

Cecal volvulus in situs inversus totalis accompanied with pancreatic malrotation

Ahmet Önder, Nazan Okur, Ertan Bülbüloğlu, Mehmet Fatih Yüzbaşıoğlu

ABSTRACT

Situs inversus totalis is a rare anomaly that is characterized by mirror image anatomy of the abdominal and thoracic organs. This anomaly is often discovered incidentally in adults during an emergent surgical procedure or by radiologic investigations performed for other medical reasons. Cecal volvulus is a surgical emergency requiring urgent laparotomy. Coexistence of these two abnormalities poses a challenge in establishing a definitive diagnosis because of the alteration of normal anatomy; this may cause a delay in emergent surgical intervention. We present a case of situs inversus totalis complicated by cecal volvulus. In our case, a different type of pancreatic malrotation, which we called "folded", also accompanied these abnormalities. To our knowledge, this is the first case in the literature involving situs inversus totalis, cecal volvulus, and a "folded" type of pancreatic malrotation.

Key words: • intestinal volvulus • situs inversus
• pancreas, abnormalities

Situs inversus totalis is a congenital anomaly (1). It is defined as a complete mirror image of the thoracic and abdominal viscera. It does not seem to affect normal health or life expectancy, and it is not considered to be premalignant (1–5). It is often detected incidentally by radiological investigations. In fact, there is little information about the abdominal manifestations of situs anomalies in adult (1, 2).

Volvulus of the cecum is a surgical emergency caused by the axial twist of the cecum, distal ileum, and proximal colon in the absence of normal cecal fixation (6, 7). It is a rare cause of intestinal obstruction (6, 7). Coexistence of situs inversus totalis and cecal volvulus poses a challenge in establishing an accurate diagnosis, and may cause a delay in emergent surgical intervention. Thus, the mortality and morbidity risk may increase. We report a case of situs inversus totalis complicated with cecal volvulus, accompanied by a different type of pancreatic malrotation.

Case report

A 26-year-old female was admitted to our hospital with a 4-day history of crampy, diffuse abdominal pain, nausea, and vomiting. There had been no gas or stool passage during this period. The patient had been treated conservatively in another hospital for two days. She reported having similar complaints two months earlier, with relief of symptoms following conservative treatment. On physical examination, heart sounds were heard on the right side of the chest. The abdomen appeared to be slightly distended, with diffuse tenderness on palpation, especially over the left upper quadrant, but there were no signs of peritoneal irritation. Bowel sounds were increased on the right side of the abdomen. The rest of the examination was normal. Initial laboratory tests were unremarkable, except for moderate ketonuria. Plain abdominal radiograph revealed dilated bowel loops and air-fluid levels throughout the abdomen, most prominently in the left upper quadrant. Dextrocardia was also noted on the plain radiograph.

Computed tomography (CT) showed mirror image transposition of the abdominal and thoracic viscera (situs inversus totalis) relative to situs solitus (Fig. 1a). Transaxial images demonstrated enlarged bowel loops with air-fluid levels. The digital topogram showed a markedly distended colonic gas shadow extending from the left upper quadrant down to the midabdomen (Fig. 1b). This is a sign of cecal volvulus called *comma-shaped gaseous distension*. There was no gas shadow to the left lateral aspect of this colonic segment. Axial images also showed remarkable distension and air-fluid level in the cecum, known as the *coffee bean sign* (Fig. 2). Also seen in the lower abdomen was the *bird's beak sign* (marking the point of torsion) (Fig. 2). Another interesting finding was the position and configuration of the pancreas. While the pancreatic uncus and

From the Department of Surgery (A.Ö.), Birecik Government Hospital, Şanlıurfa, Turkey; the Department of Radiology (N.O.), Afyon Kocatepe University School of Medicine, Afyon, Turkey; and the Department of Surgery (E.B. ✉ ertanbulbuloglu@yahoo.com, M.F.Y.), Sütçü İmam University School of Medicine, Kahramanmaraş, Turkey.

