

A RARE CASE OF SUPERIOR MESENTERIC ARTERY SYNDROME

Seetha Pramila V. V¹, Shankar², Karthik³, Anil Kumar Shukla⁴, R. Nagesh⁵

¹Professor, Department of Radiodiagnosis, Rajarajeswari Medical College, Bengaluru.

²Postgraduate Resident, Department of Radiodiagnosis, Rajarajeswari Medical College, Bengaluru.

³Postgraduate Resident, Department of Radiodiagnosis, Rajarajeswari Medical College, Bengaluru.

⁴Professor, Department of Radiodiagnosis, Rajarajeswari Medical College, Bengaluru.

⁵Professor, Department of Radiodiagnosis, Rajarajeswari Medical College, Bengaluru.

ABSTRACT

BACKGROUND

Superior Mesenteric Artery syndrome is a rare condition with an incidence of 0.013 – 0.3%. It causes obstruction at the third part of the duodenum due to compression between the Superior Mesenteric Artery and the aorta.

CASE REPORT

A 16-year-old male patient presented with complaints of vomiting, pain abdomen and loss of weight of six months' duration. Contrast CT was done and the patient was diagnosed to have Superior Mesenteric Artery syndrome.

DISCUSSION

CT is the modality of choice to diagnose SMA Syndrome. CT angiography is preferred and the two cardinal signs to look for are – AMA (Aortomesenteric Angle) and AMD (Aortomesenteric Distance).

CONCLUSION

CT is the gold standard in diagnosing Superior Mesenteric Artery Syndrome and eliminating other causes of intestinal obstruction.

KEYWORDS

SMA Syndrome, Wilkie's Syndrome, Cast Syndrome, Aortomesenteric Angle (AMA), Aortomesenteric Distance (AMD).

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BACKGROUND

Superior Mesenteric Artery (SMA) Syndrome which is synonymous with Wilkie's Syndrome, Cast Syndrome is a rare condition with an incidence of 0.013 – 0.3%.¹ It is caused due to obstruction of third part of duodenum as a result of reduced angle between the superior mesenteric artery and aorta.² Normal Aortomesenteric Angle (AMA) is 38 – 65 degrees.³ Barium studies of upper gastrointestinal tract, CT, Conventional Angiography, CT Angiography and MR Angiography are useful tools in the diagnosis of SMA syndrome.⁴

We are presenting a case of SMA Syndrome where a young male patient presented with complaints of vomiting, pain abdomen and loss of weight of six months' duration. Patient was referred to the Department of Radiodiagnosis for Contrast CT of the abdomen to find out the level of intestinal obstruction and the cause for the same.

CASE REPORT

A 16-year-old male patient presented with complaints of vomiting, dull aching epigastric pain and loss of weight of six months' duration. Patient had 1-2 episodes of vomiting per day

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Corresponding Author:

*Dr. Seetha Pramila V. V,
#728, Panchavati, 23rd Cross,
Ideal Homes Township,
Raja-Rajeswari Nagar,
Bengaluru-560098.*

E-mail: varadaseetha@gmail.com



which was not blood tinged, non-projectile and non-bilious. The vomitus contained food particles. Patient also complained of dull aching epigastric pain which was insidious in onset, non-radiating & with no significant aggravating or relieving factors. Patient also lost 8 kilograms of weight in the past six months.

Plain radiograph of erect abdomen showed few prominent bowel loops and barium meal study showed dilated first and second parts of duodenum with delayed transit through the third part. Patient was referred to our department for CT Abdomen. CT scan was performed on a 128-slice Siemens Somatom scanner after oral & IV contrast. Post-processing was done.

Axial CT scan images revealed dilated proximal part of duodenum [Figure – 1] with compression of third part of duodenum between the aorta and the superior mesenteric artery. CT angiography [Figure - 2] was performed with iodinated contrast and two cardinal signs were looked for – AMA (Aortomesenteric Angle) and AMD (Aortomesenteric Distance).⁵ Aortomesenteric Angle (AMA) was found to be 10 degrees [Figure – 3,4] and the Aortomesenteric Distance (AMD) was found to be 5 mm.

Patient was managed conservatively with nasogastric tube to decompress the stomach and proximal duodenum. Parenteral nutrition was given. Patient was counselled about the importance of lying down in the left lateral decubitus position. Patient was relieved of his symptoms and discharged.

The patient presented again with similar complaints three weeks later. Patient underwent open duodeno-jejunostomy and imaging findings were confirmed per operatively. Patient has been doing well post-surgery.



