

CASE REPORT

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Congenital absence of omentum with short bowel syndrome: a case report

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Abstract

Introduction Short bowel syndrome can manifest as either an inherited or acquired condition, with the inherited form occurring sporadically. Furthermore, the complete absence of the omentum at birth (congenital absence) is a highly uncommon event.

Case report This case report presents a unique confluence of these rare conditions in a 38-year-old Iranian male with a prior history of intestinal obstruction requiring right hemicolectomy and ileostomy. He subsequently presented to the emergency department experiencing intense abdominal pain and swelling in his lower extremities. Laboratory investigations revealed an elevated white blood cell count and metabolic alkalosis. During the surgical exploration prompted by his acute presentation, both congenital absence of the omentum and short bowel syndrome were confirmed. A jejunostomy was performed, but unfortunately, this intervention resulted in severe malabsorption and subsequent cachexia. This case sheds light on the rare occurrence of anastomotic leakage and subsequent peritonitis following right hemicolectomy and ileostomy in a patient with the combined conditions of congenital short bowel and congenital absence of the omentum.

Conclusion This unique presentation highlights the potential complexities that can arise due to the convergence of these rare medical conditions.

Keywords Short bowel syndrome (SBS), Congenital absence of omentum, Inflammatory bowel disease, Intestinal adaptation

Background

In accordance with the 2010 guidelines from the International Study Group of Rectal Cancer, anastomotic leakage (AL) is defined as a defect in the intestinal wall at the anastomotic area, resulting in a connection between the

luminal interior and exterior [1]. AL, a critical and life-threatening complication specific to colorectal surgery, can increase postoperative morbidity, mortality, and hospitalization duration [2]. Severe leaks may lead to peritonitis, sepsis, and necessitate emergency surgery. Risk factors for AL include malnutrition, weight loss, hypoalbuminemia, cardiovascular disease, corticosteroid use, peritonitis, bowel obstruction, and chronic obstructive pulmonary disease [3, 4]. AL after right hemicolectomy potentially leads to diffuse peritonitis throughout the abdomen. Laparotomy is the preferred method of treating diffuse peritonitis [5, 6]. No clear and uniform definition of short bowel syndrome (SBS) has been established. In this syndrome, malnutrition arises from a reduction in the length of the small intestine below the expected

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amount (less than half the size expected for gestational age) [7].

Children with SBS often require total parenteral nutrition (TPN), predisposing them to various liver disorders [8, 9].

SBS can be congenital or acquired, with conditions such as necrotizing enterocolitis, gastroschisis, midgut volvulus, or intestinal atresia causing it secondarily [10]. Key symptoms of SBS, characterized by reduced absorptive intestinal length, include diarrhea, vomiting, failure to thrive, and manifestations consistent with intestinal obstruction [11]. Congenital short bowel (CSB) is exceedingly rare, with a prevalence estimated to be greater than 1 in 1,000,000 [12].

Complete congenital absence of the omentum is exceptionally rare. Previous case reports in 1958 and 2016 described cases involving double omental herniation and surgical management of a peritoneal pseudocyst causing acute kidney injury, both attributed to congenital absence of the omentum [13, 14].

Case presentation

A 38-year-old Iranian male patient presented to the emergency department of Kowsar Hospital, Semnan, on 2 October 2021, with chief complaints of severe abdominal pain and lower extremity edema. The pain, originating in the umbilical region and spreading across the entire abdomen, prompted the visit. Vital signs were recorded as follows: blood pressure (BP) = 130/90 mmHg, heart rate (HR) = 80 bpm, and temperature (T) = 37 °C. Abdominal examination revealed normal bowel sounds, slight tenderness in the inguinal region, and tympanic sounds. Also, he was noted to have congenital SBS with a measured total small bowel length of approximately 185 cm, significantly shorter than the normal range, and an absence of the omentum.

