Med & Health Jan 2026; 21(1): 563-569 https://doi.org/10.17576/MH.2026.2101.39

CASE REPORT

Fatal Wilkie's Syndrome Complicated by Aspiration Pneumonia in an Adolescent with Autism: A Case Report

NURHUDA NADZRI[1], ISMAIL MOHD SAIBOON[1,2]*

[1]Department of Emergency Medicine, Faculty of Medicine, Universiti Kebangsaan Malaysia, 56000 Kuala Lumpur, Malaysia

[2]Hospital Canselor Tuanku Muhriz, Universiti Kebangsaan Malaysia, 56000 Kuala Lumpur, Malaysia.

Received: 24 September 2024 / Accepted: 24 October 2025

ABSTRAK

Sindrom arteri mesenteri superior (SMAS), atau sindrom Wilkie, merupakan punca penghalang duodenum yang jarang berlaku, di mana duodenum terhimpit antara arteri mesenteri superior dan aorta abdomen akibat kehilangan lapisan lemak mesenteri. Keadaan ini dikaitkan dengan kadar morbiditi dan kematian yang tinggi, terutamanya apabila berlaku kelewatan dalam diagnosis, yang boleh menyebabkan kepada komplikasi seperti perut bocor atau aspirasi radang paru-paru. Seorang kanak-kanak lelaki berusia 14 tahun dengan sejarah gangguan spektrum autism tahap tinggi, mengadu muntah dan sakit perut selama dua hari, yang menunjukkan penghalang outlet perut, kemudian menyebabkan kandungan laktat dalam darah yang tinggi dan asidosis metabolik. Tomografi berkomputer dengan peningkatan kontras menunjukkan pengembangan ketara pada esofagus bawah hingga ke segmen D3 duodenum, penyempitan sudut aortomesenterik, serta pemendekan jarak aortomesenteric, iaitu penemuan yang menunjukkan penyumbatan duodenum disebabkan SMAS. Walau bagaimanapun, pesakit kemudiannya telah muntah berterusan yang membawa kepada aspirasi radang paru-paru. Keadaan pesakit semakin merosot kepada kejutan sepsis dengan kegagalan pelbagai organ, yang akhirnya membawa kepada kematian. Laporan kes ini bertujuan untuk menyoroti cabaran dalam diagnosis dan rawatan SMAS terutamanya pada kanak-kanak austistik kerana mereka mungkin mempunyai kesukaran menyatakan kesakitan atau ketidakselesaan, di samping risiko kemerosotan yang cepat yang memerlukan penjagaan rapi yang segera.

Kata kunci: *Aspirasi radang paru-paru; gangguan spectrum autism; kesudahan maut; sindrom arteri mesenteri superior*

ABSTRACT

Superior mesenteric artery syndrome (SMAS), or Wilkie's syndrome, is a rare cause of duodenal obstruction, whereby the duodenum is compressed between the superior mesenteric artery (SMA) and abdominal aorta (AA) due to a loss of mesenteric fat pad. This condition is associated with high morbidity and mortality, particularly if the diagnosis is delayed, leading to complications such as gastric perforation or aspiration pneumonia. A 14-year-old boy with an underlying severe autism spectrum disorder, complained of a two-day history of abdominal pain and vomiting, suggestive of gastric outlet

Correspondence: Ismail Mohd Saiboon. Department of Emergency Medicine, Faculty of Medicine, Universiti Kebangsaan Malaysia, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Kuala Lumpur, Malaysia. Tel: +6019 3200297
E-mail: fadzmail69@yahoo.com.my

obstruction, leading to hyperlactatemia and metabolic acidosis. An immediate contrast-enhanced computerised tomography revealed a significant dilatation of the lower esophagus until the D3 segment of the duodenum, narrowing of the aortomesenteric angle, and shortening of the aortomesenteric distance, findings suggestively of duodenal obstruction secondary to SMAS. However, patient subsequently developed persistent vomiting leading to aspiration pneumonia. He rapidly deteriorated into septic shock with multiorgan failure, ultimately leading to his death. This case report aimed to highlight the challenges in diagnosing and managing SMAS particularly in autistic children as they may have difficulty communicating their distress, and there is a potential of rapid deterioration that requires prompt escalation of care.

