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Case Report

Synchronous Volvulus of the Transverse Colon and Cecum Associated with Scleroderma and Lupus^{☆,☆☆}

Nicholas T. Manasewitsch, B.S.^{a,*}, Michael T. Lee, B.S.^a, Anthony N. Khuu, M.D., M.B.A.^b

^a University of Nevada, Reno School of Medicine, 1664 N. Virginia Street, Reno, NV 89557, USA

^b Department of Radiology, Renown Regional Medical Center, Reno, NV, USA

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ABSTRACT

Colonic volvulus may infrequently occur in the transverse colon, and synchronous double volvulus is a rarely reported phenomenon in the literature. Additionally, intestinal volvulus is a rare but serious complication that has been reported in scleroderma and systemic lupus erythematosus (SLE) patients. We report a 26-year-old patient with a history of scleroderma-SLE overlap syndrome who presented with an acute abdomen. CT imaging revealed a transverse colon volvulus and a cecal bascule (cecal volvulus). To our knowledge, this is the first reported case of synchronous double volvulus of the transverse colon and cecum. Additionally, this is the second reported case of transverse colon volvulus occurring in a patient with scleroderma and the first case in a patient with scleroderma-SLE overlap syndrome.

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Introduction

The sigmoid colon and cecum are the most frequently reported sites of colonic volvulus. Transverse colon volvulus (TCV) is rare and is estimated to comprise 3% of all cases of colonic volvulus [1]. There are approximately 100 cases of TCV reported in the medical literature with many of them discovered at the time of surgery [2]. Synchronous double volvulus of the sigmoid colon and cecum has been reported in the literature [3,4], but never a synchronous double volvulus of the transverse colon and cecum. We present the case of a 26-year-old woman with a history of scleroderma-systemic erythe-

matusus lupus (SLE) overlap syndrome who was discovered to have synchronous volvulus of the transverse colon and cecum on CT imaging. Additionally, we review the radiographic findings of TCV.

Case presentation

A 26-year-old American Indian woman presented with a 4-day history of generalized abdominal pain, nausea, vomiting, and constipation. Her past medical history was significant for scleroderma-SLE overlap syndrome, critical lower

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* Corresponding author.

E-mail address: nmanasewitsch@gmail.com (N.T. Manasewitsch).

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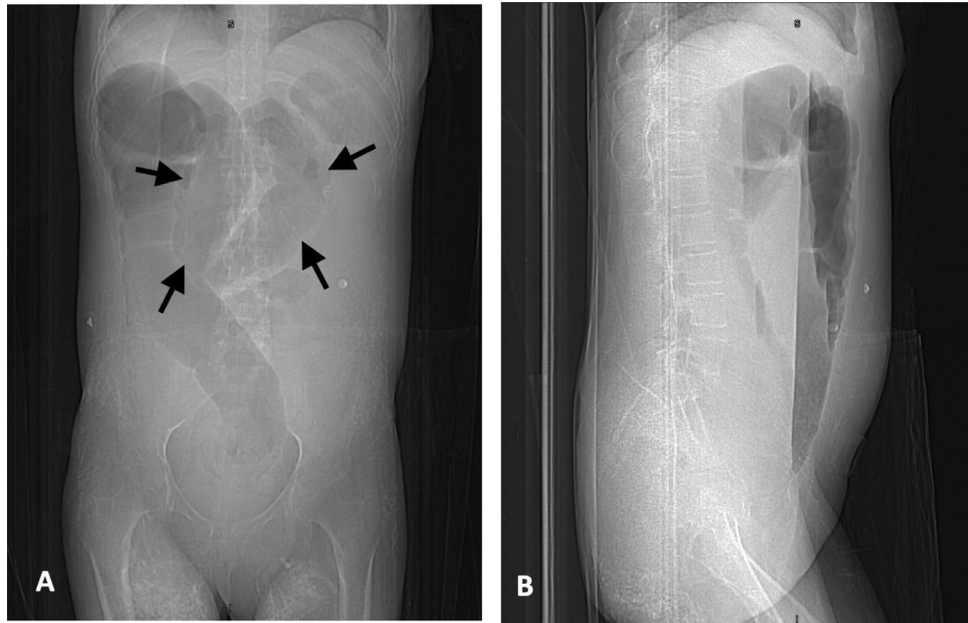


Fig. 1 – CT scout images of the abdomen and pelvis. Coronal view demonstrates a markedly dilated loop of transverse colon (arrows). The colon proximal to the loop is dilated and filled with stool. (B) Sagittal view demonstrates markedly distended bowel.

