Yang L, Liu Z, Rao J, Liu L, Yang J. Diagnosis and surgical treatment of nutcracker syndrome: a single-center experience. Urology 2009;73(4):871–6.
11. Wang H, Guo YT, Jiao Y, He DL, Wu B, Yuan LJ, *et al*. A minimally invasive alternative for the treatment of nutcracker syndrome using individualized three-dimensional printed extravascular titanium stents. Chin Med J 2019;132(12):1454.

Submitted: October 4, 2023	*Revised: November 14, 2023*	*Accepted: December 21, 2023*

Address for Correspondence:

Prof. Shahzad Hussain Waqar, Department of General Surgery, PIMS, Islamabad-Pakistan
Cell: +92 333 513 1365
Email: waqardr@yahoo.com