Acute abdominal pain in midgut malrotation
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Section: Abdominal imaging
Area of Interest: Abdomen Gastrointestinal tract
Mesentery
Imaging Technique: CT
Case Type: Clinical Cases
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Patient: 20 years, male

Clinical History:
A 19-year-old man was admitted in the emergency department with an history of severe cramping abdominal pain and anorexia for two days. Upon examination, the abdomen was grossly distended. Lab results demonstrated leukocytosis (17,000/mL) with mild neutrophilia. Abdominopelvic contrast-enhanced computed tomography (CT) examination was performed.

Imaging Findings:
Scanogram showed a 9-cm dilated air-filled loop of large bowel. Axial CT scan showed a markedly distended fluid- and gas-filled ectopic caecum, located in the left upper and mesogastric region of the abdomen, rising suspicious of caecal volvulus (Fig. 1). Moreover, other two features were observed: a) the “whirlpool sign” of jejunal loops and superior mesenteric vein (SMV) with its tributaries wrapped around the superior mesenteric artery (SMA) (Fig. 2); b) and the “bird beak sign”, i.e., tapered efferent and afferent bowel loops at the level of the volvulus seen in a more caudal region (Fig. 3). A right-sided small bowel, a left-sided distended caecum and a dilated SMV to the left of the SMA, a reversal of their normal anatomic relationship (Fig. 4) led to the diagnosis of caecal volvulus with midgut malrotation.

Discussion:
-Background: The caecum is the proximal portion of the colon following the ileocaecal valve, it measures 7.5–8 cm in length and since it is normally covered by peritoneum, has limited mobility [1]. Congenital anomalies of colonic fixation due to an improper fusion of the caecal mesentery with the retroperitoneum and consequently an abnormal motility of the right colon can predispose to volvulus [2]. Caecal volvulus is a rare cause of large bowel obstruction, it occurs when the caecum twists around its own mesentery and accounts for 25–40% of all cases of colonic volvulus [2] and for 1–3% of cases of intestinal obstruction in adults [3]. Usually, it affects patients younger (30–60 years of age) than those affected by sigmoid volvulus [4] and it is slightly predominant in males [5]. Caecal volvulus is also one of the possible clinic presentations of intestinal malrotation [6], a congenital anomaly consisting in lack of or incomplete rotation of the fetal intestines around the axis of the superior mesenteric artery.
-Clinical perspective: Symptoms of malrotation typically manifest within the first year of life, with bilious vomiting and intestinal obstruction [4]. More rarely, it may manifest in adulthood (0.2–0.5% of cases) where it has an insidious onset with nonspecific symptoms [7]. In an adult caecal volvulus as a cause of intestinal obstruction secondary to intestinal malrotation, is rare. Typically, symptoms mimic those of intestinal obstruction and consist in cramping pain, distention, constipation and vomiting [6]. This condition is associated to a high rate of mortality and morbidity, because of the common diagnostic delay due to the rarity of the clinical scenario and the anomalous anatomy, and because of the possible severe complications such as intestinal ischaemia and perforation [8].

-Imaging perspective: Investigations usually start with plain X-ray of the abdomen which shows a dilated gas-filled large bowel segment located ectopically (in the left upper quadrant or mid abdomen) producing the classic “coffee bean” image [5]. Abdominal CT is then mandatory for an accurate preoperative diagnosis. CT is the best diagnostic modality for differentiating caecal volvulus from its mimics, determining its location, evaluating associated anatomic variants, and identifying potentially fatal complications such as ischaemia or perforation [4]. On CT images, the abnormally positioned caecum often appears in the upper mid and left abdomen and can be traced back to the level of the volvulus, which appears as an area of swirling of the bowel and its mesentery: this finding is also known as the whirl sign [2]. Another common CT finding is the bird beak sign, referring to the tapering efferent and afferent bowel loops at the point of volvulus [4]. CT can also point out other coexistent abnormalities. In our case, the evidence of a reversal position of the SMV on the left of SMA instead on the right, a right-sided small bowel and left-sided colon were highly suspicious for midgut malrotation associated to caecal volvulus [9].

-Outcome: Surgery is the only effective treatment of caecal volvulus, and the prognosis depends mainly on the viability of the caecum [4,5]. In our case, an urgent midline laparotomy was performed and a grossly dilated right colon, twisted along its mesentery, was visualised. Moreover, the small bowel loops were seen to be pushed in the right side of the abdomen and the Treitz ligament was not clearly visible, while the colon was mainly located in the left-side of the abdomen with an anomalous mobile mesentery (Fig. 5a). These findings confirmed the diagnosis of caecal volvulus associated with midgut malrotation from mesenterium ileocolicum commune. Since there was no evidence of vascular damage, detorsion of the caecal volvulus and colon cecopexy was performed (Fig. 5b). To note, when diagnostic delay leads to intestinal ischaemia, right colectomy is the only surgical option.

-Take home message: As is for most congenital gastrointestinal anomalies, intestinal malrotation usually manifests in early childhood. However, some patients may remain asymptomatic until adulthood. In adults, congenital anomalies are often mistaken for common abdominal conditions since they normally manifest with acute abdomen and non-specific symptoms. Imaging, especially CT, is particularly useful in identifying congenital intestinal anomalies in adults. It is important to know how they manifest to establish a prompt correct diagnosis and appropriate treatment.

Written informed patient consent for publication has been obtained.

Differential Diagnosis List: Caecum volvulus in a patient with congenital midgut malrotation, Appendicitis, Sigmoid volvulus , Small intestinal obstruction

Final Diagnosis: Caecum volvulus in a patient with congenital midgut malrotation

References:


Figure 1

Description: A dilated air-filled colon in the central region of the abdomen. Origin: © Unit of Radiology, IRCCS Policlinico San Donato, San Donato Milanese, Italy
Figure 2

Description: The whirlpool sign (arrow) of jejunal loops, SMV with its tributaries and mesenteric fat, wrapped around the SMA. Right-sided small bowel (*). Origin: © Unit of Radiology, IRCCS Policlinico San Donato, San Donato Milanese, Italy
Figure 3

Description: Dilated caecum (*) and appendix (arrowhead). Bird beak sign is also seen (arrow). Origin: © Unit of Radiology, IRCCS Policlinico San Donato, San Donato Milanese, Italy
Figure 4

Description: SMV (long arrow) is on the left of SMA (short arrow): a reversal of their normal anatomical relationship. Origin: © Unit of Radiology, IRCCS Policlinico San Donato, San Donato Milanese, Italy
Description: After derotation, right colon (RC) and caecum (C) standing on the left part of the abdomen, with ileum (IL) on the right. Dashed line shows the axis of rotation. Origin: © Unit of General and Foregut Surgery, IRCCS Policlinico San Donato, San Donato Milanese, Italy
Description: Colopexy with multiple running sutures (arrows) fixing right colon (RC), left colon (LC) and sigmoid (S). Origin: © Unit of General and Foregut Surgery, IRCCS Policlinico San Donato, San Donato Milanese, Italy