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Case Report

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Congenital Duodenal Membrane Requiring Differential Diagnosis from Superior Mesenteric Artery Compression Syndrome: A case report

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Abstract

Background Congenital duodenal membrane is a rare congenital gastrointestinal anomaly, typically manifesting during infancy, with fewer cases diagnosed in adulthood. Superior mesenteric artery syndrome (SMAS) shares clinical manifestations with congenital duodenal membrane, often leading to misdiagnosis.

Case presentation We report a case of a 16-year-old female patient presenting with "abdominal pain and distension for 2 months." These symptoms had been present since the neonatal period, suggesting a congenital partial duodenal obstruction. The patient had a history of feeding difficulties, postprandial vomiting, and growth retardation since birth. Abdominal enhanced CT showed a 16° angle between the superior mesenteric artery and the abdominal aorta. Upper gastrointestinal radiography indicated signs of obstruction at the beginning of the horizontal portion of the duodenum, with significant dilation of the duodenal bulb and descending part. Gastroscopy revealed marked dilation of the duodenal bulb, with a small opening visible at the distal end, though the endoscope could not pass through. Initial diagnosis considered duodenal stenosis, but exploratory laparotomy revealed a

diaphragm in the descending duodenum, leading to a diagnosis of congenital duodenal membrane. Partial resection of the diaphragm was performed, and the patient's symptoms were completely relieved post-surgery. Follow-up after six months showed improvement in eating status, with the patient able to consume solid food without abdominal pain or distension, and her weight was within the normal range for her age.

Conclusion This case highlights that, in patients presenting with upper gastrointestinal obstruction, even when imaging suggests SMAS, the possibility of congenital duodenal membrane should be considered. Detailed medical history and endoscopic examinations are crucial for accurate diagnosis, and surgical exploration may be necessary for confirmation and treatment.

Keywords: Congenital Duodenal Membrane; Superior Mesenteric Artery Syndrome; Upper Gastrointestinal Obstruction; Differential Diagnosis; Case report

1 Introduction

Congenital duodenal membrane is a rare congenital gastrointestinal anomaly with an incidence of approximately 1 in 10,000 to 1 in 40,000 live births, typically presenting during the neonatal or infantile period [1]. Clinical manifestations can range from complete obstruction to partial obstruction, depending on the size of the perforation in the diaphragm. Complete obstruction often presents as bile-stained vomiting and abdominal distension in neonates, while partial obstruction may remain asymptomatic until adulthood [2].

Superior mesenteric artery syndrome (SMAS) occurs when the angle between the superior mesenteric artery and the abdominal aorta decreases (usually <25°), leading to compression of the third portion of the duodenum and causing symptoms of upper gastrointestinal obstruction [3]. SMAS is often seen in individuals who have recently undergone significant weight loss and presents with symptoms such as postprandial bloating, nausea, and vomiting [4].

Due to the clinical similarities between congenital duodenal membrane and SMAS, and the possibility of concurrent occurrence, careful differentiation is required during diagnosis. This case reports a patient initially diagnosed with SMAS

but ultimately confirmed to have a congenital duodenal membrane, aiming to provide clinical insights and diagnostic approaches for physicians.

2 Case presentation

2.1 Chief complaints

A 16-year-old Chinese female presented to the clinic with a complaint of abdominal pain and distension for 2 mo.

2.2 History of past illness

The patient has no significant past medical history.

2.3 History of present illness

The patient had feeding difficulties and vomiting after eating since birth. The caregivers could not reliably recall whether the vomitus was bilious or non-bilious, so the exact level of obstruction relative to the ampulla of Vater could not be determined. The local hospital once suggested giving up treatment, presumably due to suspected congenital duodenal obstruction, but no definitive diagnosis was documented according to caregivers' recollection. The family insisted on raising the child, maintaining nutrition with liquid diet. The patient had delayed growth and development. Recently, the symptoms worsened with significant abdominal pain and distension after eating.Two months prior to admission, after attempting to transition from a purely liquid diet to semi-liquid and solid foods, she developed aggravated postprandial abdominal pain and distension. This likely explains the solid food residue observed in the duodenal bulb during endoscopy.

2.4 Personal and family history

The patient denied any family history of congenital duodenal membrane.