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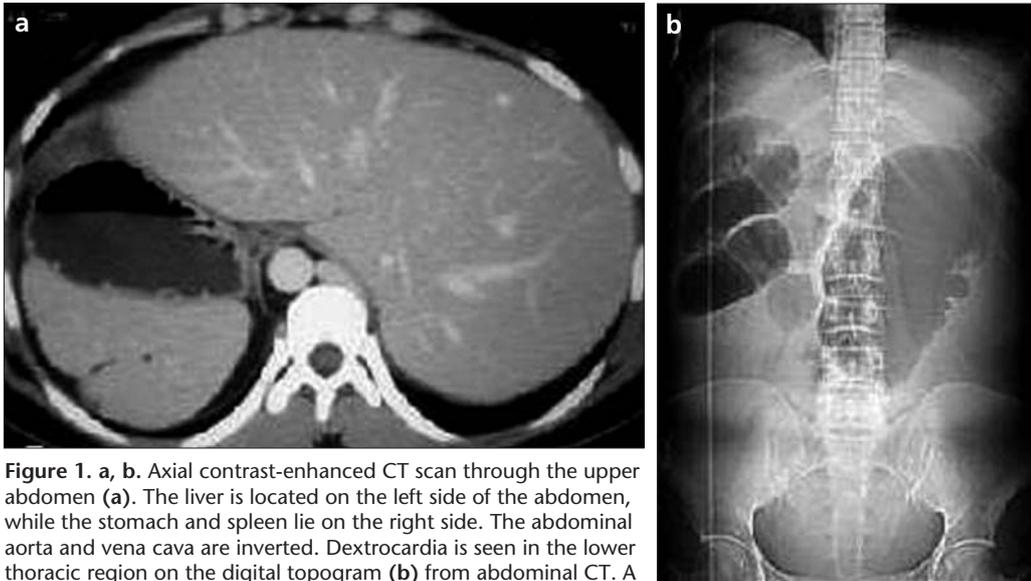


Figure 1. a, b. Axial contrast-enhanced CT scan through the upper abdomen (a). The liver is located on the left side of the abdomen, while the stomach and spleen lie on the right side. The abdominal aorta and vena cava are inverted. Dextrocardia is seen in the lower thoracic region on the digital topogram (b) from abdominal CT. A very dilated cecal gas shadow is seen extending from the left upper quadrant down to the midabdomen (comma-shaped distension). Haustral creases are preserved. There is no gas shadow to the left of this segment of the colon.

