

Midgut Volvulus with Cavernous Transformation of the Portal Vein in a Middle-aged Patient: A Case Report

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ABSTRACT

INTRODUCTION - Midgut volvulus and cavernous transformation of the portal vein (CTPV) are rare conditions in adults. Midgut volvulus, a congenital anomaly of the intestines, can cause a life-threatening intestinal obstruction. CTPV, on the other hand, is a complication of chronic portal vein thrombosis. In adult patients, these conditions are rare and manifest with a collection of non-specific symptoms, posing significant diagnostic challenges.

CASE PRESENTATION - We report a case of A 46-year-old male who had severe central abdominal pain, vomiting, and obstipation for three days, indicating intestinal obstruction. The CT with oral contrast revealed midgut malrotation with small bowel obstruction due to volvulus. The post-operative CT with contrast findings reveals the presence of multiple collaterals of the portal vein, which is suggestive of cavernous transformation. During the procedure, intra-operative findings revealed a Ladd's band, the duodenojejunal (DJ) junction in the right upper quadrant, and a gangrenous bowel segment measuring approximately 30 centimeters in length and located 50 centimeters distal to the DJ junction. The surgical approach included a Ladd's procedure, lysis of the band, resection of the gangrenous segment, with a side-to-side anastomosis, and an appendectomy.

CONCLUSION - A unique case study presents an adult with two uncommon conditions—midgut volvulus and cavernous transformation of the portal vein. Their occurrence is rarely seen in adults and requires a multidisciplinary approach for diagnosis and management, emphasizing early detection and intervention to prevent severe complications. This case report contributes novel insights into these conditions and underscores the need for swift action, potentially marking the first instance documented in medical literature.

KEYWORDS - midgut volvulus, midgut malrotation, cavernous transformation of the portal vein, Intestinal obstruction, case report.

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INTRODUCTION

Malrotation is a congenital anomaly occurring during embryonic development, leading to an abnormal positioning of the intestines. Among its complications, midgut volvulus frequently emerges within the initial months of life, often marked by episodes of bilious vomiting [1]. This critical condition involves the clockwise rotation of the bowel around the mesenteric vascular pedicle's axis, leading to an atypical orientation of the superior mesenteric artery (SMA) and superior mesenteric vein (SMV). This characteristic narrowing and rotation create the diagnostic hallmark known as the "whirlpool sign" [2].

However, diagnosing midgut volvulus in adolescents and adults poses challenges due to its predominately nonspecific presentation, often resulting in overlooked malrotation. The incidence of malrotations is estimated to be approximately 1 in 6000 live births [3], whereas in adults, reported incidences range from 0.0001% to 0.19% [4]. Due to their rarity, the documented cases of malrotation in adults are relatively rare, with only about 100 cases reported in the available literature [5].

An obstruction or blockage of the main portal vein causes a condition known as cavernous transformation of the portal vein, which results in the formation of multiple small, looping venous channels, or collaterals, surrounding the liver. Usually, chronic portal vein thrombosis causes this condition. A limited number of documented cases discuss the cavernous transformation of the portal vein (CTPV) in adults [6]. It is noteworthy that CTPV is observed more frequently in children than in adults and stands as the primary cause of portal hypertension among pediatric populations [7].

This scarcity of reported cases of these two conditions in adults accentuates the complexity of diagnosing and treating them beyond infancy. The lack of specific symptoms and the rarity of occurrences in this age group contribute to the diagnostic challenges, potentially leading to delayed recognition and management of these conditions.