2.5 Physical examination

At admission, the patient's height was 150 cm (<10th percentile), weight 43 kg (<15th percentile), and BMI 19.1 kg/m² (WHO Z-score ≈ -1.0), indicating mild chronic undernutrition. The physical examination shows thin habitus, reduced subcutaneous fat. Abdomen soft and non-tender at rest, with mild postprandial distension and visible peristaltic waves. Bowel sounds were hyperactive during symptomatic episodes; no guarding, rebound, or palpable mass.

2.6 Laboratory examinations

Laboratory results were within normal limits.

2.7 Imaging examinations

Enhanced CT of the small intestine showed that the angle between the superior mesenteric artery and the abdominal aorta was 16° (normal range: 38°-65°) (Figure 1). The distance between the SMA and the aorta was not measured in this case, which is a limitation. Upper gastrointestinal endoscopy revealed significant dilation of the duodenal bulb with some food residue distally, and an apparent opening was observed. However, the endoscope could not be advanced beyond the distal duodenal bulb due to obstruction, which prevented direct visualization of the suspected membrane-like structure in the distal duodenum (Figure 2). Because of the possibility of other causes of obstruction and the rarity of congenital duodenal membrane in adolescents, surgical exploration was considered necessary to obtain a definitive diagnosis and provide treatment. Upper gastrointestinal contrast study showed obstruction at the beginning of the horizontal segment of the duodenum, but without the typical "pencil sign" of superior mesenteric artery syndrome. The duodenal bulb and descending portion showed significant dilation with fluid accumulation. The imaging also demonstrated a characteristic ‘double bubble’ sign, which is a classic indicator of congenital duodenal obstruction and supports the final diagnosis (Figure 3).