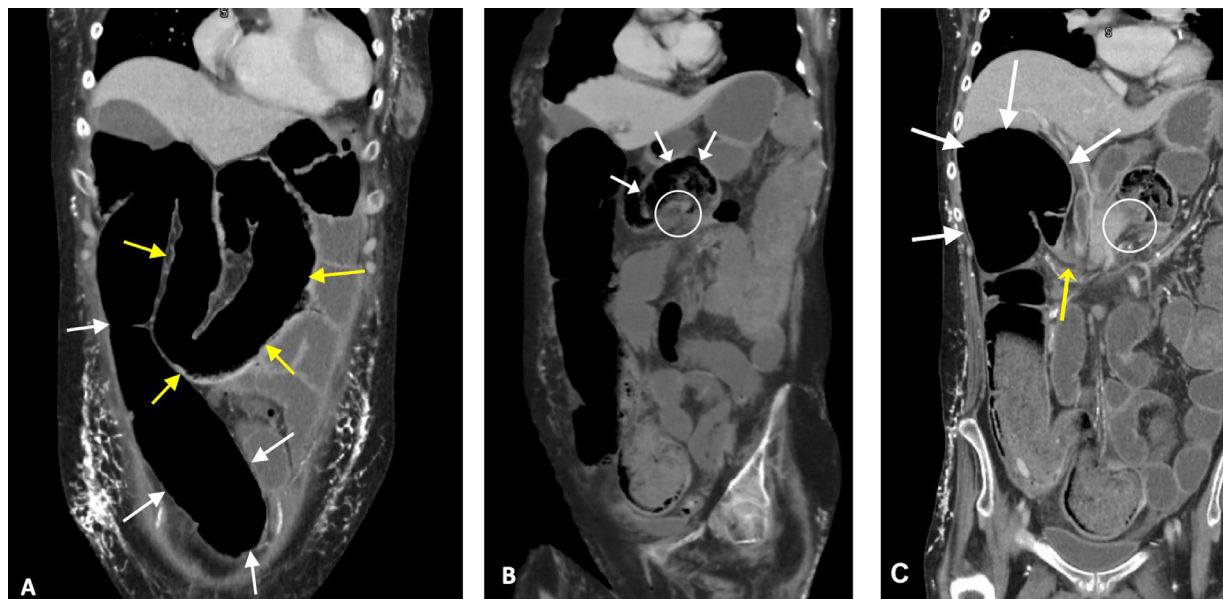


Fig. 2 – CT of the abdomen and pelvis with IV contrast on presentation. (A) Coronal view demonstrates extensive bowel dilatation and loop of transverse colon with “coffee bean” appearance (yellow arrows). There is extensive dilatation of the redundant ascending colon (white arrows). The descending and sigmoid colon are severely decompressed. (B) Oblique coronal view demonstrates a swirling appearance of the mesentery about the transverse colon (circles), consistent with the “swirl” sign. Additionally, there is a small closed loop of transverse colon (arrows). (C) Coronal view demonstrates a cecum that is folded anteromedially and displaced superiorly, consistent with a cecal bascule (white arrows). At its widest portion, the cecal bascule measures 10.0 cm in diameter. There is an abrupt cut-off with “bird’s beak” tapering at the distal cecum (yellow arrows). The “swirl” sign is again noted (circles). The small closed loop of transverse colon is again seen. The redundant ascending colon is filled with stool. (D) Axial view demonstrates a “swirl” sign of the terminal ileum, indicating bowel rotation around its mesentery (arrow). The colon is markedly distended. (E) Sagittal view again demonstrates markedly distended ascending colon which is filled with stool. Subcutaneous calcifications are noted throughout the lower abdominal wall and pelvis secondary to scleroderma (arrows).