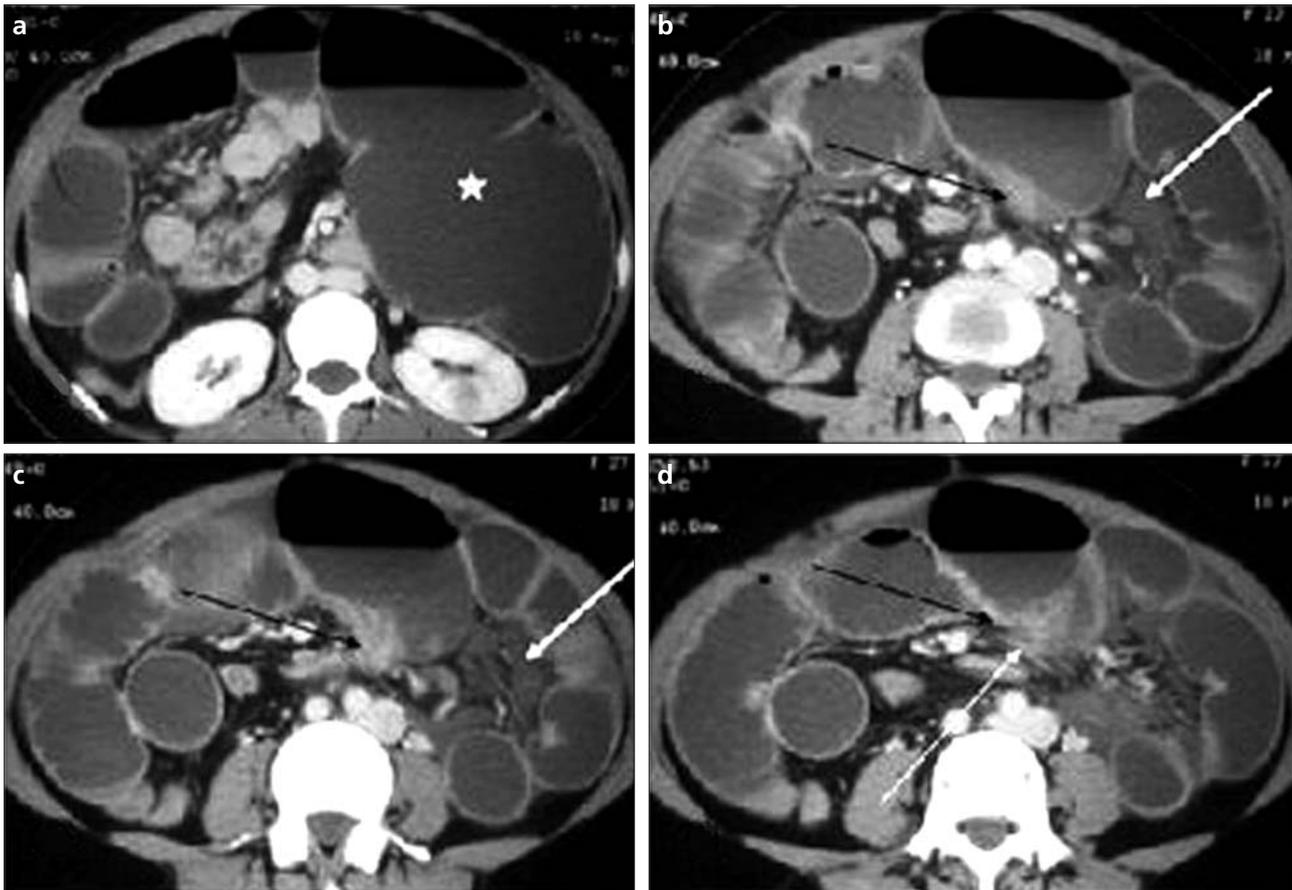


Figure 2. a–d. Axial abdominal CT images (a–d). Through the level of the kidneys, markedly distended cecum filled with air and fluid is demonstrated in the left half of abdomen (coffee bean sign, *star*) (a). Inferiorly, the cecum is terminated, tapering towards the point of torsion (bird's beak sign, *black arrow*) (b–d). Also note the distortion of the mesenteric vessels (*white arrow*, d) and fluid attenuation of the fatty mesentery (*thick white arrow*, b, c). There are dilated, fluid-filled small intestinal loops interposed between the abdominal wall and the cecum.

head were to the left of the midline, being positioned left posterolateral to the inverted superior mesenteric vein,

the pancreatic body and tail were also positioned on the left side, extending obliquely down to the neighborhood

of the left renal vein. The pancreatic tissue was folded on itself at the neck (pancreatic malrotation) (Fig. 3).