On 10 August 2021, the patient underwent a right hemicolectomy and ileostomy owing to abdominal pain associated with a diagnosis of intestinal obstruction. Subsequently, on 21 September 2021, the patient had a colostomy removed and was hospitalized for 6 days (including 3 days in the intensive care unit [ICU]). Post-discharge, the patient experienced abdominal pain and edema, with an escalation of symptoms on 1 October 2021. Nausea was reported upon admission, but without vomiting, and there were no complaints related to urination. Due to postprandial pain and poor appetite, the patient's nutritional intake was compromised. No fever, chills, or dyspnea were reported. Lower extremity edema was noted (2+), with no clubbing or cyanosis observed. Other vital signs were within normal ranges (pulse rate 82 beats/minute, blood pressure 110/70 mmHg, respiratory rate 18 breaths/minute, temperature 37 °C). Abdominal

tenderness hindered a complete examination owing to severe pain. Examination of the head, neck, and chest revealed no specific findings.

Full blood count indicated leukocytosis (hemoglobin 11.7 g/dL, white cell count $19.23 \times 10^3/\mu\text{L}$, platelets $406 \times 10^3/\mu\text{L}$, RBC $4.30 \times 10^6/\mu\text{L}$, and hematocrit [HTC] 34.3%), while arterial blood gas analysis showed metabolic alkalosis (pH 7.56, partial pressure of oxygen [PO₂] 204.5 mmHg, partial pressure of carbon dioxide [PCO₂] 31.9 mmHg, base excess [BE] 7.0 mmol/L, and bicarbonate [HCO₃] 29.1 mmol/L).

Abdominal and chest X-rays of the 38-year-old male with congenital short bowel syndrome and absence of the omentum. Supine abdominal radiograph demonstrating dilated loops of bowel and absence of the omentum, with no evidence of air-fluid levels (Fig. 1). Also, chest X-ray showed no significant abnormalities, confirming normal cardiopulmonary findings (Fig. 2). Upright abdominal radiograph revealed evidence of gaseous distension and loops consistent with the patient's diagnosis (Fig. 3).

A contrast-enhanced computed tomography (CT) scan of the abdomen and pelvis was recommended. Fecal secretion from the patient's sutures raised concerns about postoperative peritonitis due to anastomotic leakage, prompting immediate surgical intervention. The patient underwent emergency laparotomy, drainage, and double barrel jejunostomy (Figs. 4, 5, 6, and 7).

The patient's recovery was uneventful, with oral intake of clear fluids permitted on postoperative day 2 and full fluids on postoperative day 4.

For congenital absence of omentum and short bowel syndrome, treatment options encompass nutritional support, such as total parenteral or enteral nutrition. Severe cases may necessitate surgical intervention to lengthen the remaining bowel or perform an intestinal

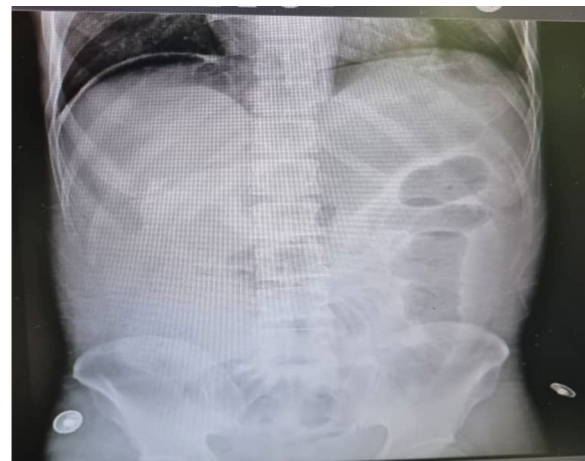


Fig. 1 X-ray of the abdomen and pelvis



Fig. 2 X-ray of the abdomen and pelvis



Fig. 3 X-ray of the abdomen and pelvis

transplant. Strategies for short bowel syndrome resulting from congenital absence of omentum may involve nutritional support, along with enhancing the expression of transport proteins (such as Na^+ /glucose cotransporters, Na^+ / H^+ exchangers) to facilitate intestinal adaptation. Human interventions include glutamine and growth hormone treatment or glucagon-like peptide administration, though multicenter studies are required for conclusive evidence.