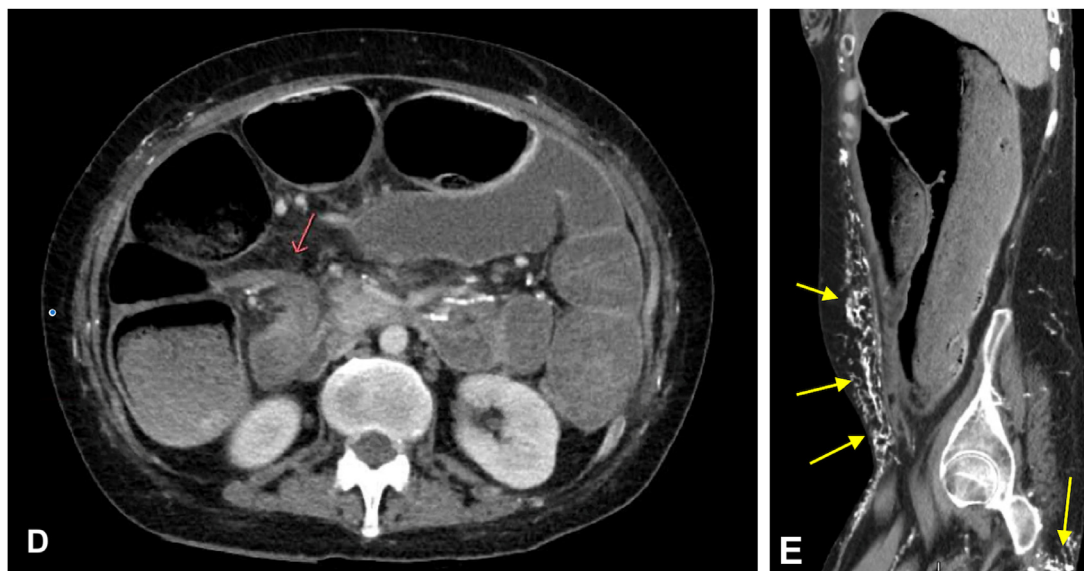


Fig. 2 – Continued

leg ischemia secondary to vasculitis, hypothyroidism, chronic kidney disease, alcoholic pancreatitis, and diet-controlled noninsulin dependent (type II) diabetes. Previous rheumatologic workup was remarkable for the presence of antinuclear antibody, anti-dsDNA, anti-U1-RNP, antiparietal cell, antiscleroderma-70 (antitopoisomerase I), and antismooth muscle antibodies. Rheumatologic symptoms included mask-like facies, diffuse skin tightening, sclerodactyly, Raynaud's phenomenon, multiple finger ulcerations, calcinosis, facial rash, oral ulceration, and photosensitivity. Five days prior, she underwent an amputation of vasculitis-induced necrosis of the left toes. She had no prior history of abdominal surgery. Her medication regimen included prednisone, hydroxychloroquine, mycophenolate mofetil, levothyroxine, and nifedipine.

Vital signs were unremarkable. The patient was ill-appearing in acute distress, and physical exam was remarkable for abdominal distension and left-sided tenderness to palpation with guarding. Laboratory investigation (reference range in parentheses) revealed white blood cell count: 14.9 K/ μ L (4.8–10.8 K/ μ L), neutrophils: 86.3% (44–72%), hemoglobin: 13.3 g/dL (12–16 g/dL), sodium: 129 mmol/L (135–145 mmol/L), potassium: 3.2 mmol/L (3.6–5.5 mmol/L), lactic acid: 2.7 mmol/L (0.5–2 mmol/L), and anion gap: 24 mmol/L (7–16 mmol/L).