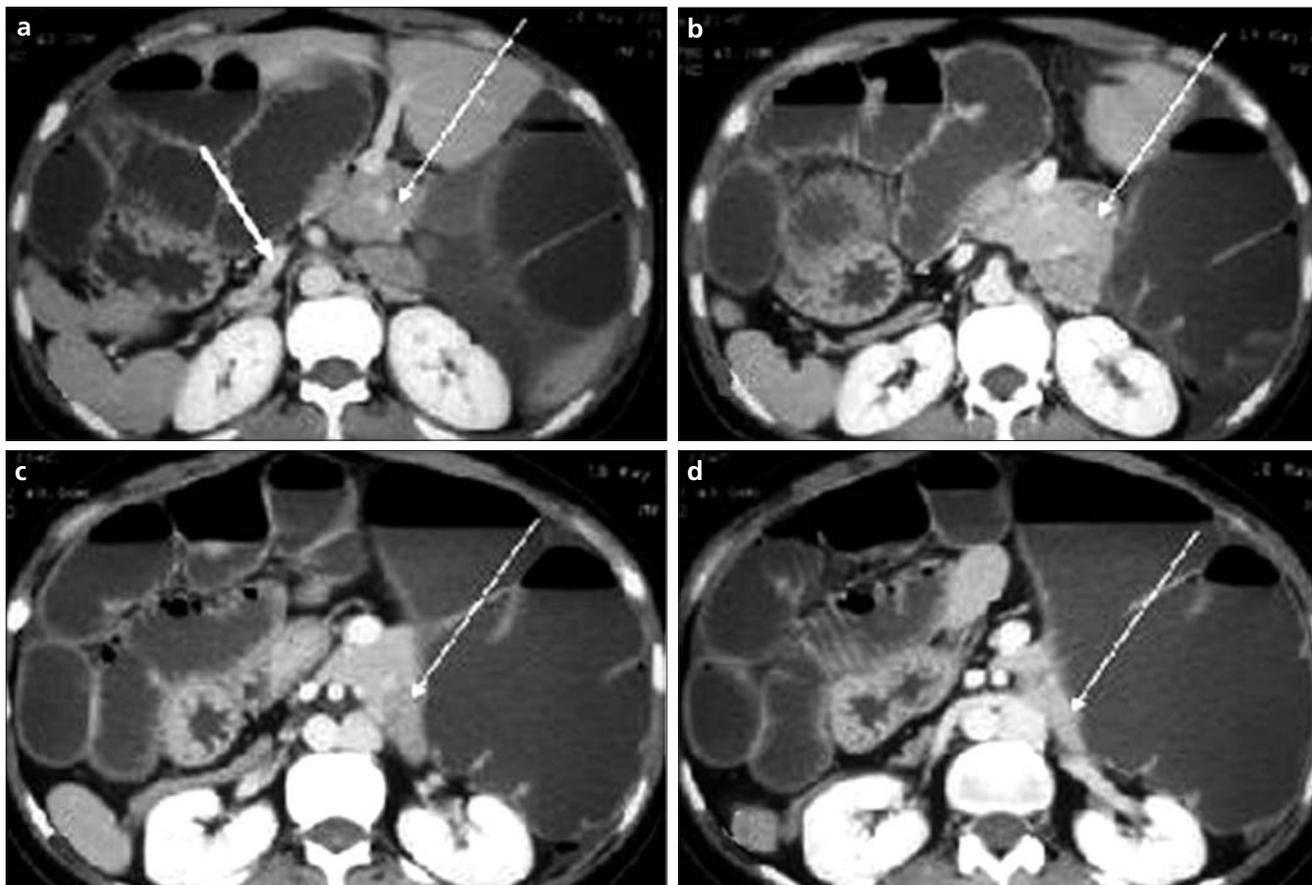


Figure 3. a–d. Axial abdominal CT images through the pancreas (a–d). Mirror image reversal of the pancreatic uncus is seen (*thin arrow, a*). There is no pancreatic tissue anterior to the splenic vein (*thick arrow, a*). Pancreas is folded on itself at the level of neck (*arrow, b*), which lies to the left of the superior mesenteric vein, and then extends caudally to the neighborhood of the left renal vein (*arrow, c, d*) (pancreatic malrotation).

On the second day of admission, the patient developed mild tachycardia (92 beats/minute), elevated body temperature (38.2 °C), and leukocytosis (white blood cells 16,300/ μ L). Laparotomy was performed, establishing the diagnosis of cecal volvulus. During surgical exploration, although the cecum was twisted and distended in the left upper quadrant, it was viable with no signs of ischemia or necrosis. Following detorsion of the cecum, appendectomy was performed. The lumen of cecum was aspirated through the appendiceal stump. After establishing the viability of the cecum, cecopexy was performed. The recovery was uneventful and the patient was discharged from the hospital on the third postoperative day. She has been followed as an outpatient for the last 10 months and has had no problems.

Discussion

Situs anomalies are variable, and it is important to define the relevant

anatomy before making more specific diagnosis, especially in complicated cases. Situs solitus is defined as the normal position of the heart and abdominal viscera, with the cardiac apex, spleen, stomach, and aorta located on the left, and the liver and inferior vena cava located on the right (1). Situs inversus is the mirror-image location of the viscera relative to situs solitus. It may occur with dextrocardia (the more common configuration) (2, 3), as was the case in our patient, or it may be associated with levocardia, a very rare variant. Situs ambiguus, or heterotaxia, on the other hand, is the abnormal arrangement of organs and vessels as opposed to the orderly arrangement typical of situs solitus and situs inversus. It represents a spectrum of abnormalities, and it may be associated with polysplenia or asplenia. Situs anomalies may be associated with intestinal rotational disorders (4). Although situs anomalies are embryologically related to each other, it is suggested that situs

inversus be differentiated from others because of its significantly different pathophysiologic and clinical implications (5). It is a rare anomaly with an incidence ranging from 1:35,000 to 1:1,400 (1, 2). Its recognition is important to avoid confusion, especially in the emergency setting, as in our case. Cecal volvulus is the result of abnormal mobility of the cecum caused by developmental improper fusion of cecal mesentery with the posterior parietal peritoneum (6). Patients with this condition may present with highly variable clinical presentations ranging from intermittent, self-limiting abdominal pain to acute abdominal pain associated with strangulation and sepsis (7). When strangulation is present, resection is mandatory. In the absence of strangulation, there are many treatment modalities recommended in the literature (8, 9). Endoscopic detorsion has a low overall success rate (12.5%) (10). Resection, cecopexy, cecostomy, and cecostomy with cecopexy may be