Figure 1. Axial CT images showing dilated stomach & 1st part of duodenum



Figure 2. CT sagittal recon image showing reduced Aorto-mesenteric angle



Figure 3. CT Angiogram showing the Aorta & its branches

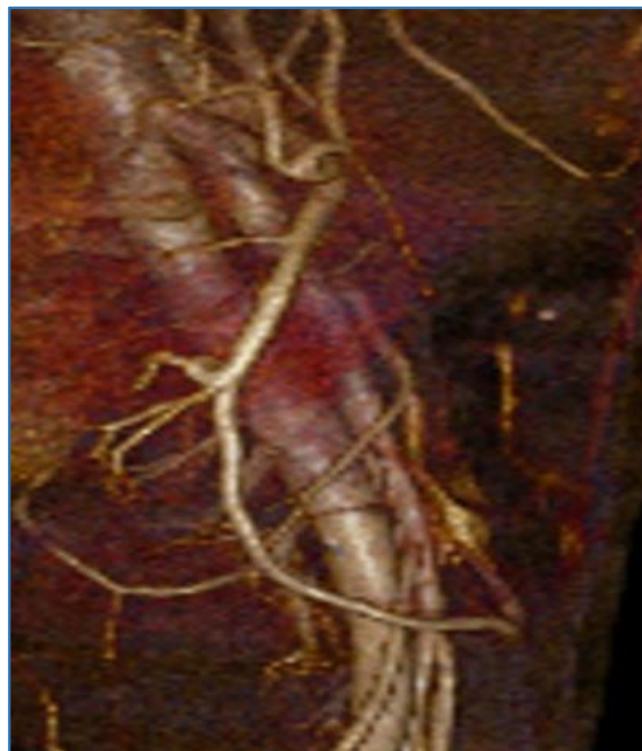


Figure 4. VRT image showing the Aorto-mesenteric angle

DISCUSSION

Anatomy & Pathogenesis: Superior Mesenteric Artery is the second of the three ventral visceral branches of the abdominal aorta. The origin of this artery is at the L1- L2 level. It courses anteriorly and inferiorly forming an angle of about 38 - 65 degrees with the abdominal aorta.^{3,5}

Duodenum which is the first of the three parts of small intestine has four parts, D1 - D4. The third part (D3) traverses between the aorta and the superior mesenteric artery at the level of L3 vertebral body. The other structures which traverse between these two vessels is the left renal vein, compression of which gives rise to a syndrome called the 'nutcracker syndrome'.⁵

The third part of the duodenum is usually cushioned by the retroperitoneal fat, thus preventing it from getting compressed between the superior mesenteric artery and aorta.⁶

The following Conditions can give rise to SMA Syndrome-

- A) Loss of retroperitoneal fat as a result of chronic weight loss due to anorexia nervosa, malabsorption, AIDS, malignancies, drug abuse and others.^{2,7}
- B) Post-operative cases in patients who have undergone corrective surgery for scoliosis- causes lengthening of spine which in turn increases the tension on mesentery.^{8,9}
- C) In individuals who have high insertion of ligament of Treitz, low origin of SMA.¹⁰
- D) Application of external body cast such as hip spica cast.¹¹

Demographics: The incidence of SMA syndrome is found to be 0.013 - 0.3% with a female preponderance.¹ The age group commonly diagnosed with SMA syndrome is between 10 and 39 years.^{12,13}

Clinical features: The patients may present with acute-on-chronic symptoms such as postprandial epigastric pain, nausea, vomiting, abdominal distension.^{2,14-16}

Chronic patients present with vague abdominal pain, weight loss which may be present from several years.^{2,14-16}

Exclusion of other causes such as tumours, strictures, pancreatitis should be done in order to arrive at the diagnosis of SMA syndrome.⁵

Radiologic findings: Diagnosis of SMA syndrome can be done by conventional methods like barium studies.

The Signs of Barium study which suggest SMA Syndrome are-

- A) Dilatation of D1 and D2 with dilatation of stomach
- B) Delayed gastroduodenal emptying.
- C) Anti-peristaltic flow of barium proximal to obstruction.
- D) Relief of obstruction on prone/left lateral decubitus position.^(17,18)

CT is the modality of choice to diagnose SMA syndrome.^{19,20}

CT angiography is preferred with iodinated contrast and two cardinal signs are looked for – AMA (Aortomesenteric Angle) and AMD (Aortomesenteric Distance)⁵

The normal AMA has been found to be 38 – 65 degrees. AMA <22 degrees is diagnosed as SMA.^{3,5}

The normal AMD is between 10 and 28 mm. In SMA syndrome, AMD < 8 mm is diagnostic.^{17, 21-22}

Recent study of four cases of SMA syndrome shows the mean AMA and AMD to be 13.5 degrees and 4.4 mm.¹⁹

A note should be made that the diagnosis of SMA Syndrome should merely not be made in cases with decreased AMA and AMD with absence of clinical symptoms.⁵

Treatment: The initial line of management is conservative, especially in acute cases which includes nasogastric tube to decompress stomach and duodenum, electrolyte resuscitation, parenteral nutrition, postpyloric feeding, mobilisation of patient to prone and left lateral decubitus position and small liquid meals.³

If patient does not respond to conservative management, then surgical approach is the treatment opted for. Open duodenojejunostomy has been the operation of choice traditionally, laparoscopic duodenojejunostomy has also been found to be safe and effective with a 100% success rate.^{6,23}

Another minimally invasive procedure used in SMA syndrome is Strong's procedure which includes lysis of ligament of Treitz with mobilisation of duodenum.²

CONCLUSION

Contrast-enhanced CT Abdomen with 3D reconstruction is the choice for diagnosing SMA Syndrome. It is a non-invasive investigative modality and diagnosis of SMA can be made when other causes of duodenal obstruction have been ruled out. Diagnosis of SMA syndrome cannot be made when no clinical symptoms are present.