A CT scan of the abdomen and pelvis with intravenous contrast revealed volvulus of the transverse colon and a cecal bascule (Figs. 1 and 2). There was extensive bowel dilatation and a loop of transverse colon with a “coffee-bean” or “bent inner tube” appearance (Fig. 2A). The CT scan also showed a twisting appearance, or “swirl” sign, and a closed loop obstruction of the transverse colon (Fig. 2B and D). There was also a cecal bascule with the terminal ileum and cecum displaced superiorly in the right upper quadrant with associated cecal volvulus (Fig. 2C). In addition, there was an abrupt cut-off with “bird’s beak” tapering at the distal cecum (Fig. 2C). Incidentally, extensive subcutaneous calcifications were noted throughout the

lower abdominal wall and pelvis which was secondary to patient’s known scleroderma-SLE overlap diagnosis (Fig. 2E).

The patient was urgently taken to the operating room for an exploratory laparotomy and extended right hemicolectomy. Intraoperative findings confirmed volvuli of both the transverse colon and cecum caused by a densely adherent omentum. There was no evidence of mass, stricture, or adhesions. Pathology specimen examination was consistent with resection of a necrotic cecum and transverse colon. Two days later, the patient underwent a second operation for creation of end-ileostomy and primary abdominal fascial closure. The patient was monitored in the intensive care unit.

The patient remained critically ill with gradual improvement until hospital day 10. A CT scan of the abdomen and pelvis with intravenous contrast (Fig. 3) was performed due to nausea, vomiting, and leukocytosis. This scan revealed portal venous gas, pneumatosis of the duodenum, and multiple loops of dilated small bowel. The patient had progressive and significant respiratory failure, and an emergency intubation was performed on hospital day 13. The patient was unable to be oxygenated despite the intubation, and she developed cardiac instability, hypotension, and cardiac arrest. The patient could not be resuscitated and unfortunately passed away.

Discussion

To the best of our knowledge, this is the first reported case of synchronous double volvulus of the transverse colon and cecum. The diagnosis was made radiographically and subsequently confirmed intraoperatively. There are few reported cases of synchronous double volvulus involving the sigmoid colon and cecum [3,4]; however, none have reported concurrent involvement of the transverse colon and cecum.

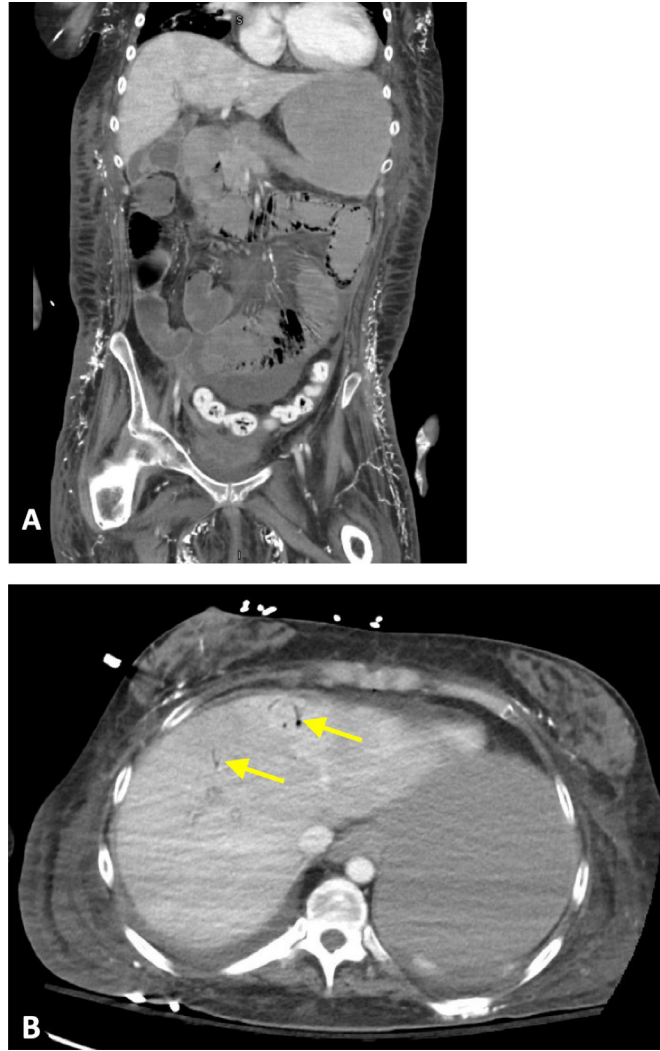


Fig. 3 – CT of the abdomen and pelvis with IV contrast on hospital day 10. (A) Coronal view reveals pneumatosis intestinalis with multiple loops of dilated small bowel. (B) Axial view demonstrates portal venous gas (arrows).