Discussion

This case report details a 38-year-old male with congenital absence of the omentum and short bowel syndrome (SBS), highlighting the complex interplay between these rare conditions. The patient underwent surgical intervention for intestinal obstruction, which ultimately led to severe malabsorption and cachexia due to SBS. This



Fig. 4 Computed tomography scan revealed an image of ileus and intramural gas in the small intestine

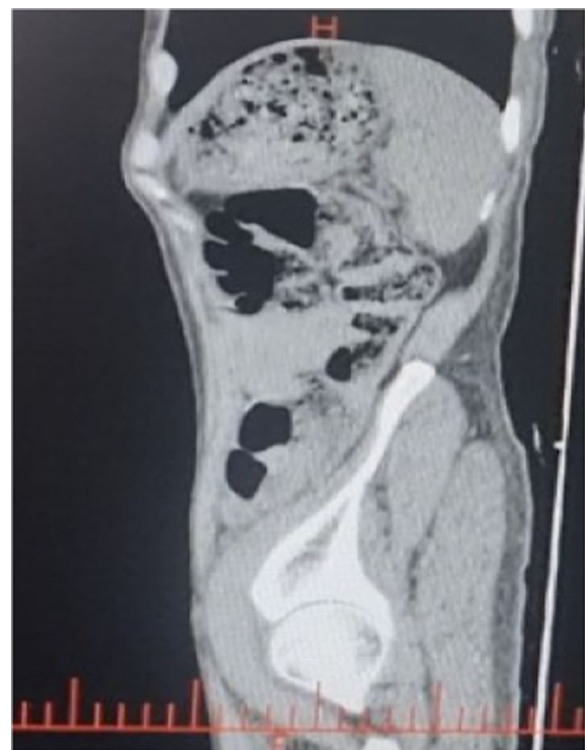


Fig. 5 Computed tomography scan revealed an image of ileus and intramural gas in the small intestine



Fig. 6 Computed tomography scan revealed an image of ileus and intramural gas in the small intestine

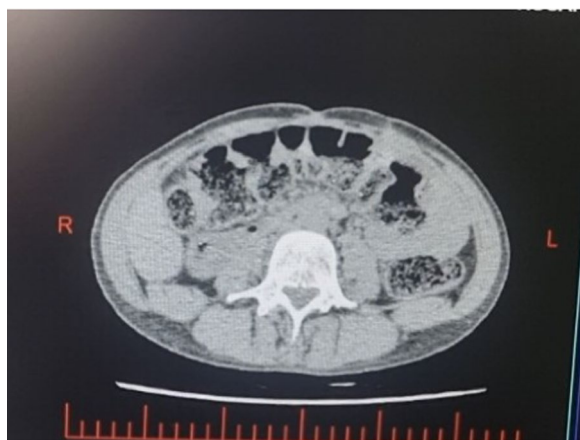


Fig. 7 Computed tomography scan revealed an image of ileus and intramural gas in the small intestine

unique presentation raises critical questions regarding the prognosis and management strategies for patients with SBS complicated by congenital anomalies.

The findings from this case indicate that the absence of the omentum significantly complicates the clinical

management of SBS. The omentum plays a crucial role in abdominal organ support, immune response, and fat storage, which are vital for recovery and adaptation following intestinal surgeries [7]. In this patient, the lack of omental tissue likely exacerbated postoperative complications such as anastomotic leakage and peritonitis, leading to a more challenging recovery process. Previous studies have shown that patients with SBS often require total parenteral nutrition (TPN) to manage malabsorption effectively [15]. However, the absence of the omentum may further impair nutritional absorption and increase susceptibility to infections and inflammatory responses.