REFERENCES

- [1] Rokitansky C. Handbuch der pathologischen Anatomie, Vol. 3. 1st edn. Wien, Austria: Braunmuller & Seidel 1842.
- [2] Merrett ND, Wilson RB, Cosman P, et al. Superior mesenteric artery syndrome: diagnosis and treatment strategies. *J Gastrointest Surg* 2009;13(2):287-92.
- [3] Zaraket V, Deeb L. Wilkie's syndrome or superior mesenteric artery syndrome: fact or fantasy? *Case Rep Gastroenterol* 2015;9(2):194-9.
- [4] Ünal B, Aktas A, Kemal G, et al. Superior mesenteric artery syndrome: CT and ultrasonography findings. *Diagn Interv Radiol* 2005;11(2):90-5.
- [5] Lamba R, Tanner DT, Sekhon S, et al. Multidetector CT of vascular compression syndromes in the abdomen and pelvis. *Radiographics* 2014;34(1):93-115.
- [6] Fong JK, Poh AC, Tan AG, et al. Imaging findings and clinical features of abdominal vascular compression syndromes. *AJR* 2014;203(1):29-36.
- [7] Welsch T, Büchler MW, Kienle P. Recalling superior mesenteric artery syndrome. *Dig Surg* 2007;24(3):149-56.
- [8] Sapkas G, O'Brien JP. Vascular compression of the duodenum (cast syndrome) associated with the treatment of spinal deformities: a report of six cases. *Arch Orthop Trauma Surg* 1981;98(1):7-11.
- [9] Griffiths GJ, Whitehouse GH. Radiological features of vascular compression of the duodenum occurring as a complication of the treatment of scoliosis (the cast syndrome). *Clin Radiol* 1978;29(1):77-83.
- [10] Strong EK. Mechanics of arteriomesenteric duodenal obstruction and direct surgical attack upon etiology. *Ann Surg* 1958;148(5):725-30.
- [11] Hughes JP, McEntire JE, Setze TK. Cast syndrome: duodenal dilation or obstruction in a patient in a body cast, with review of the literature. *Arch Surg* 1974;108(2):230-2.
- [12] Lee TH, Lee JS, Jo Y, et al. Superior mesenteric artery syndrome: where do we stand today? *J Gastrointest Surg* 2012;16(12):2203-11.
- [13] Biank V, Werlin S. Superior mesenteric artery syndrome in children: a 20-year experience. *J Pediatr Gastroenterol Nutr* 2006;42(5):522-5.
- [14] Mandarry M, Zhao L, Zhang C, et al. A comprehensive review of superior mesenteric artery syndrome. *Eur Surg* 2010;42(5):229-36.
- [15] Ahmed AR, Taylor I. Superior mesenteric artery syndrome. *Postgrad Med J* 1997;73(866):776-8.
- [16] Tsirikos AI, Anakwe RE, Baker AD. Late presentation of superior mesenteric artery syndrome following scoliosis surgery: a case report. *J Med Case Rep* 2008;2:9.
- [17] Gustafsson L, Falk A, Lukes PJ, et al. Diagnosis and treatment of superior mesenteric artery syndrome. *Br J Surg* 1984;71(7):499-501.
- [18] Hines JR, Gore RM, Ballantyne GH. Superior mesenteric artery syndrome. Diagnostic criteria and therapeutic approaches. *Am J Surg* 1984;148(5):630-2.
- [19] Agrawal GA, Johnson PT, Fishman EK. Multidetector row CT of superior mesenteric artery syndrome. *J Clin Gastroenterol* 2007;41(1):62-5.
- [20] Konen E, Amitai M, Apter S, et al. CT angiography of superior mesenteric artery syndrome. *Am J Roentgenol* 1998;171(5):1279-81.
- [21] Mansberger AR, Hearn JB, Byers RM, et al. Vascular compression of the duodenum. Emphasis on accurate diagnosis. *Am J Surg* 1968;115(1):89-96.

[22] Lukes PJ, Rolny P, Nilson AE, et al. Diagnostic value of hypotonic duodenography in superior mesenteric artery syndrome. *Acta Chir Scand* 1978;144(1):39-43.

[23] Munene G, Knab M, Parag B. Laparoscopic duodenojejunostomy for superior mesenteric artery syndrome. *Am Surg* 2010;76(3):321-4.