Keyword: Aspiration pneumonia; autism spectrum disorder; fatal outcome; superior mesenteric artery syndrome

INTRODUCTION

Superior mesenteric artery syndrome (SMAS) was first described by Carl Von Rokitansky in 1842, and later was further elaborated by D. Wilkie in 1927, after which the eponym "Wilkie Syndrome" emerged (Guerra et al. 2023). SMAS is a rare but disabling condition with the reported incidence ranging from 0.013% to 0.78% (Guerra et al. 2023). It occurs when the third part of duodenum (D3) is compressed between the superior mesenteric artery (SMA) and the abdominal aorta (AA). This causes non-specific symptoms such as abdominal pain, bloating and vomiting, and can present as acutely or episodically in chronic cases. This condition is typically associated with rapid growth in children or rapid weight loss in adults (Salehi et al. 2022). Diagnosing SMAS can be challenging, as the clinical presentations are similar to other gastrointestinal diseases, often leading to misdiagnosis. Therefore, a high index of suspicion is necessary, and performing computerised tomography (CT) imaging is crucial for an accurate diagnosis. Despite its rarity, SMAS can lead to significant morbidity and mortality.

CASE REPORT

A 14-year-old Indian boy with severe autism spectrum disorder (ASD), which affects his ability to communicate verbally and nonverbally, presented to the Emergency Department (ED) with a two-day history of abdominal pain and abdominal distension. He had two episodes of non-bilious, post-prandial vomiting per day, no bowel movement, reduce oral intake and fever. There was no history of eating outside food or similar complaints among family members. There were no complaints of weight loss, anorexia, intermittent abdominal pain or vomiting in the past few months. He had no respiratory and urinary symptoms. His father was uncertain if the boy had experienced a seizure or any trauma.

Physical examination revealed a tall, thin and restless boy with tachypnoea (respiratory rate (RR) of 22 breaths/minute), oxygen saturation (SPO2) of 97%, elevated blood pressure (BP) of 154/98 mmHg, heart rate (HR) of 117 beats/minute (bpm) and temperature 37.9°C. His Glasgow coma scale (GCS) was E4V2M6 (12/15), similar as his baseline. Lungs' auscultation was equal air entry bilaterally without any crepitations. Heart sounds were normal. His abdomen was distended, generalised tenderness with no bowel sounds. Rectal examination found no abnormalities.

Initial blood tests showed leukocytosis, but renal function tests, liver function tests and other electrolytes were within normal limits. Detailed blood test results were listed in Table 1. The initial venous blood gas (VBG) results however revealed high anion gap metabolic acidosis with hyperlactatemia (Table 2). A bedside ultrasound showed a significantly distended stomach, without intraperitoneal free fluid. A chest radiograph showed an elevated left hemidiaphragm but no lung consolidation (Figure 1A). Abdominal radiograph suggested a proximal

TABLE 1: Initial blood investigations

Test	Results	Reference range
Hemoglobin (g/dL)	17.6	13.0-17.0
White cell count ($x10^9$/L)	25.4	4-10
Hematocrit (%)	51.4	45.7
Platelet ($x10^9$/L)	254	150-410
Urea (mmol/L)	4.8	3.2-7.4
Creatinine (micromole/L)	71.8	64-104
CRP (mg/dl)	0.18	<0.5
APTT (seconds)	33.4	30.3-46.5
Blood glucose (mmol/L)	13.0	4.0-7.8
Sodium (mmol/L)	143	136-145
Potassium (mmol/L)	4.3	3.1-5.1
Albumin (g/L)	52	35-50
Corrected calcium (mmol/L)	2.39	2.2-2.6
Magnesium (mmol/L)	1.01	0.66-1.07
Phosphate (mmol/L)	1.63	0.74-1.52
Prothrombin time (seconds)	18.6	11.6-14.9
INR	1.37	0.8-1.1
Serum ketone (mmol/L)	0.2	<0.6

CRP: C-Reactive Protein; APTT: Activated partial thromboplastin time; INR: International normalised ratio

bowel obstruction (Figure 1B).