Additionally, this is the first reported case of TCV occurring in a patient with scleroderma-SLE overlap syndrome. There are few reports of intestinal volvulus in pure scleroderma patients and SLE patients [5–9], and only one previous case involved the transverse colon [6].

Predisposing factors leading to TCV include anatomic, mechanical, and physiologic causes. Anatomic causes include colon redundancy and nonfixation. Mechanical causes include distal obstruction, adhesions, inflammatory strictures, carcinoma, pregnancy, malrotation, and Chilaiditi syndrome. Physiologic causes include bowel distension due to chronic constipation [1,10,11].

Radiographic findings of a TCV include a “coffee bean” sign or “bent inner tube” sign representing the appearance of a closed-loop obstruction [1,12]. In our patient, the “coffee bean” sign (Fig. 2A) was seen in the upper abdomen with decompression of the descending and sigmoid colon. Importantly, TCV extends superiorly in the abdomen in a U-shape and does not originate in the pelvis, distinguishing it from the more

common sigmoid colon volvulus [12]. TCV may also present with a “swirl” or “whirlpool” sign. The “swirl” sign illustrates bowel twisted around a singular point of mesentery that may compromise intestinal blood flow leading to ischemia, and the sign is highly specific for volvulus [1,13]. The “swirl” sign was present in our patient (Fig. 2B and D) and was seen located inferiorly to the TCV. The “bird’s beak” phenomenon may be present radiographically in TCV [11,14] and was also present in our patient (Fig. 2C). This sign demonstrates narrowing of the bowel lumen proximal to an obstruction. Although the “bird’s beak” sign is most clearly demonstrated with barium enema, this study is not recommended as it may delay treatment unnecessarily and increase morbidity [15].

TCV has a 33% mortality rate, which is higher than the mortality rate of cecal or sigmoid volvulus (10% and 21%, respectively) [15]. Bowel resection in the form of transverse colectomy or extended right colectomy is the treatment of choice and preferred over colopexy and endoscopic decompression [1,11].

Our case was unique because of the additional presence of a cecal bascule (Fig. 2C). A cecal bascule is a variant of cecal volvulus that lacks the classic axial twisting. Rather, a cecal bascule is caused by anterior or antero-medial folding of the cecum over itself. This creates a flap valve that causes an obstruction [16].

There are few reports in the literature of intestinal volvulus in patients and SLE patients [5–9], with one report of a TCV in a pure scleroderma patient [6]. Overlap syndrome refers to patients who fulfill the criteria for at least two rheumatologic disorders including SLE, rheumatoid arthritis, scleroderma, polymyositis, dermatomyositis, and Sjögren syndrome [17,18]. Our patient met the criteria for scleroderma-SLE overlap syndrome given her clinical symptoms and antibody tests. A combination of scleroderma-induced colonic dysmotility, postoperative ileus, and postoperative opioid use following recent surgery may have led to colonic elongation and redundancy which predisposed to her double volvulus. Although many cases of TCV are diagnosed intraoperatively, our patient was diagnosed radiographically which allowed for urgent surgical intervention.

Conclusion

Synchronous double volvulus of the transverse colon and the cecum has never been reported in the medical literature. This diagnosis requires a high degree of suspicion to diagnose and necessitates urgent surgical intervention. It is important to be aware of the radiologic differences between different volvuli. Additionally, colonic volvulus is a rare but serious complication in scleroderma and SLE patients, and intestinal volvuli should be considered in the differential diagnosis for such patients presenting with acute abdomen. Further research is required to explore the pathophysiology and mechanism behind intestinal volvuli in scleroderma and SLE patients.

