

Superior mesenteric artery syndrome. An often overlooked cause of abdominal pain!

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ABSTRACT

Objectives: To determine the prevalence of superior mesenteric artery (SMA) syndrome in patients presenting with abdominal pains, and to evaluate computed tomographic (CT) findings needed for its diagnosis.

Methods: This retrospective record-based study was carried out at the radiology department, from January 2016 to January 2021. All young patients (aged under 25) who underwent CT scans for abdominal pains were reviewed. Post-surgery, tumor, and trauma cases were excluded. Imaging findings for SMA syndrome were recorded as 'suggestive' (reduced aortomesenteric angle and distance with proximal duodenal dilatation), 'possible' (reduced angle and distance without proximal duodenal dilatation) and 'probable' (reduction of either angle or distance). Two radiologists interpreted the findings and consensus reporting was made. Diagnoses were confirmed on clinical grounds (symptomatic improvement by specific treatment and exclusion of other diagnoses), or barium studies. Imaging findings were compared to final diagnoses. McNemar's Chi-square test was used to determine association.

Results: Out of 141 patients (mean age=10.8, standard deviation=4), 7 (4.9%) patients mostly females were having SMA syndromes based on 'suggestive' imaging criteria ($p=0.0005$), and one patient underwent surgery.

Conclusion: Superior mesenteric artery syndrome is not an uncommon condition and should be considered in differential diagnosis of acute abdomen in young patients after excluding other diagnoses.

Keywords: SMA syndrome, computed tomography, aortomesenteric angle

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Superior mesenteric artery (SMA) syndrome, also known as Wilkie syndrome, cast syndrome or aortomesenteric duodenal compression syndrome, is an acquired vascular compression disorder in which acute angulation of the SMA results in compression of the third part of the duodenum leading to signs of obstruction or recurrent post-prandial pain.¹ Upper gastrointestinal fluoroscopy can demonstrate dilatation of the first and second part of the duodenum, extrinsic compression of the third part of the duodenum, and a relatively collapsed small bowel distal to point where the SMA crosses the duodenum.² Computed tomography (CT) enables clear visualization of vascular compression of the third horizontal part of duodenum by the SMA and also helps in measuring aortomesenteric angle and distance.^{1,3} Normally, the aortomesenteric angle and aortomesenteric distance measure 28-65° and 10-34 mm; in SMA syndrome, both parameters are reduced with values of 6° to 22° and 2 to 8 mm.⁴ Reduced fat between the SMA and aorta either due to poor nutritional intake or weight loss results in reduced aortomesenteric angle and distance.^{2,4}

Most of the patients diagnosed with SMA syndromes are managed conservatively, with surgery reserved for refractory cases not responding to medical treatment. Superior mesenteric artery syndrome is often an over-looked entity, and diagnosis can be delayed or even missed if not clinically suspected or known by the attending physician.⁵ Therefore, we sought to determine the prevalence of SMA syndrome in young patients presenting with acute abdominal pains and to evaluate specific CT imaging findings that could help in establishing this diagnosis.

Methods. This retrospective record-based study was carried out in the Radiology Department, King Fahad Military Medical Comple, Dhahran, Saudi Arabia from January 2016 to 2021. Clinical information and imaging of all children and young adults (aged under 25 years) presented with abdominal pains and underwent CT scans were reviewed. Post-surgery, tumor, and trauma cases were excluded. Research protocol was approved from the Institutional Review Board and need for informed written consents for the study was waived off considering retrospective nature of the study, and non-disclosure of patients' information. All information was kept strictly confidential. The research was conducted in accordance with the Helsinki Declaration.

Computed tomography images were reviewed for aortomesenteric angle (measured on mid sagittal image between abdominal aorta and SMA), aortomesenteric distance (measured on axial image at

level of horizontal third part of duodenum between abdominal aorta and SMA), and proximal duodenal dilatation. An aortomesenteric angle below 25° and aortomesenteric distance below 8 mm were considered positive for SMA syndrome. Duodenal distension (proximal to compressed third part) of more than 3 times the diameter of compressed duodenal segment was considered positive for SMA syndrome. Imaging findings in each case were classified as: i) 'Suggestive', if both angle and distance were found reduced along with proximal dilatation of duodenum; ii) 'Possible', if both angle and distance were found reduced without proximal duodenal dilatation; and iii) 'Probable', if either angle or distance was found reduced. Two radiologists interpreted the images and consensus was reached for final documentation of findings. Diagnoses were confirmed on clinical grounds (symptomatic improvement following specific treatment and exclusion of other diagnoses), or confirmation of obstructive findings on subsequent barium studies. Demographic information (age and gender), and imaging findings were compared to final diagnoses. The McNemar's Chi-square test was used to determine association.