A provisional diagnosis of gastric outlet obstruction was made. Intravenous (IV) isotonic saline, IV fentanyl 50 mcg and IV pantoprazole 50 mg were immediately administered. A nasogastric tube was inserted to decompress the gastric cavity. He was referred to the surgical team. A contrast-enhanced computerised tomography (CECT) of the abdomen was performed, with oral contrast fluid administered through the nasogastric tube.

Following CECT, he experienced vomiting, sudden decline in consciousness (E2V2M5) and developed respiratory distress, requiring high-flow oxygen. Lung auscultation revealed fine crepitations. The arterial blood gas (ABG) indicated type 2 respiratory failure with worsening metabolic acidosis, as shown in Table 2. He was intubated for impending respiratory arrest secondary to aspiration pneumonia. Empirical antibiotics (IV Rocephin 1 g and IV metronidazole 500 mg) were initiated. A post-intubation chest radiograph revealed bilateral alveolar infiltrates suggestive of aspiration (Figure 1C).

The CECT findings revealed significant dilatation of the lower esophagus, stomach and duodenum with a transition point at the D3 segment of the duodenum (Figure 2), where the aortomesenteric angle (AMA) was narrowed (11.0°), and the aortomesenteric distance was 0.4 cm. These findings were suggestive of duodenal obstruction secondary to SMAS. No

TABLE 2: Serial blood gas results

Test	Reference range	Initial VBG result	Serial ABG result
pH	7.35-7.45	7.278	7.118
pO_2 (mmHg)	80-100	-	108
pCO_2 (mmHg)	35-45	37.4	64.4
Bicarbonate ion (mmol/L)	22-26	17.7	16.8
Serum lactate (mmol/L)	<2.0	7.9	9.2

VBG: venous blood gas; ABG: arterial blood gas

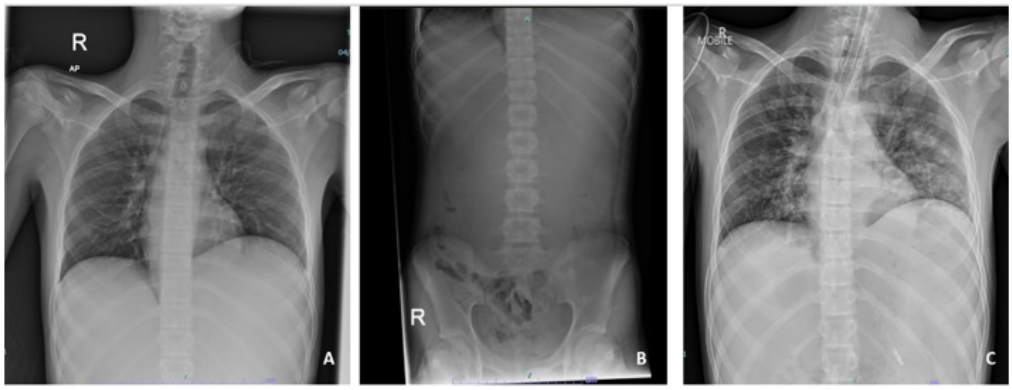

FIGURE 1: (A) Chest radiograph revealed an elevated left hemidiaphragm with absent of gastric bubble; (B) Abdominal radiograph revealed a paucity of small bowel gases that suggest proximal intestinal obstruction; (C) Chest radiograph taken post-intubation revealed bilateral alveolar opacities after the patient aspirated

obstructing mass or volvulus seen. An urgent oesophagoduodenoscopy (OGDS) revealed undigested food at the gastroesophageal junction extending to the body of the stomach, along with multiple Forrest III ulcers (Figure 3).

He was diagnosed with gastric outlet obstruction secondary to SMAS with aspiration pneumonia complicated by adult respiratory distress syndrome, metabolic acidosis and concurrent acute kidney injury (AKI). He was admitted to the intensive care unit (ICU), but succumbed to septic shock with multiorgan failure after two days.