CASE PRESENTATION

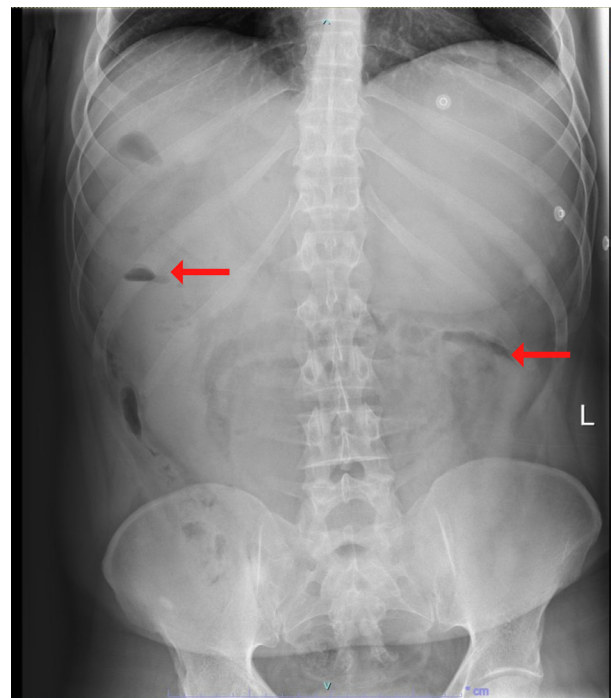
A 46-year-old male presented to the emergency department (ED) reporting persistent central abdominal pain and discomfort lasting six days, intensifying notably within the last three days. The pain exhibited colicky characteristics, exacerbating post-consumption of food or liquids, albeit partially alleviated with over-the-counter

painkillers. Within the three days leading up to admission, the patient reported experiencing vomiting of a greenish substance immediately following oral intake, accompanied by a significant decrease in appetite. Additionally, he suffered from obstipation during these 3 days.

There was no documented medical history indicating prior hospitalizations for similar symptoms. The patient reported being a smoker with no dependence on alcohol or any drugs. Furthermore, he did not have any pre-existing medical conditions or a history of abdominal surgeries. Moreover, there were no indications or past occurrences suggestive of intestinal obstruction. During the general physical examination, the patient demonstrated full consciousness and orientation to time, place, and person, exhibiting a Glasgow Coma Scale (GCS) score of 15.

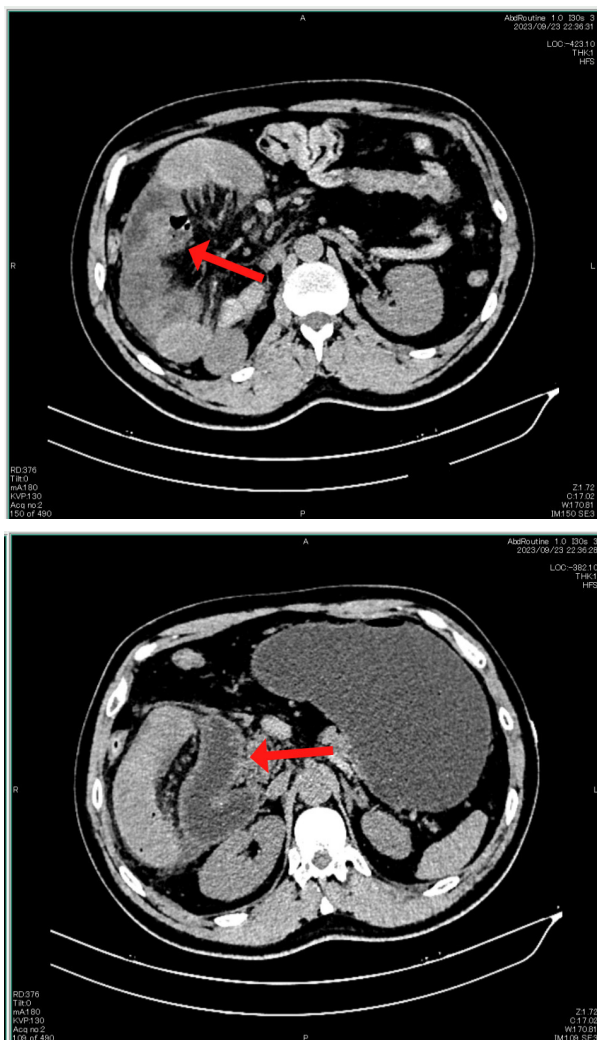
The abdominal examination demonstrated a lack of distention and mild tenderness observed in the periumbilical and suprapubic areas, as well as noted sluggishness in bowel sounds. The abdominal X-ray [Figures. 1] revealed the presence of two air-fluid levels without any indication of air accumulation beneath the diaphragm. Vital signs recorded included a body temperature of 36.7, pulse rate of 82 beats per minute, a blood pressure reading of 129/87 mmHg, a respiratory rate of 20 breaths per minute, oxygen saturation levels at 95%, and a body mass index (BMI) of 23.7 Kg/m².

