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Absence of Ascending Colon, Persistent Transverse Colon, and Highly Movable Cecum: A Rare Congenital Abnormality with High Risk of Volvulus Formation

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Background	A male patient presented with severe abdominal pain, constipation, nausea and vomiting six years following diagnosis of congenital abnormality of the colon.
Summary	Cecal volvulus is uncommon in adults and there are limited reports that describe the underlying embryologic pathology. Here, we report the case of a 43-year-old male patient who presented with severe abdominal pain. He had a similar episode earlier in life and was told he had a rare congenital abnormality but no surgical interventions were done at that time. Upon his current presentation computed tomography (CT) scan without contrast enhancement showed dilation of the transverse colon with congenital absence of the right colon. Given this rare congenital abnormality suspicion for cecal volvulus was made. The patient underwent an emergent surgery that revealed cecal volvulus. Cecal volvulus usually occurs following intestinal malrotation, previous surgery or absence of fixation of the ascending colon to the posterior abdominal wall. However, in this patient there was a total absence of ascending colon. This abnormality of the right colon might have contributed to highly movable cecum that resulted in volvulus.
Conclusion	Various authors described anatomical variations of the colon, but absence of ascending colon is rare and has scant previous reports. We present a rare case of congenital anomaly where the ascending colon was totally absent, which contributed to development of cecal volvulus.
Keywords	Cecal volvulus, congenital abnormality, absent ascending colon

DISCLOSURE STATEMENT:
The authors have no conflicts of interest to disclose.

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Case Description

Congenital anomalies such as volvulus of the cecum can cause an incomplete obstruction, which may not manifest until later in life. These abnormalities are frequently found incidentally on CT or MRI. Specifically, cecal volvulus is uncommon in adults and extremely rare in childhood. It accounts for <1% of all cases of intestinal obstruction and 25 to 40% of the cases of volvulus of the colon.[1] Presence of mesocolon for ascending or descending colon and very long colon are some of the predisposing factors for a volvulus formation.[2] We report a rare case of a congenitally absent right colon, which resulted in a volvulus at later stage in life.

The patient is a 43-year-old male with history of hypertension and no previous surgeries. The patient reported a history of severe persistent abdominal pain, nausea, vomiting, and constipation. He had one similar episode six years ago at which point he was told that he had a congenital abnormality, but no surgery was done at that time. A CT scan was requested, which showed a definite cecal volvulus with the cecum up to 13 cm in size (Figure 1).

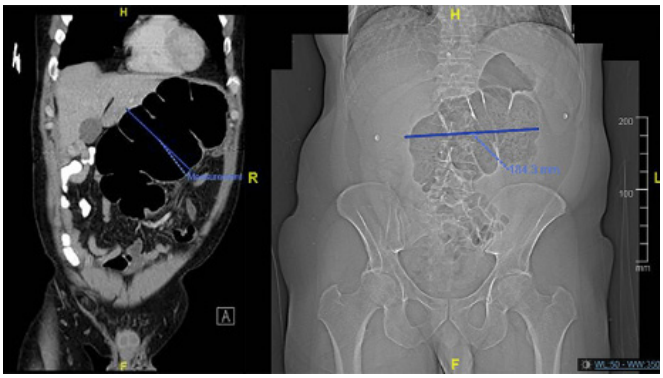

Figure 1. Congenital malrotation of the bowel with cecal volvulus measuring 13 cm in size

Nonsurgical management (e.g., colonoscopy, reduction barium enema) for correction of a cecal volvulus is not recommended, as these treatments have an increased risk of colon perforation and have failure rates approaching 95%.[3] Thus, an emergent surgery was planned. He had a normal white count. At the time of surgery, he was afebrile.