DISCUSSION

Wilkie's Syndrome or SMAS arises as a result of loss of mesenteric fat pad between the AA and

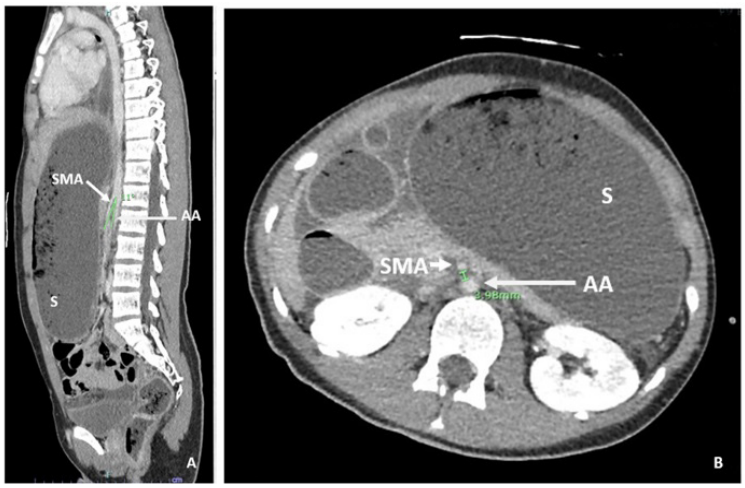

FIGURE 2: The abdominal CT scan showing SMA syndrome. (A) Sagittal view revealed prominent distension of stomach that reaches the pelvis. The aortomesenteric angle between the abdominal aorta (AA) and the superior mesenteric artery (SMA) was 11°; (B) The axial view showing a reduced aortomesenteric distance of 3.98 mm, compressing the third part of duodenum

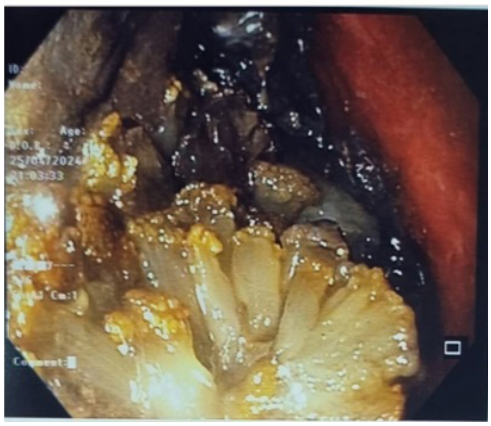

FIGURE 3: Oesophagoduodenoscopy showed stomach was filled with undigested food particles at gastroesophageal junction

SMA, which leads to a narrower AMA of less than 25° (normally ranges from 38° to 56°) and a shorter aortomesenteric distance of less than 10 mm (normally ranges about 10 to 20 mm) (Navandhar et al. 2024). This causes external compression of the D3 between the SMA and the AA (Guerra et al. 2023). This was the issue in our patient, where the AMA and aortomesenteric distance were markedly reduced.

Any condition that reduces the fat cushioning effect can lead to SMAS (Guerra et al. 2023). Acquired factors such as rapid growth in children, rapid weight loss due to chronic illness in adults, dietary disorders, substance abuse, immunosuppressed and malnutrition were reported as common causes (Salehi et al. 2022; Shi et al. 2019). SMAS can also occur after prolonged bed rest or post spinal surgery, or local pathology such as tumours (Payus et al. 2019). Anatomical variants too can cause SMAS such as congenitally low origin of the SMA, high insertion of the ligament of Treitz at the duodenojejunal flexure or a short mesenteric root (Navandhar et al. 2024). Nevertheless, SMAS is uncommon in healthy individuals (Mohammad Kazmin et al. 2020), but it is more prevalent in adolescents and young adults aged 10 and 39 years, and often found in females (Ganss et al. 2019; Mohammad Kazmin et al. 2020).

Diagnosing SMAS is exceedingly challenging as patients can present with a broad spectrum of symptoms that are non-specific, either acute or chronic, depending on the etiology. It is often a diagnosis of exclusion, requiring a thorough evaluation and investigations for other more common diagnoses such as gastroenteritis, peptic ulcer diseases , pancreatitis, cholecystitis, mesenteric ischemia and inflammatory intestinal disorders (Mohammad Kazmin et al. 2020; Navandhar et al. 2024). Patients with chronic presentation usually have a history of episodic, prolonged, vague abdominal pain with vomiting and epigastric bloating (Danushka et al. 2023). Hence, emergency physicians (EPs) should consider SMAS as a potential diagnosis in patients presenting with clinical features of bowel obstruction to ensure timely and appropriate management.