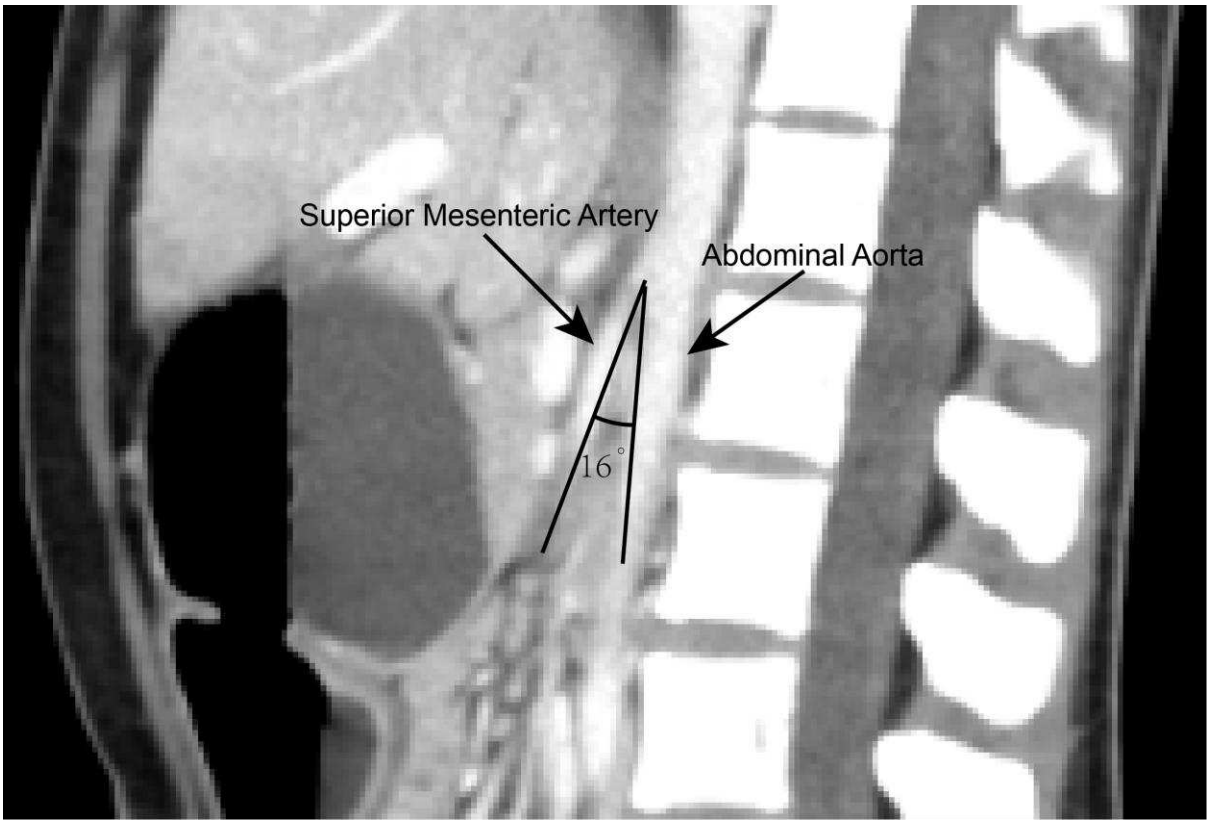


Figure 1 Reduced aortomesenteric angle (16°) on contrast-enhanced CT of the

small intestine.

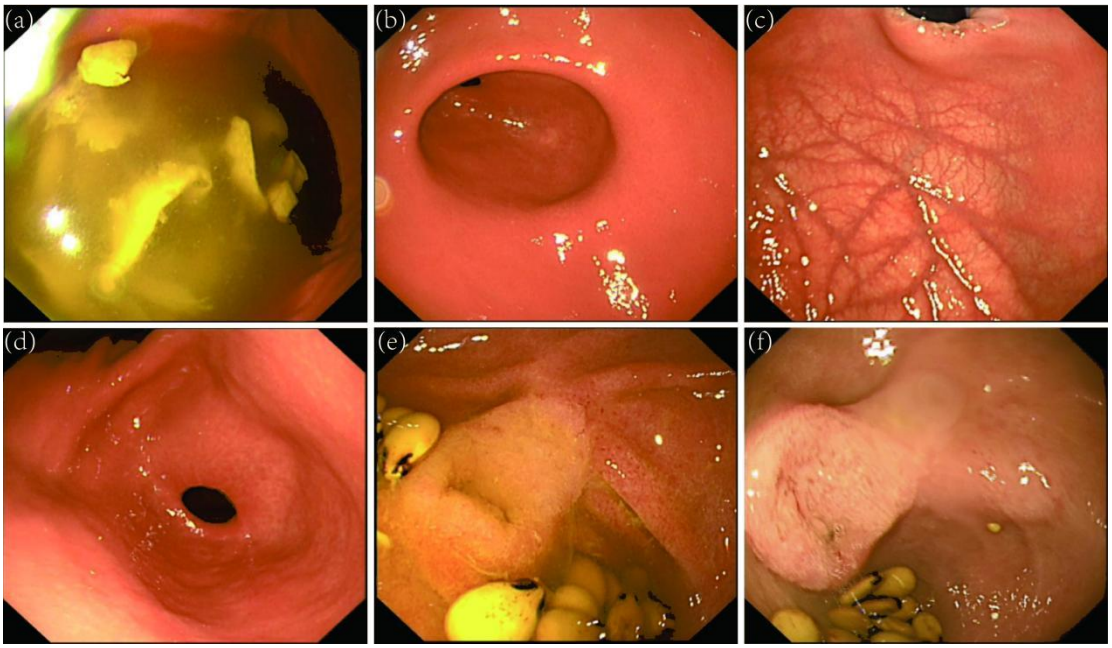


Figure 2 Endoscopic examinations. (a) The endoscopic examination of the gastric fundus revealed a large amount of bile and some food remnants, with no significant abnormalities in the mucosa. (b) The gastric antrum showed mucosal congestion, but no significant lesions were observed. (c) Endoscopic examination of the duodenal bulb revealed significant dilatation, with a small amount of food remnants at the distal end. A possible opening was observed. (d) The endoscopic view of the distal duodenal bulb showed significant expansion of the duodenal lumen, making it difficult to further advance the endoscope into the area.
(e) The pyloric opening was round, with normal opening and closing, and no abnormalities were noted. (f) The gastric angle appeared normal, with no abnormalities observed.