Upon entering the abdomen, there was a very large cecal mass that was the size of a small football (Figure 2). This was gently mobilized into the operative area. The terminal ileum was located using an appendix, which appeared to be normal. An attempt was made to mobilize the right colon, but it was not until the colon was traced from the transverse colon that the abnormality was discovered. There was no hepatic flexure and no right colon in the actual position. The transverse colon was mobilized, but it did not seem to go up into the right upper quadrant. It was seen that two mesenteries were continuous and the right end of the transverse colon was more mobile than normally. Once the adhesions where lysed the splenic flexure was identified by going along the white line of Toldt and also dividing some omentum. The left colon ascended up to splenic flexure and the transverse colon was not in proper position. It descended straight down in the lower abdomen connecting to the cecum in almost a diagonal fashion. After this was fully mobilized, terminal ileum was divided about 5 to 6 cm proximal to the cecum with GIA-80 stapler. The colon was also divided with GIA-80 and was just removed. A small enterotomy was made and the transverse colon was attached to the terminal ileum with GIA-80 stapler and then closing with TA60 stapler. The anastomosis was widely patent; several 3-0 Vicryl sutures were put at the apex to take off any tension. The suture line was also reinforced with 3-0 Vicryl sutured. The mesenteric defect was with continuous suture of 3-0 Vicryl suture. The small bowel was placed back in the abdomen, what would be considered an anatomic position as the transverse colon actually went across the lower abdomen.

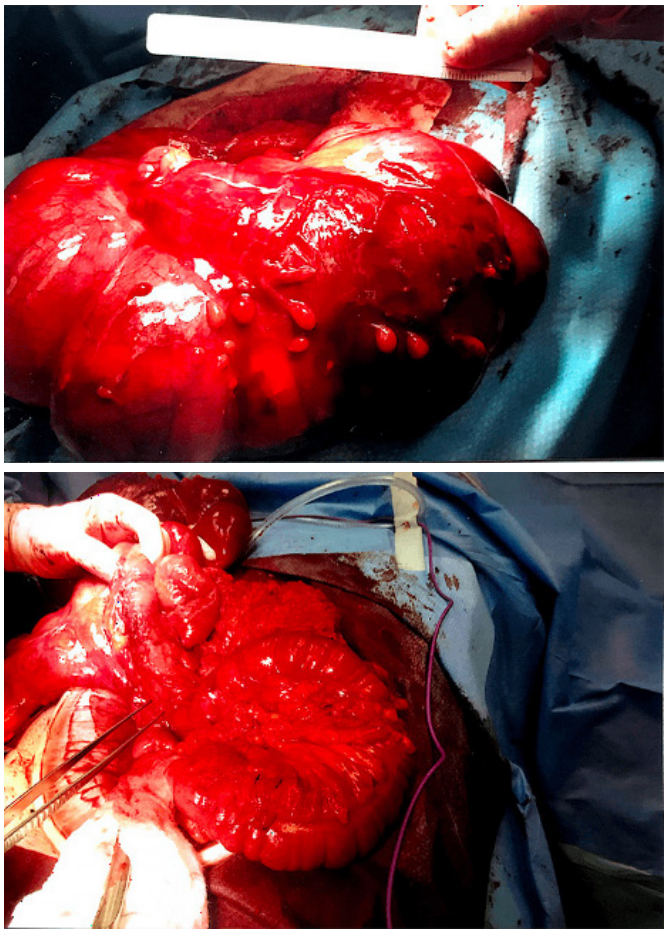

Figure 2. Anterior view of anomalous colon in situ after reflection of anterior abdominal wall. Greater omentum has been reflected upwards. Note the size of the cecum. Jejunum and ileum were occupying right part of the abdomen and transverse colon was found in a diagonal position.