Consent for publication

Written informed consent for publication of their clinical details and clinical images was obtained from the patient's next of kin.

REFERENCES

[1] Kapadia Muneera R. Volvulus of the small bowel and colon. *Clin Colon Rectal Surg.* 2017;30(1):40–5. doi:10.1055/s-0036-1593428.

- [2] Hasnaoui Hamza, et al. Transverse colon volvulus presenting as bowel obstruction: a case report. *J Med Case Rep.* 25 May. 2019;13(1):156. doi:10.1186/s13256-019-2080-1.
- [3] Islam S, Hosein D, Bheem V, Dan D. Synchronous volvulus of the sigmoid colon and caecum, a very rare cause of large bowel obstruction. *BMJ Case Rep.* 2016;2016:bcr2016217116. Published 2016 Oct 14. doi:10.1136/bcr-2016-217116.
- [4] Roy SP, Tay YK, Kozman D. Very rare case of synchronous volvulus of the sigmoid colon and caecum causing large-bowel obstruction. *BMJ Case Rep.* 2019;12(1) bcr-2018-227375. Published 2019 Jan 28. doi:10.1136/bcr-2018-227375.
- [5] Anderson M F, Pickney L. “The traveling volvulus. *Dis. Colon Rectum* 1979;22(4):276–8. doi:10.1007/BF02586895.
- [6] Budd D C, et al. “Transverse colon volvulus associated with scleroderma. *Am. J. Surg.* 1977;133(3):370–2. doi:10.1016/0002-9610(77)90547-5.
- [7] Hendy M S, et al. “Small-bowel volvulus in association with progressive systemic sclerosis. *Br. Med. J.* 1979;1(6170):1051–102. doi:10.1136/bmj.1.6170.1051.
- [8] Plummer J M, et al. “Splenic flexure volvulus associated with systemic lupus erythematosus: A case report. *West Indian Med. J.* 2006;55(2):125–6. doi:10.1590/s0043-31442006000200013.
- [9] Yang Yunfei, et al. Lupus causing small bowel obstruction. *BMJ case reports* 2018;2018 bcr2018225886. doi:10.1136/bcr-2018-225886.
- [10] Abdulla Hussain Adnan, et al. “Transverse colon volvulus in a patient with sickle cell disease. *BMJ case reports* 2019;12(3) e228863. doi:10.1136/bcr-2018-228863.
- [11] Sage Mark J, et al. “Colopexy as a treatment option for the management of acute transverse colon volvulus: a case report. *J Med Case Rep.* 2012;6:151. doi:10.1186/1752-1947-6-151.
- [12] Feldman D. The coffee bean sign. *Radiology* 2000;216(1):178–9. doi:10.1148/radiology.216.1.r00j117178.
- [13] Duda JB, Bhatt S, Dogra VS. Utility of CT whirl sign in guiding management of small-bowel obstruction. *AJR Am J Roentgenol* 2008;191(3):743–7. doi:10.2214/AJR.07.3386.
- [14] Sparks Dorothy A, et al. “Ischemic volvulus of the transverse colon: A case report and review of literature. *Cases journal* 2008;1(1):174. doi:10.1186/1757-1626-1-174.
- [15] Loke K L, Chan C S. “Case report: transverse colon volvulus: unusual appearance on barium enema and review of the literature. *Clin. Radiol.* 1995;50(5):342–4. doi:10.1016/s0009-9260(05)83431-2.
- [16] Shah F, Elsayed M, Sekhar A. Cecal bascule as a cause of postoperative nausea and abdominal pain. *Radiol Case Rep.* 2019;14(6):697–9 Published 2019 Mar 27. doi:10.1016/j.radcr.2019.02.010.
- [17] Foocharoen Chingching, et al. Clinical characteristics of scleroderma overlap syndromes: comparisons with pure scleroderma. *Int J Rheum Dis.* 2016;19(9):913–23. doi:10.1111/1756-185X.12884.
- [18] Sharma Sanchit, Kumar Uma. “Scleroderma overlap syndromes. *Int J Rheum Dis.* 2016;19(9):831–3. doi:10.1111/1756-185X.13011.