EJCRIM
European Journal
of Case Reports
in Internal Medicine

ACHALASIA-ASSOCIATED SUPERIOR MESENTERIC ARTERY SYNDROME WITH MASSIVE GASTRIC DILATATION

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Received: 18/04/2025
Accepted: 28/04/2025
Published: 20/05/2025

Conflicts of Interests: The Authors declare that there are no competing interests.
Patient Consent: Written informed consent was obtained from the patient for the publication of his clinical data and accompanying images.
Acknowledgments: The team would like to thank the surgery department for great cooperation after the diagnosis was made.
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How to cite this article: Gashi Z, Shabani L, Shatrolli E. Achalasia-associated superior mesenteric artery syndrome with massive gastric dilatation. *EJCRIM* 2025 ;**12**:doi:10.12890/2025_005461

ABSTRACT

Background: Achalasia and superior mesenteric artery (SMA) syndrome are both uncommon gastrointestinal disorders. When they occur together—a rare clinical scenario—they can cause serious complications, including severe gastric distension.

Case Presentation: We report the case of a 62-year-old man with a known history of achalasia, previously managed with botulinum toxin injections. He presented with progressively worsening abdominal bloating, unintentional weight loss, and abdominal discomfort. Endoscopy revealed a strikingly dilated oesophagus and stomach, filled with nearly 9 litres of fluid. Further imaging with computed tomography angiography showed a narrowed aortomesenteric angle measuring just 16.7°, consistent with a diagnosis of SMA syndrome. Remarkably, despite the massive distension, no perforation was identified.

Discussion: This case highlights a rare but important overlap between achalasia and SMA syndrome, leading to extreme gastric dilatation in the absence of rupture. Despite the extreme degree of gastric dilatation observed on both endoscopy and imaging, the stomach wall remained intact, and there were no signs of ischemia or perforation. Although SMA syndrome may occur secondary to other health conditions, this case underscores how variable and potentially dangerous this dual pathology can be.

Conclusion: Clinicians should remain vigilant for the possibility of SMA syndrome in patients with achalasia who develop new or worsening upper gastrointestinal symptoms, particularly if there is significant gastric dilatation. Early imaging and recognition can be crucial in preventing severe outcomes like perforation.

KEYWORDS

Achalasia, SMA syndrome, massive gastric dilatation

LEARNING POINTS

- The patient has a very rare combination of achalasia and superior mesenteric artery syndrome, presenting with massive dilatation of the stomach with no perforation.
- Early detection through computed tomography angiography and early decompression are essential to prevent potentially fatal complications, including gastric rupture.
- This case adds to the limited literature describing achalasia-superior mesenteric artery overlap and reinforces the need for a high degree of clinical suspicion when symptoms diverge from baseline.

EFIM
European Federation of Internal Medicine

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INTRODUCTION

Achalasia is an uncommon oesophageal motility disorder defined by impaired relaxation of the lower oesophageal sphincter (LES) and loss of normal peristalsis in the oesophageal body[1]. Superior mesenteric artery (SMA) syndrome is another rare condition, caused by compression of the third portion of the duodenum between the aorta and the overlying superior mesenteric artery[2]. While both entities are well-described individually, their co-occurrence is exceptionally rare. We describe an unusual case of a patient with a history of achalasia who developed SMA syndrome, leading to extreme gastric dilatation and fluid retention, yet without gastric perforation.

CASE DESCRIPTION

A 62-year-old male, previously diagnosed with achalasia and treated with botulinum toxin injections 2 years earlier in another foreign centre, presented to our clinic with progressive abdominal distension, significant unintentional weight loss, and abdominal discomfort. He also reported worsening difficulty with oral intake, accompanied by a sensation of bloating and fullness.

On examination, his abdomen was markedly distended but soft, with no signs of peritonitis. Laboratory results were unremarkable except for mild electrolyte imbalances. An upper gastrointestinal endoscopy revealed a grossly dilated oesophagus filled with fluid and food residue, in keeping with his prior diagnosis of achalasia. The endoscope encountered resistance at the LES *(Fig. 1)*. After gentle passage into the stomach, a striking amount of retained fluid—estimated at around 9 litres—was suctioned *(Fig. 2)*. The major curvature extended approximately 55 cm beyond the cardia, suggesting significant gastric elongation. The duodenal bulb and the second portion of the duodenum also appeared dilated, although full visualization beyond this point was limited due to the length of the scope. Given the extent of the gastric dilation, a distal obstructive process was suspected. A contrast-enhanced computed tomography angiography (CTA) of the abdomen was performed, revealing a significantly narrowed aortomesenteric angle of approximately 16.7° *(Fig. 3)*, consistent with a diagnosis of SMA syndrome. The imaging also demonstrated compression of the third part of the duodenum, leading to proximal duodenal and gastric dilatation with gastric ptosis *(Fig. 4)*.