Positional variations of the colon are dependent on midgut rotation during its development. There are three stages of midgut development (Figure 3). Initially the midgut loop rotates 90° in an anticlockwise manner as it enters the umbilical cord. During week 10 of intrauterine life, the intestines return back to the abdomen from physiologic hernia. At this point the cecum and appendix lie near the right lobe of the liver. Lastly, the proximal portion of the colon lengthens, giving rise to the hepatic flexure and ascending colon as the cecum descends from the upper to the lower right side of the abdomen into the right iliac fossa. The total range of rotation around superior mesenteric artery is about 270°. As the intestines settle into their final positions, their mesenteries undergo a process of fixation, where mesenteries press against the back wall of the abdominal cavity.[4] Defective fixation can result in ascending and descending colon to retain their mesentery. In the current case, the ascending colon has failed to descend to the right and lose its mesentery. This could be the possible reason for the absence of right ascending colon and abnormal position of the transverse colon. In 1912 Mummery[6] has described a case in which both the cecum and ascending colon have failed to develop. Moreover, cases of right sided descending and sigmoid colons,[7] sessile ileum and subhepatic caecum[8] have been reported. However, in our literature survey, we could not come across any specific case of isolated absence of ascending colon and we assume that this is the first report on the absence of ascending colon.

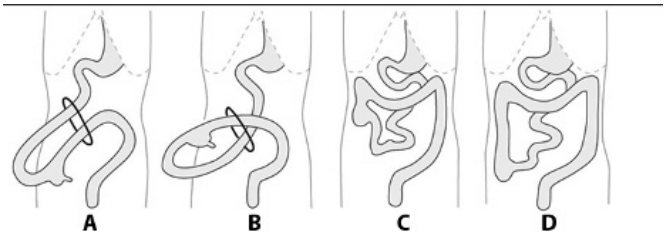

Figure 3. Developmental stages of midgut rotation. In A, the duodenum has completed a 90° counterclockwise rotation to position to the right of the superior mesenteric artery. In B, the duodenum has rotated another 90° counterclockwise. In C, the final the 90° counterclockwise rotation is completed by duodenum. The cecum continues to rotate and future to be ascending colon descends. In D, the normally rotated bowel is depicted.[5]

In this report, absence of ascending colon resulted in a diagonal position of transverse colon, with the proximal part being attached to cecum. When cecum fails to descend and undergo fixation it retains its own mesentery making it freely movable. This abnormality gives rise to the cases of cecal volvulus. A much unusual condition is for the cecum to be arrested under the liver and for there to be no ascending colon. A cecal volvulus is defined as the rotation of a flexible cecum and ascending colon, frequently progressing to bowel obstruction, ischemia, necrosis, and perforation. Congenital mobile cecum is hypothesized to result from failed fusion of the ascending colon mesentery to the posterior parietal peritoneum. Based upon autopsy studies, approximately 10 to 25 percent of the population has a cecum and ascending colon with sufficient mobility to develop a volvulus.[9]

In general, the mobility of the right colon is very great. Instead of being fixed, the right colon is found to possess a mesentery of varying length and, in addition to the true mesentery, there usually develops a backward continuation and elongation of it, the pseudomesentery, a firm dense structure coming from the fascia of posterior abdominal

wall.[10] The small bowel mesentery is normally broad based, with its attachment extending from the ligament of Trietz to the ileocecal valve. This wide base prevents the small intestine from twisting around the superior mesenteric artery. However, in our case the mesentery of the small intestine was very short and ileum and jejunum occupied most of the right part of the abdomen. Short mesentery and limited space for the small intestine in the abdomen might also lead to its abnormal movements leading to the formation of a volvulus.[11,12]

To the best of our knowledge, total absence of ascending colon has not been reported yet. Absence of ascending colon, presence of a diagonally placed transverse colon and displacement of small and large intestines as reported here result in abnormal movements predisposing to volvulus. Absence of ascending colon poses problems in investigation, diagnosis, and intervention. The knowledge of this variation is very useful for the radiologist, gastroenterologists and surgeons in general.

Conclusion

Various authors described anatomical variations of the colon, but absence of ascending colon is rare and has not been reported. We present a rare case of congenital anomaly where the ascending colon was totally absent, which contributed to development of cecal volvulus.

Lessons Learned

Congenital abnormalities predispose to intestinal malrotations. Normal embryologic development has three stages followed by fixation of the mesentery to the abdominal wall. Failure of fixation and decent contributed to the absence of the right ascending colon with highly mobile cecum, which later developed into volvulus.

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