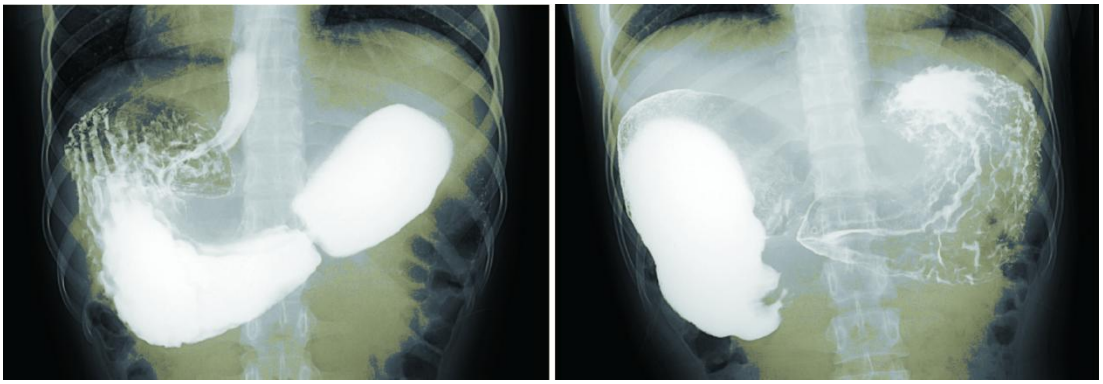


Figure 3 Upper gastrointestinal contrast study showing duodenal obstruction with proximal dilation.

3 Final diagnosis

Based on the patient's imaging examinations, the final diagnosis was duodenal stenosis: SMAS? Others?

4 Treatment

After multidisciplinary discussion, an exploratory laparotomy was decided. Open surgery was selected over laparoscopy because of significant duodenal dilatation, uncertainty regarding the obstruction’s exact nature, and the need for tactile assessment. Alternative options such as laparoscopic membrane resection or duodenojejunostomy were considered but deemed less appropriate in this setting. Exploration revealed significant dilation of the duodenal bulb and descending portion. The retroperitoneum was incised, and the duodenum at the junction of the descending and horizontal portions was mobilized at the antimesenteric border. Examination up to the bulb showed no obvious spasm, stenosis, or mass. Therefore, a duodenojejunostomy was planned. When attempting to pass a nasogastric tube orally, it could not traverse the junction between the descending and horizontal portions of the duodenum. Small openings were made at the antimesenteric borders of the jejunum (15 cm distal to the Treitz ligament) and the horizontal portion of the duodenum. A cutting stapler was inserted and fired along the antimesenteric border. Through the common opening, examination of the descending duodenum revealed a mucosal diaphragm with no apparent opening, suggesting congenital duodenal diaphragm. The opening size was not measured during surgery. The center of the diaphragm was incised, and a portion was resected and sent for examination. The remaining mucosal edges were sutured, and the duodenal incision was closed. Histopathological examination of the specimen showed chronic mucosal inflammation with erosion, vascular dilation with hemorrhage, and focal areas of necrosis.

5 Outcome and follow-up

The patient recovered well after surgery, with no symptoms during the 6-month

follow-up period. The patient's eating condition improved significantly, with normal intake of solid food and no symptoms of abdominal pain or distension. Her weight also reached the normal range for females of the same age.

6 Discussion

Congenital duodenal membrane is a rare congenital gastrointestinal anomaly linked to abnormal recanalization of the duodenal lumen during embryonic development [5]. In weeks 5-6 of embryogenesis, the duodenal lumen is filled by rapidly proliferating epithelial cells, and by weeks 8-10, apoptosis and vacuolization form the duodenal lumen. Incomplete recanalization leads to the formation of a duodenal membrane [6]. The disease's incidence is extremely low, and patients with a small perforation may not exhibit symptoms until adulthood, with approximately 30% of congenital duodenal membrane patients being diagnosed in adulthood [7]. In this case, the 16-year-old adolescent presented with a small perforation, causing partial obstruction with gradually worsening symptoms over the past year. Because the bilious nature of vomiting was not documented, the precise level of obstruction before surgery remained uncertain.