Figure 1. Abdominal X-ray demonstrating two air-fluid levels



The laboratory findings indicated that Hemoglobin (HGB) levels were within normal limits at 15.2 g/dL. White blood cell count (WBC) was markedly elevated at 30,000 cells/mm³. Neutrophils (NEUT) were elevated at 87%, while Platelet count (PLT) was at 230 K/ μ L. The erythrocyte sedimentation rate (ESR) was 70 mm/hr. indicative of increased inflammatory activity. C-reactive protein (CRP) levels were significantly elevated at 350 mg/L, suggesting acute inflammation. The International Normalized Ratio (INR) was 1.36. Electrolyte levels are within normal ranges: potassium (K) at 4.6 mmol/L, sodium (Na) at 138 mmol/L, chloride (Cl) at 97 mmol/L, and calcium (Ca) at 10.2 mg/dL.

Figure 2. Axial section CT scan without contrast



The CT scan [Figures. 2] [Figures. 3] of the abdomen and pelvis without contrast (Oral contrast was given by the treating team seen reaching the proximal jejunum) revealed abnormal positioning

of the duodenum and duodenojejunal junction in the right upper abdomen, indicating midgut malrotation. Notably, the stomach, duodenum, and proximal jejunum appeared distended and filled with fluid, measuring up to 4.5 cm in maximum diameter, accompanied by wall thickening and minimal fluid accumulation (HU 10). A transition point was observed at the right upper abdomen with associated distal small bowel collapse, consistent with small bowel obstruction likely due to volvulus. However, there was no evidence of torsion or detorsion. Minimal ascites was present without evidence of pneumoperitoneum. Additionally, mesenteric lymph nodes were prominent. No focal lesions in the liver were noted, albeit a small hiatus hernia was observed. Lower lung cuts exhibited bibasal atelectatic bands.

Figure 3. Sagittal section CT without contrast scan demonstrating abnormal positioning of the duodenojejunal junction, with distended stomach, duodenum, and proximal jejunum.



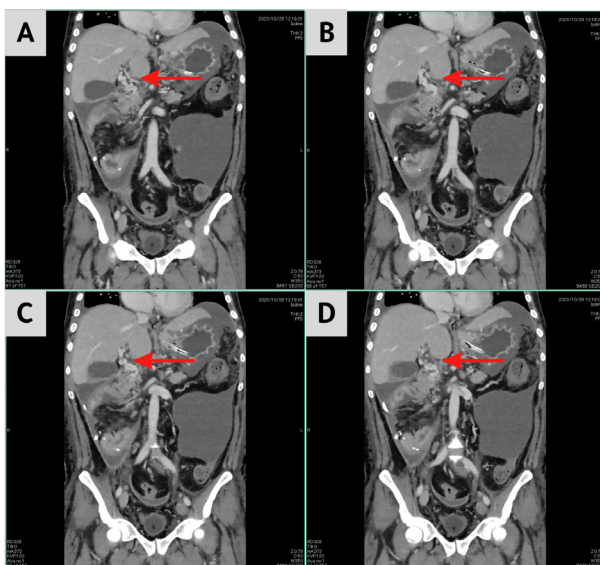
In the emergency department, the initial course of action involved the insertion of a nasogastric tube connected to Gumco suction and a urinary catheter connected to a urometer. Additionally, administering intravenous fluids and initiating broad-spectrum antibiotic therapy were implemented as part of the management plan.