Our patient was a male adolescent with severe ASD, which made it more challenging to communicate effectively his symptoms and to diagnose his condition. Similar challenges have been reported by Riera and Phalen (2013), who found that approximately 80% of children with ASD have an altered pain sensorium, often described as indifference to pain, and about 80% of them also have features of asthenic body habitus (Salehi et al. 2022). Although rare, our patient presented acutely with clinical features suggestive of gastric outlet obstruction, but his body habitus was normal. Therefore, it is important for EPs to be attentive when treating patients with intellectual and developmental disabilities.

In this case, high suspicion of gastric outlet obstruction from the abdominal radiograph prompted the physicians in the ED to get an urgent CECT, as it is a crucial diagnostic tool to evaluate small-bowel obstruction (Rajput et al. 2023), and a gold standard to diagnose SMAS (Mohammad Kazmin et al. 2020). Unfortunately, he aspirated after given the oral contrast and further deteriorated. Some studies suggest that the use of oral contrast media is still controversial in cases of suspected mechanical small bowel obstruction and should be avoided in patients

with high risk for aspiration (Pickhardt 2020). In this particular case, the oral contrast agent should be excluded. Further investigation using an OGDS helps to rule out intraluminal causes of obstruction (Mohammad Kazmin et al. 2020).

Our patient presented with high anion gap metabolic acidosis and hyperlactatemia. Acute gastric obstruction causes gastric dilatation, increasing intragastric pressure and leading to bowel hypoperfusion and ischemia (Ugras et al. 2017). Mohammad Kazmin et al. (2020) explain that ischemic tissues produce lactic acid due to anaerobic metabolism, contributing to metabolic acidosis. They also state that a massively distended stomach increases abdominal compartment pressure, reducing renal artery perfusion and venous drainage, which can cause AKI and worsen metabolic acidosis due to decreased bicarbonate reabsorption. Additionally, the raised abdominal compartment pressure compresses the inferior vena cava, impairing venous return and further contributing to hypotension (Ugras et al. 2017). Recurrent vomiting from the obstruction leads to loss of gastric content, hypovolemia, electrolyte imbalance and potentially causing metabolic alkalosis (Mohammad Kazmin et al. 2020) .

Our patient received initial conservative medical management such as IV hydration, gastric decompression through nasogastric tube insertion, and early initiation of broad-spectrum antibiotics to treat aspiration pneumonia. Additionally, nutritional support through naso-jejunal feeding or total parenteral nutrition (TPN) is vital to alleviate the obstruction by increasing the mesenteric fat cushion, thereby increasing the AMA (Salehi et al. 2022). Some patients may require surgical interventions such as lysis of ligament of Treitz, gastrojejunostomy, duodenojejunostomy or enteral tube placement (Akel et al. 2024).

SMAS has a high mortality rate of 33% due to delays in diagnosis (Danushka et al. 2023). Gastric perforation and hemorrhage, electrolyte abnormalities, AKI, and aspiration pneumonia are the common causes of death associated with SMAS (Guerra et al. 2023). Early diagnosis and treatment are critical for a favourable prognosis (Mohammad Kazmin et al. 2020). Despite receiving appropriate initial management for duodenal obstruction and septic shock, he unfortunately deteriorated rapidly after developing aspiration pneumonia, and ultimately succumbed after 2 days in the ICU. Perhaps, a more vigilant in anticipating and managing the potential complications associated with SMAS could lead to a different outcome.

CONCLUSION

This case highlights the challenges and awareness in managing nonverbal ASD patients with rare conditions like SMAS. Despite its rarity, SMAS should be considered in young adults presenting with upper intestinal obstruction. Early abdominal CECT scans are essential for diagnosing SMAS and ruling out other potential causes. However, the use of oral contrast should be extra cautiously considered especially in patients with other co-morbid apart from just SMAS (e.g. non-verbal ASD) since it can cause aspiration pneumonia which can be fatal. Prompt diagnosis, timely intervention and increased vigilance are crucial to improve patient outcomes.