Research indicates that congenital short bowel syndrome is exceedingly rare, with a prevalence greater than 1 in 1,000,000 [12]. While some studies have documented improved outcomes in patients with SBS through innovative surgical techniques and nutritional support [3], this case diverges from typical outcomes owing to the dual presence of congenital anomalies. The literature suggests that patients with congenital SBS often face a higher risk of complications compared with those with acquired forms [11]. This case aligns with earlier findings that emphasize the need for meticulous postoperative care in such patients, as they are at an increased risk for severe malnutrition and related complications [9] (Table 1).

The primary limitation of this case report is its singular nature; it represents an isolated instance of a rare combination of conditions. This restricts the ability to generalize findings across broader populations. In addition, the retrospective analysis relies on limited clinical data from one patient, which may introduce bias. The absence of long-term follow-up data regarding nutritional status and quality of life post-surgery further limits comprehensive conclusions about optimal management strategies.

Future research should focus on multicenter studies to gather data from a larger cohort of patients exhibiting similar rare conditions. Investigating the long-term outcomes associated with congenital absence of the omentum in patients with SBS could provide valuable insights into their prognosis and management strategies. In addition, exploring adjunctive therapies such as glutamine supplementation or growth hormone

Table 1 Impact of congenital absence of the omentum on the prognosis of patients with short bowel syndrome (SBS), along with relevant references:

Aspect	Impact of congenital absence of omentum on SBS prognosis	References
Nutritional absorption	The absence of the omentum may worsen malabsorption, complicating nutritional support strategies	[7, 15]
Postoperative complications	Increased risk of anastomotic leakage and peritonitis following surgical interventions	[1, 3]
Recovery challenges	Patients may experience prolonged recovery due to complications associated with SBS and omental absence	[11, 12]
Management strategies	Requires tailored nutritional support, possibly including total parenteral nutrition (TPN)	[9]
Long-term outcomes	Potential for severe malnutrition and growth disturbances if not properly managed	[7, 15]

treatment could enhance intestinal adaptation and improve nutritional outcomes [9]. Understanding the genetic underpinnings of congenital short bowel syndrome may also lead to targeted therapies that could mitigate complications associated with these rare presentations.

Conclusion

Patients with acute mesenteric ischemia undergo laparotomy owing to obstruction of the small intestine if the length of the small intestine remains intact. If the small intestine is too short then resection and anastomosis are often not recommended. However, according to the above patient, it is recommended for these patients, or in any other condition in which the colon is healthy, when it is possible to keep 10–15 cm at the beginning of the jejunum and 20–30 cm at the end of the ileum intact. The patient should have a complete resection and anastomosis, with a high likelihood of the patient surviving without nutritional dependence. Complications of short bowel syndrome can include malabsorption of nutrients, fluid and electrolyte imbalances, liver dysfunction, bacterial overgrowth in the remaining small bowel, and growth disturbances.

Abbreviations

SBS	Short bowel syndrome
CSB	Congenital short bowel
AL	Anastomotic leakage
CBC	Complete blood count
ICU	Intensive care unit
TPN	Total parenteral nutrition
BP	Blood pressure
HR	Heart rate
RBC	Red blood cell
HTC	Hematocrit
PO ₂	Partial pressure of oxygen
PCO ₂	Partial pressure of carbon dioxide
BE	Base excess
HCO ₃	Bicarbonate
CT	Computed tomography

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Author contributions

RM and MSK discovered this case and wrote the manuscript. ZKF and PB collected and analyzed the data. SS reviewed and edited the manuscript. All authors have read and approved the final manuscript.

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Availability of data and materials

These data are considered confidential patient information and are not publicly available due to privacy concerns. However, access to the anonymized data can be provided upon reasonable request from qualified researchers interested in further analysis for scientific purposes. Requests should be directed to the corresponding authors.

Declarations

Ethics approval and consent to participate

This case report was prepared following the ethical guidelines for publishing patient information. Written informed consent for the publication of this report was obtained from the patient, ensuring anonymity and confidentiality. No formal ethics committee approval was deemed necessary for this case report as it does not involve manipulation or experimentation on the patient.

Consent for publication

Written informed consent was obtained from the patient 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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