The surgical intervention commences utilizing a laparoscopic approach, with a primary objective of mobilizing the colon and performing adhesiolysis. However, the easily bleeding inflamed tissue and the severely distended proximal bowel loops necessitate the conversion to an open proce-

dure, prompted by the challenges associated with achieving hemostatic control. Intraoperatively, several critical findings were noted, including the identification of a Ladd's band, the presence of the DJ (duodenojejunal) junction located in the right upper quadrant, and the alarming discovery of a segment of gangrenous bowel measuring approximately 30 centimeters in length. This necrotic bowel segment was situated about 50 centimeters distal to the DJ junction. The surgical approach employed a comprehensive strategy, beginning with a Ladd's procedure to address the anatomical abnormalities. Subsequently, lysis of the band was conducted, followed by resection of the gangrenous bowel segment. Followed by side-to-side anastomosis to restore bowel continuity and function. Additionally, considering the situation, an appendectomy was performed as part of Ladd's procedure.

The postoperative course was complicated by secondary peritonitis (infected ascites) and bilateral pleural effusions. Additionally, a suspected cavernous transformation of the portal vein underscores the complexity of the patient's condition. This condition was suggested by the absence of clear visualization of the portal vein at the porta hepatis and the presence of multiple collateral vessels. Further clinical correlation and management are warranted to address these observed complications and optimize the patient's recovery.

Figure 4. Post-operative CT scan with contrast revealing the absence of clear visualization of the portal vein at the porta hepatis and the presence of multiple collateral vessels.



The post-operative CT with contrast [Figures. 4] was done to investigate the cause of the ascites and rule out anastomotic leak. The findings

revealed persistent lymph node enlargement and fluid-filled ileal loops without signs of obstruction. Additionally, intra-abdominal and pelvic free fluid was present, focal lesions were present in multiple organs, and pleural effusion was present. However, no drainable collections were visible, and there was no evidence of pneumoperitoneum or bilateral hydronephrosis.

The cause of peritonitis, in this case, sparked controversy between spontaneous bacterial peritonitis (SBP) and secondary bacterial peritonitis. Following surgery, the patient developed ascites, which was drained percutaneously. Laboratory results supported the diagnosis of secondary peritonitis, and the post-operative CT scan did not reveal any ischemic lesions.

This was treated with percutaneous drainage and intravenous antibiotics tailored to the patient's symptoms and signs. The patient was subsequently initiated on full anticoagulation therapy, and a thrombophilia workup was conducted since the patient had extensive portal thrombosis extended into SMV. However, the results of the thrombophilia workup turned out to be negative.

DISCUSSION

The atypical presentation of midgut volvulus in adults contributes to diagnostic dilemmas, necessitating a high index of suspicion for timely intervention. Imaging modalities such as computed tomography (CT) scans and magnetic resonance imaging (MRI) play a crucial role in visualizing the twisted bowel segments and confirming the diagnosis.[8] However, misdiagnoses or delays in imaging may occur, emphasizing the need for a comprehensive understanding of the condition's clinical nuances.

Midgut volvulus in adults manifests with a spectrum of symptoms that can vary in intensity. Patients may experience severe abdominal pain, often colicky in nature, which gets worse by eating, accompanied by nausea and vomiting.[9] [10][11] Other common symptoms include bloating, distension of the abdomen, and the potential for bowel obstruction. In some cases, there may be visible signs of abdominal tenderness upon examination. It is crucial to note that midgut volvulus in adults can present with nonspecific symptoms, making it challenging to differentiate from other abdominal conditions.[10][11] Therefore, prompt medical attention and diagnostic evaluation are essential for accurate diagnosis and timely intervention.

Midgut volvulus necessitates urgent surgical intervention as a crucial step for a favorable

outcome. Swift diagnosis and immediate surgical measures are pivotal. Ladd's procedure is the established surgical approach for managing midgut volvulus and intestinal malrotation.[10] [12] As part of Ladd's procedure, an appendectomy is commonly performed to avert potential misdiagnosis of appendicitis in the future.[12] The mortality rate associated with Ladd's procedure demonstrates variability. A retrospective study spanning two decades involving eleven adult malrotation patients who underwent this surgical intervention reported an absolute mortality rate of zero[13]. However, it is important to note that postoperative complications were observed in up to 60% of adults, and these cases exhibited a substantially higher rate of reoperation following the initial procedure.[14][15]