Results. Out of 141 young patients with mean age of 10.8 years (standard deviation=4.0), most of these were males (n=81, 57.45%). Seven (4.9%) patients were found to have SMA syndromes based on our 'suggestive' (3/3) imaging criteria ($p=0.0005$) (Figure 1) and confirmed on subsequent barium studies (Table 1).

Twenty-one patients (14.8%) had normal scans. One hundred and thirteen (80.1%) had other diagnoses including acute appendicitis (n=36), Crohn's disease (n=16), acute gastroenteritis (n=18), Meckel's diverticulitis (n=3), acute intussusception or bowel obstruction (n=5), colonic diverticulitis and non-specific colitis (n=3), hernia-related bowel obstruction (n=11), acute urinary tract obstruction or infection (n=8), acute hepatitis (n=7), and acute cholecystitis (n=6).

All SMA patients had thin body habitus and reduced body mass indices (mean 16.1 ± 1.4). Most of these patients (5/7) were females, with mean aortomesenteric angle of 19 degrees and distance of 5.5 mm. Six of the SMA syndromes were treated medically (or conservatively) while one syndromic patient was treated surgically (derotation procedure) (Figure 2).

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Table 1 - TImaging criteria for SMA syndrome and clinical outcome.

Imaging criteria	NAD	OD	SMA syndrome
Suggestive	0	0	7
Possible	5	22	0
Probable	2	12	0
Not detected	14	79	0

NAD: no abnormality detected, OD: other diagnosis, SMA: superior mesenteric artery

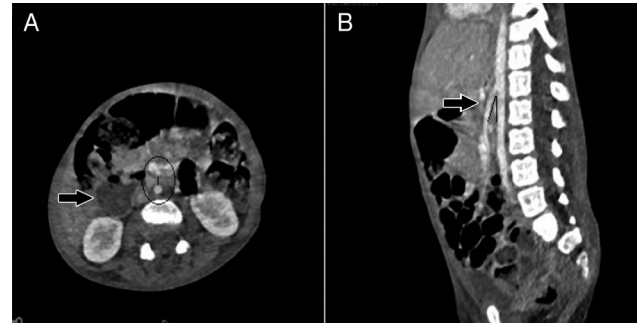


Figure 1 - Computed tomography criteria suggestive of SMA syndrome. Selected contrast enhanced A) mid abdominal axial and B) mid sagittal images demonstrating reduced aortomesenteric distance (small vertical line in region of interest in image A), dilated proximal second part of duodenum (arrow head in image A), and reduced aortomesenteric angle (arrow head in image B).

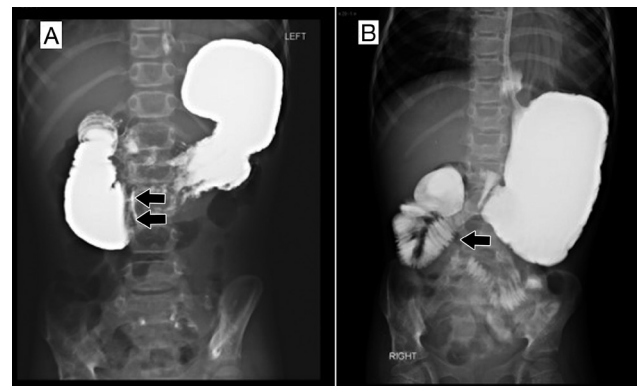


Figure 2 - Barium meal studies. Spot images of A) pre-operative study demonstrating curvilinear extrinsic lucency (arrow heads) corresponding to superior mesenteric artery impression upon the third part of duodenum, while B) post-operative (duodenal de-rotation) study showing unobstructed flow of barium through the third duodenal part (arrow head).