The patient was referred for surgical management to relieve the duodenal obstruction, as well as LES myotomy to prevent future recurrence or progression.

DISCUSSION

The simultaneous occurrence of achalasia and SMA syndrome is extremely uncommon. In this case, the patient's chronic weight loss—most likely due to long-standing achalasia—likely led to depletion of mesenteric fat, contributing to narrowing of the aortomesenteric angle and subsequent duodenal compression.

Despite the extreme degree of gastric dilatation observed

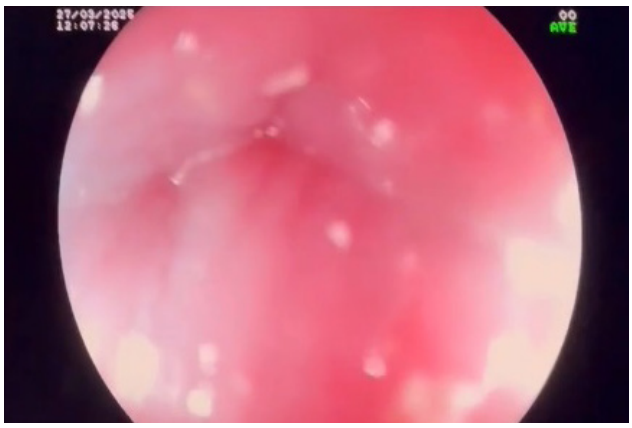


Figure 1. Tight lower oesophageal sphincter.



Figure 2. Bile content evacuated from the patient's stomach.



Figure 3. Narrowed aortomesenteric angle.

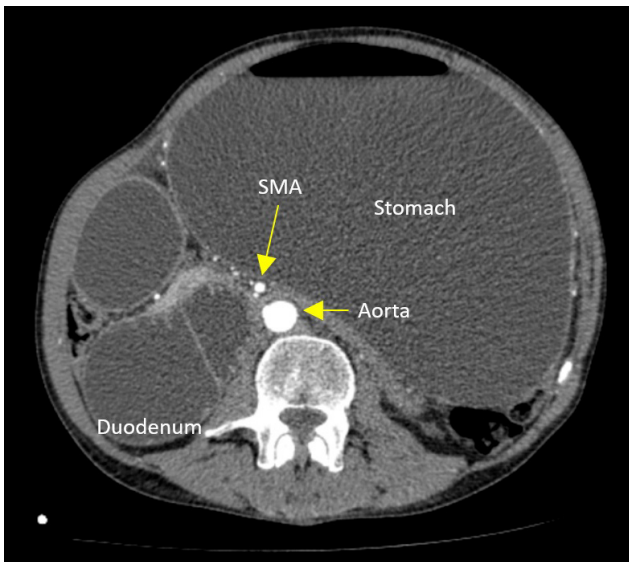


Figure 4. Significant gastric dilatation, as well as proximal duodenal dilatation with severe narrowing of the duodenum when passing in between the superior mesenteric artery (SMA) and the aorta.

on both endoscopy and imaging, the stomach wall remained intact, and there were no signs of ischemia or perforation. This contrasts with a case described by Kang and Williams[3], where a similar dual pathology resulted in gastric perforation. On the other hand, Sawano et al.[4] reported a case resembling ours, in which they reported the detailed dilation of the duodenum, stomach and oesophagus in a patient with significant weight loss, reinforcing the same overlapping pathophysiology of functional and mechanical obstruction. Moreover, Foster and Choudhary[5] described an adolescent with triple A syndrome whose delayed achalasia diagnosis led to severe malnutrition and subsequent SMA syndrome, highlighting the risk of vascular compression in malnourished achalasia patients. Although SMA syndrome may occur secondary to other underlying health conditions, a comprehensive review of the existing literature across multiple medical databases, including PubMed, Scopus, and Google Scholar, revealed that this appears to be only the third closely related case reported to date. The rarity of the case underscores the clinical significance of our findings and highlights the need for continued documentation and awareness of such atypical presentations.

These cases, including ours, highlight an important clinical consideration: in patients with achalasia who develop sudden or worsening obstructive symptoms associated with weight loss, SMA syndrome should be included in the differential—particularly if significant weight loss is present. Imaging studies, especially CTA, are essential for timely diagnosis.

CONCLUSION

This case emphasizes the need for heightened awareness of secondary causes of gastric outlet obstruction in patients with pre-existing motility disorders. Early imaging and recognition of SMA syndrome can prevent serious complications and guide appropriate intervention.

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