Cavernous transformation of the portal vein (CTPV) typically arises as a consequence of prolonged portal vein thrombosis (PVT) or obstruction, leading to the development and enlargement of numerous small collateral vessels in proximity to a blocked portal vein [16]. This condition may result from extrahepatic portal vein obstruction, which presents as a rare vascular anomaly, often manifesting symptoms during childhood, frequently leading to incidental detection in adult cases and concurrent illnesses [17]. It is noteworthy that there is a scarcity of documented instances of CTPV in the adult population, as the diagnosis of CTPV is very rare in this population. [6] In adults, recognized causes of portal vein thrombosis encompass liver cirrhosis, hepatocellular carcinoma, pancreatitis, and splenectomy. Conversely, in children, identified causes involve intraabdominal infection, dehydration, and coagulation disorders [16].

However, Computed tomography (CT) serves as an efficient method for assessing the portal-mesenteric venous system [18].

CTPV manifests across a spectrum of clinical presentations. Remarkably, most patients exhibit asymptomatic manifestations at the point of diagnosis, [6] akin to our patient who exhibited symptoms of volvulus but lacked antecedent CTPV symptoms before admission. The characterization of CTPV as an infrequent condition predominantly relies on insights from clinical series and case reports, engendering substantial divergence in therapeutic approaches.[16]

CONCLUSION

This case report illustrates a rare clinical scenario of an adult patient who was presented with two uncommon conditions not usually seen in adults: midgut volvulus and cavernous transformation of

the portal vein. Midgut volvulus is a life-threatening condition that results from congenital malrotation of the intestines, causing intestinal obstruction and ischemia. Cavernous transformation of the portal vein is a chronic complication of portal vein thrombosis or obstruction, leading to the formation of collateral vessels that bypass the occluded portal vein. The etiology and pathophysiology of these two conditions are different and their simultaneous occurrence in the same patient is extremely unusual. Diagnosing and managing these two conditions require a multidisciplinary approach involving radiologists, surgeons, gastroenterologists, and hepatologists. This case report adds to the existing knowledge of these rare conditions. It highlights the importance of early detection and prompt intervention to prevent serious complications and improve patient outcomes.

AUTHORS' CONTRIBUTIONS

All authors contributed to the literature review, data collection, writing, and manuscript review. MR led the team and revised the manuscript.

INFORMED CONSENT

Verbal informed consent was obtained from the patient for the publication of this case report and any accompanying images.