SMAS occurs when the angle between the superior mesenteric artery and the abdominal aorta decreases (typically <25°), leading to compression of the third portion of the duodenum [8]. However, more recent studies suggest stricter thresholds may be more predictive, with symptomatic cases often showing an angle of around 22° and an aortomesenteric distance of about 8 mm [9,10]. In patients with severe undernutrition or anatomical variation, even smaller angles (6–15°) and distances (2–8 mm) have been reported [11]. The normal angle is between 38°-65°, with a distance between the two arteries of 10-28 mm [12]. In this case, the CT showed a 16° angle between the superior mesenteric artery and the abdominal aorta, suggesting the possibility of SMAS. The absence of the typical 'pencil sign' on the upper GI series reduced the likelihood of SMAS, but given the markedly reduced aortomesenteric angle and clinical severity, exploratory laparotomy was ultimately chosen to rule out other obstructive lesions.

Congenital duodenal membrane and SMAS share clinical features, including

upper gastrointestinal obstruction symptoms such as nausea, vomiting, and bloating [13]. Radiological findings may also overlap, with dilation of the duodenal bulb and descending portion. However, they differ in etiology, pathogenesis, and treatment methods [3].

Detailed imaging, especially upper gastrointestinal radiography and gastroscopy, is essential for differential diagnosis. Upper gastrointestinal radiography can show the specific location and degree of duodenal obstruction, while gastroscopy can directly visualize membrane-like structures, providing direct evidence for diagnosis [14]. In this case, gastroscopy revealed a typical membrane-like structure in the descending duodenum. However, due to limited clinical experience and the rarity of this disease in adults, this diagnosis was initially overlooked.

CT angiography is an important diagnostic method for SMAS, as it accurately measures the angle and distance between the superior mesenteric artery and the abdominal aorta [15]. However, it is worth noting that a reduced angle between the superior mesenteric artery and the abdominal aorta does not necessarily indicate SMAS. The reduced SMA – aorta angle in this patient may have been secondary to chronic undernutrition leading to loss of mesenteric fat, or simply represent an anatomic variant. This highlights the need to interpret SMA – aorta angle measurements cautiously in patients with low BMI. Agrawal et al. [8] reported that about 20% of asymptomatic individuals also exhibit a reduced angle between the superior mesenteric artery and the abdominal aorta. In this case, despite the reduced angle on CT, the final diagnosis was congenital duodenal membrane, emphasizing that relying solely on angiographic findings could lead to misdiagnosis. To facilitate clinical differentiation between congenital duodenal membrane (CDM) and superior mesenteric artery syndrome (SMAS), we summarized their key distinguishing features in Table 1.

Table 1. Differential Diagnosis between Congenital Duodenal Membrane (CDM) and Superior Mesenteric Artery Syndrome (SMAS)

Feature	Congenital duodenal membrane (CDM)	Superior mesenteric artery syndrome (SMAS)
Symptoms	Chronic/intermittent vomiting (sometimes bilious), feeding difficulties	Acute/subacute vomiting, postprandial pain, weight loss
Imaging	Duodenal bulb dilatation, membrane sign, double bubble	Narrowed aortomesenteric angle (<25°), short aortomesenteric distance (<8–10 mm)
Endoscopy	Possible visualization of web, difficulty advancing scope	External compression, no intraluminal membrane
Surgery	Membrane excision or duodenojejunostomy	Duodenojejunostomy

7 Conclusion

Congenital duodenal membrane is a rare congenital gastrointestinal anomaly that should be differentiated from SMAS and other diseases. In patients presenting with upper gastrointestinal obstruction, even when imaging suggests SMAS, congenital duodenal membrane should be considered in the differential diagnosis. Patient history and endoscopic findings are of critical value for confirmation.

Abbreviations

SMAS Superior mesenteric artery syndrome

CDM Congenital duodenal membrane

SMA Superior mesenteric artery

CT Computed tomography

BMI Body mass index

GI Gastrointestinal

mo Month(s)

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Author contributions

Qi HB and Jin XR contributed to manuscript writing and editing; Luo SQ, Ding X, Li XS, Zhang MF, Yang YS, Zhou YJ, Liang BC, Chen JJ and Wang Q contributed to literature search and figure preparation; Wang HB and Dai L contributed to conceptualization and supervision; all authors have read and approved the final manuscript.

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Data availability

Data used to support the fndings and the conclusions of this manuscript are available from the corresponding author upon request.

Declarations

Ethics approval and consent to participate

The privacy of the patient was considered, and the manuscript does not display any identifying information.

Consent for publication

Written informed consent was obtained from the patient's parents for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare that they have no competing interests.

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