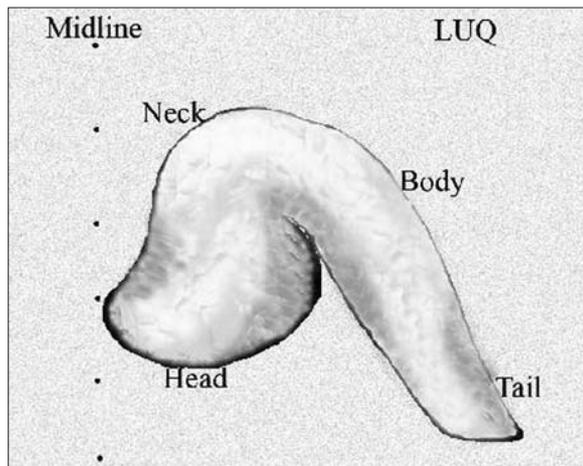


Figure 4. Schematic representation of the folded type of pancreatic anomaly (LUQ, left upper quadrant).

performed after laparotomy and detorsion. Resectional procedures have a significantly lower recurrence rate (8, 9). We avoided cecal resection and performed cecopexy because the cecum was edematous, and there was a considerable risk of anastomosis leakage if performed.

In cecal volvulus, the radiological findings that can be detected on plain radiographs are cecal dilatation, air-fluid levels, small bowel dilatation, absence of gas in distal colon, and dilated small bowel loops localized lateral to a dilated caecum (7, 11). Since CT is more commonly used as the imaging technique of choice in the evaluation of patients with acute abdominal pain, familiarity with the findings of cecal volvulus will help avoid confusion and delay in surgery. The bird's beak sign is the gradual tapering and convergence of the obstructed loop limbs to the point of torsion (12). The *whirl sign* has been described to be composed of spiraled loops of collapsed cecum, distal ileum, and enhancing engorged vessels (12, 13); and it is suggested that the degree of cecal rotation can be predicted by the tightness of the whirl (12). Though we detected the bird's beak sign, we could not appreciate a significant *whirl sign* in our case, suggesting a lesser degree of rotation; and, accordingly, the cecum was found to be viable at surgery. Although there are several case reports in the literature explaining cecal volvulus and situs anomalies independently, to the best of our knowledge, association of cecal volvulus with situs inversus totalis (from the clinical and radiological

points of view) has not previously been reported in the English literature. This makes our case an interesting and unusual one.

The diagnosis of cecal volvulus may be challenging even in patients with situs solitus. We aimed at both reviewing the radiological findings of cecal volvulus and also noting that cecal volvulus should be kept in mind in the differential diagnosis in patients with situs inversus totalis, presenting with acute abdominal pain. It may be helpful to adapt and interpret the radiological findings as mirror image reversal of those seen in situs solitus. For example, the configuration of the comma-shaped distension of the cecum in the digital topogram in our case was reversed, and the bird's beak sign at the point of torsion as well as mesenteric engorgement was to the left of the lower abdomen. The coffee bean sign was demonstrated in the left half of abdomen. Since it may be visualized anywhere within the abdominal cavity (7), being related to the type of the cecal volvulus (14), its location may not be helpful in assessing an intestinal rotation anomaly.

Another interesting feature of our case was the position and configuration of the pancreas, which was consistent with malrotation. Although pancreatic malrotation has been described in patients with heterotaxy syndromes (1, 15), to our knowledge, there is no reported pancreatic malrotation type in patients with situs inversus totalis. This makes our case unique. The most commonly encountered pancreatic anomaly in heterotaxy syndromes is

truncation; others include pancreatic cleft created by vessels traversing the pancreas, and short pancreas (1). Pancreatic truncation is defined as the presence of only the pancreatic head, or as the presence of the head and a small portion of the body. In patients with situs inversus totalis, the pancreas is regarded as normal when the head of the pancreas is to the left of the midline, with the tail extending towards the right upper quadrant. In our patient, the pancreatic head was to the left of the midline as expected, but the body and the tail were also located on the left. This folded type of pancreatic anomaly, as we named it, is schematically depicted on Figure 4. To our knowledge, this type of pancreatic malrotation has not been reported in the literature about situs inversus totalis or heterotaxy syndromes. Information regarding the pancreatic anomalies may be important for the adequate interpretation of clinical signs, radiological examination, and also for operator orientation when needed (16).

In summary, we reported an adult case of situs inversus totalis with a folded type of pancreatic malrotation not previously described. The patient also presented with cecal volvulus. Familiarity with the overlapping features of situs anomalies and also the possible anatomical alterations may be helpful in avoiding confusion in cases with acquired diseases.

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