Author contributions: All authors contribute to the concept, data analysis, draft and review of manuscript

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

Funding: No funding was received for the study.

Acknowledgement: The authors thank the Department of Radiology for providing the radiography of the patient and Department of Surgery for the photography of the oesophagoduodenoscopy.

Ethical statement: Not applicable

REFERENCES

Akel, R., Daou, I., Jamal, D., Hobeika, E., Aoun, R., Nawfal, G. 2024. Deadly aspiration pneumonia secondary to superior mesenteric artery syndrome. *Case Rep Gastrointest Med* **2024**(1): 5055948. https://doi.org/10.1155/2024/5055948.

Danushka, P.G.N., Jayasinghe, R., Wijemanne, A. 2023. Superior mesenteric artery syndrome treatment strategies: A case report. *SAGE Open Med Case Rep* **11**: 1-4. https://doi.org/10.1177/2050313X231184587.

Ganss, A., Rampado, S., Savarino, E., Bardini, R. 2019. Superior mesenteric artery syndrome: A prospective study in a single institution. *J Gastrointest Surg* **23**(5): 997-1005. https://doi.org/10.1007/s11605-018-3984-6.

Guerra, J.D.M.B., Neto, J.S.D.M., Freitas, S.R.G.D., Nogueira, K.D.M., Souza, L.K.M.D. 2023. Superior mesenteric artery syndrome: A systematic review. *Int J Innov Educ Res* **11**(3): 1-18. https://doi.org/10.31686/ijier.vol11.iss3.4014.

Mohammad Kazmin, N.E., Kamaruzaman, L., Wong, Z., Fong, V.K., Mohd, R., Mustafar, R. 2020. Acute kidney injury caused by superior mesenteric artery syndrome. *Case Rep Nephrol* **2020**: 8364176. https://doi.org/10.1155/2020/8364176.

Navandhar, P.S., Shinde, R.K., Gharde, P., Nagtode, T., Badwaik, N. 2024. Understanding superior mesenteric artery syndrome: Etiology, symptoms, diagnosis, and management. *Cureus* **16**(6): e61532. https://doi.org/10.7759/cureus.61532.

Payus, A.O., Rahim, M.F.A., Remli, R. 2019. A rare and life-threatening but treatable cause of intractable vomiting: A case report on superior mesenteric artery syndrome. *Gulhane Med J* **61**(4): 186-8. https://doi.org/10.26657/gulhane.00082.

Pickhardt, P.J. 2020. Positive oral contrast material for abdominal CT: Current clinical indications and areas of controversy. *Am J Roentgenol* **215**(1): 69-78. https://doi.org/10.2214/AJR.19.21989.

Rajput, M.Z., Kapoor, S., Wright, A.J., Friedman, D.D., Patlas, M.N., Mellnick, V.M. 2023. The use of enteric contrast in the emergency setting. *Radiol Clin N Am* **61**(1): 37-51. https://doi.org/10.1016/j.rcl.2022.09.002.

Riera, D.C., Phalen, J.A. 2013. Superior mesenteric artery syndrome in a patient with autism spectrum disorder: Case report and review of the literature. *J Autism Dev Disord* **43**(1): 244-6. https://doi.org/10.1007/s10803-012-1548-2.

Salehi, A.M., Salehi, H., Hasanzarrini, M., Khanlarzadeh, E. 2022. Report of a misleading case of the superior mesenteric artery syndrome. *Case Rep Gastrointest Med* **2022**: 6013579. https://doi.org/10.1155/2022/6013579.

Shi, Y., Shi, G., Li, Z., Chen, Y., Tang, S., Huang, W. 2019. Superior mesenteric artery syndrome coexists with Nutcracker syndrome in a female: A case report. *BMC Gastroenterol* **19**(1): 15. https://doi.org/10.1186/s12876-019-0932-1.

Ugras, M., Bicer, S., Coskun, F.T., Romano, E., Ekci, B. 2017. Superior mesenteric artery syndrome: A rare but life threatening disease. *Turk J Emerg Med* **17**: 70-2. https://doi.org/ 10.1016/j.tjem.2017.01.001.