Discussion. Superior mesenteric artery syndrome has increasingly been recognized as a differential cause of abdominal pain particularly in young thin patients even though it remains a diagnosis of exclusion.⁶ This study demonstrates important diagnostic imaging criteria needed to identify SMA syndrome using CT scanning. All patients who met imaging criteria of reduced aortomesenteric angle, reduced aortomesenteric distance, and proximal duodenal dilatation, particularly when neither water nor contrast was provided orally before the CT examination, scored 3/3, with exclusion of other diagnoses, were found to have SMA syndromes. This has not been categorically emphasized in previous reports. Sinagra et al⁶ presented data of diagnosed SMA syndrome cases after suspicions of such compressions on upper gastrointestinal endoscopies in 2074 patients, and found both reduced aortomesenteric angles and distances to be significantly associated with SMA syndromes. They also observed that narrowed SMA distance was more accurate than narrowed angle for the diagnosis. Although we found 2/3 findings ('Possible' for SMA syndrome) in 2 of the diagnosed Crohn's disease (CD) patients, yet these findings were attributed secondary to malnourishment or weight loss primarily due to CD as acute flare-ups (terminal ileitis) were found on imaging at the time of presentations. Certain findings like weight loss can be attributed to recurrent episodes of vague abdominal pain particularly after intake of food (such as, post-prandial), however, most cases are diagnosed on acute or emergency presentations accompanied by more severe abdominal pain, nausea, vomiting and electrolyte imbalances.^{2,5} All 7 diagnosed SMA syndrome cases in our study presented in emergency for their acute abdominal pains. Recognition of such entity in differential diagnoses of abdominal pain is of importance particularly for the primary care physicians to detect these at earlier stages.

It is interesting to note that Nutcracker syndrome, another vascular compression syndrome, is also caused by the same etiology (such as, SMA) but a different affected structure such as, left renal vein.⁷ For this condition both the terms of syndrome (if symptomatic) and phenomenon (if asymptomatic) have been used in the literature.⁸ However, no such term as SMA phenomenon has been described to suggest similar finding of vascular compression in asymptomatic situation or an incident finding unrelated to patient's condition. We observed 2 of 7 SMA syndrome cases having the Nutcracker phenomena. Therefore, this condition or phenomenon should also be looked for and documented, considering similar location (such as,

aortic-SMA region) of both the affected structures (such as, duodenum and left renal vein) and compression by the similar structure (such as, SMA).

Most of the cases of this vasculo-anatomic syndrome in our study (6/7) were treated conservatively such as, by medical management (decompression of the stomach with a nasogastric tube and bypassing compressed duodenum by nasojejunal tube, correction of nutritional and electrolytes deficiencies by feeding tube or by total parenteral nutrition).⁵ When tolerated, oral feeding was resumed after stabilization of clinical condition of these patients and general vitality. Improved nutritional status helps to build up the fat cushion between the SMA and aorta and, hence, may help in reversing the situation. Posturing maneuvers during meals and motility agents may be helpful in some patients. Lying in the right decubitus position can relieve compression of the duodenum. Surgery may be considered if conservative treatment fails.⁹ Surgery (laparoscopic duodenal derotation) was performed in one patient of our study, and was also a syndromic child (Sanjad Sakati syndrome). Laparoscopic surgery involving lysis of the ligament of Treitz with the mobilization of the duodenum (derotation) is a minimally invasive approach and has proven effective.¹⁰

It is worth mentioning that the main pathophysiology in SMA syndrome is loss of fat pad between the SMA and aorta due to thin lean body habitus and reduced weight. We found all cases of SMA syndromes to have reduced body-mass indices depicting the same etiology behind this clinical entity. A variety of other conditions like chronic illnesses or infections, malignancies, trauma, eating disorders, substance abuse, spinal or bariatric surgeries can also result in reduced intraabdominal body fat due to weight loss resulting in SMA syndrome.^{11,12} Therefore, keeping suspicion of this condition in thin lean patients is of utmost clinical significance to timely diagnose this entity at an earlier stage without employing unnecessary imaging.

Study limitations. Our study included single-center, small sample size, and retrospective nature of study. Larger multicentric studies with stress upon both clinical and imaging predictive factors for the diagnosis and management of SMA syndrome are needed.

In conclusion, SMA syndrome is not an uncommon condition and should always be considered in differential diagnosis of acute abdomen in young lean patients after excluding other diagnoses.

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