REFERENCES

- 1 Langer JC. Intestinal rotation abnormalities and midgut volvulus. *Surg Clin North Am* [Internet]. 2017;97(1):147–59. Available from: <http://dx.doi.org/10.1016/j.suc.2016.08.011>
- 2 Nguyen HN, Navarro OM, Bloom DA, Feinstein KA, Guillerman RP, Munden MM, et al. Ultrasound for midgut malrotation and midgut volvulus: AJR expert panel narrative review. *AJR Am J Roentgenol* [Internet]. 2022;218(6):931–9. Available from: <http://dx.doi.org/10.2214/ajr.21.27242>
- 3 Butterworth WA, Butterworth JW. An adult presentation of midgut volvulus secondary to intestinal malrotation: A case report and literature review. *Int J Surg Case Rep* [Internet]. 2018;50:46–9. Available from: <http://dx.doi.org/10.1016/j.ijscr.2018.07.007>
- 4 Midgut volvulus in adult age associated with congenital malrotation – case report. *Persp In Surg* [Internet]. 2021;100(11). Available from: <http://dx.doi.org/10.33699/pis.2021.100.11.559-562>
- 5 Dhivakar, Singh SK, Das A, Katragadda S, Mishra A. Adult midgut malrotation with chronic volvulus with superior mesenteric artery (SMA) thrombosis: A recherche. *Cureus* [Internet]. 2023; Available from: <http://dx.doi.org/10.7759/cureus.43754>
- 6 Ramos. Cavernous transformation of portal vein secondary to portal vein thrombosis: A case report. *J Clin Med Res* [Internet]. 2012; Available from: <http://dx.doi.org/10.4021/jocmr775w>
- 7 Huang X, Lu Q, Zhang Y-W, Zhang L, Ren Z-Z, Yang X-W, et al. Intrahepatic portal venous systems in adult patients with cavernous transformation of portal vein: Imaging features and a new classification. *Hepatobiliary Pancreat Dis Int* [Internet]. 2023; Available from: <http://dx.doi.org/10.1016/j.hbpd.2023.01.002>
- 8 Xiong Z, Shen Y, Morelli JN, Li Z, Hu X, Hu D. CT facilitates improved diagnosis of adult intestinal malrotation: a 7-year retrospective study based on 332 cases. *Insights Imaging* [Internet]. 2021;12(1). Available from: <http://dx.doi.org/10.1186/s13244-021-00999-3>
- 9 Emami A, Asghari Y, Abedi Valukalaei H, Sakhaei M. Late presentation of incomplete jejunoileal junction atresia with recurrent midgut volvulus; A case report for chronic midgut volvulus in adults. *Caspian J Intern Med* [Internet]. 2023 Spring;14(2):380–5. Available from: <http://dx.doi.org/10.22088/cjim.14.2.380>
- 10 Ferreira MS, Simões J, Folgado A, Carlos S, Carvalho N, Santos F, et al. Recurrent midgut volvulus in an adult patient — The case for pexy? A case report and review of the literature. *Int J Surg Case Rep* [Internet]. 2020;66:91–5. Available from: <http://dx.doi.org/10.1016/j.ijscr.2019.11.037>
- 11 Fung AT, Konkin DE, Kanji ZS. Malrotation with midgut volvulus in an adult: a case report and review of the literature. *J Surg Case Rep* [Internet]. 2017;2017(5). Available from: <http://dx.doi.org/10.1093/ijscr/rjx081>
- 12 Al Smady MN, Hendi SB, AlJeboury S, Al Mazrooei H, Naji H. Appendectomy as part of Ladd's procedure: a systematic review and survey analysis. *Pediatr Surg Int* [Internet]. 2023;39(1). Available from: <http://dx.doi.org/10.1007/s00383-023-05437-7>
- 13 Kotobi H, Tan V, Lefèvre J, Duramé F, Audry G, Parc Y. Total midgut volvulus in adults with intestinal malrotation. Report of eleven patients. *J Visc Surg* [Internet]. 2017;154(3):175–83. Available from: <http://dx.doi.org/10.1016/j.jviscsurg.2016.06.010>
- 14 Durkin ET, Lund DP, Shaaban AF, Schurr MJ, Weber SM. Age-related differences in diagnosis and morbidity of intestinal malrotation. *J Am Coll Surg* [Internet]. 2008;206(4):658–63. Available from: <http://dx.doi.org/10.1016/j.jamcollsurg.2007.11.020>
- 15 Brungardt JG, Liebscher SC, Schropp KP. Malrotation correction in the adult population. *World J Surg* [Internet]. 2021;45(1):141–7. Available from: <http://dx.doi.org/10.1007/s00268-020-05790-x>
- 16 Wei B, Huang Z, Tang C. Optimal treatment for patients with cavernous transformation of the portal vein. *Front Med (Lausanne)* [Internet]. 2022;9. Available from: <http://dx.doi.org/10.3389/fmed.2022.853138>
- 17 Hirano N, Iseki M, Morikawa T, Umino Y, Aoki S, Inoue K, et al. Pancreaticoduodenectomy for pancreatic head cancer with cavernous transformation of the portal vein: a case report. *Surg Case Rep* [Internet]. 2022;8(1). Available from: <http://dx.doi.org/10.1186/s40792-022-01463-0>
- 18 Intagliata NM, Caldwell SH, Tripodi A. Diagnosis, development, and treatment of portal vein thrombosis in patients with and without cirrhosis. *Gastroenterology* [Internet]. 2019;156(6):1582-1599.e1. Available from: <http://dx.doi.org/10.1053/j.gastro.2019.01.265>