Unusual presentation of Acute Appendicitis: Report of a case

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Abstract

Developmental anomalies of mid gut are relatively rare. We present an atypical case of acute appendicitis which on exploration was found to be a sub-hepatic caecum with inflamed appendix and no development of right colon. The entire colon was found to be left sided with caecum in the sub-hepatic area and the inflamed appendix going behind the right lobe of liver. Few cases of Cadaveric non rotation of gut on autopsy have been reported but presentation as acute appendicitis is relatively rare.

Key wards: Acute appendicitis, inflamed appendix

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Introduction

Acute appendicitis can be at times challenging to operate particularly in case of non rotation of gut when it is not at its usual place.

Case report

A 35 year old male patient presented to accidental emergency as a case of acute abdomen with history of pain abdomen for last 3 days and not passing flatus since 2 days. On examination abdomen was slightly distended and there was diffuse tenderness in right side of the abdomen. Patient was having increased heart rate around 112/min and blood pressure was 132/76 mm hg without any vasopressor support There was no other significant past history of any chronic illness. On investigating hemogram revealed leucocytosis and slightly raised SGOT and SGPT. Radiograph erect view of abdomen was unremarkable. Ultrasound of abdomen revealed free fluid in pelvis with no other significant finding. On the basis of clinical findings and radiological and biochemical investigation a exploratory laprotomy was planned. On exploring we found that omentum was going in the sub hepatic space and the appendix was grossly inflamed and caecum was in the right sub hepatic space (Figure 1 and 2) with no ascending colon and only transverse colon and descending

colon were present. We did an appendicectomy. Patient recovered well and was discharge uneventfully on post operative day 5. The histopathological report revealed an inflamed appendix.

Discussion

Mid-gut malrotation is a congenital anomaly in the embryological development of the fetal intestinal rotation. It has been estimated that it affects approximately 1 in 500 live births [1]. However, the true incidence is difficult to determine as a substantial number of cases will go undetected throughout life. The vast majority of the complications associated with mid-gut malrotation present in the first month of life and 60-85% of cases are diagnosed in this age group [1, 2]. It is reported that more than 90% of patients will present within 1 year of age [3]. Adult mid-gut malrotation is very rare and its incidence has been reported to be between 0.0001% and 0.19% [3,4]. A literature review by von Flue et al. cites 40 cases from 1923 to 1992 [3].

Most adult diagnoses of mid-gut malrotation are made in asymptomatic patients; either on imaging investigations for unrelated conditions or at operations for other pathology. This scenario of incidental diagnosis is becoming increasingly common, particularly with improvements, and increased use, of diagnostic imaging techniques in modern practice. However, there are a small proportion of affected adults who may present with acute or chronic symptoms of intestinal obstruction or intermittent and recurrent abdominal pain. The true diagnosis in this age group is fraught with immense difficulty, especially because the typical presentation is with non-specific symptoms and the fact that adult Surgeons usually have low index of suspicion and may not consider the diagnosis a possibility in the initial evaluation of adult patients with abdominal pain.

Mid-gut malrotation is broadly considered a deviation from the normal 270 degree counterclockwise rotation of the gut during embryonic development. During week 4 of fetal development, the embryonic gut, consisting of a straight endo-dermal tube, develops vascular pedicles to be divided into the foregut, midgut and hindgut based on the anatomical blood supply. The midgut is supplied by the superior mesenteric artery (SMA) and by the fifth week of embryonic life, it begins a process of rapid elongation and outgrows the capacity of the abdominal cavity. This leads to a temporary physiological herniation into the umbilical cord at about the sixth week of life with re-

turn to the abdominal cavity about 4 to 6 weeks later. During this period, the midgut undergoes a 270 degree counterclockwise rotation around the SMA axis. This process leads to the formation of the duodenal C-loop, placing it behind the SMA in a retroperitoneal position and emerging at the ligament of Treitz. The progressive reduction of the physiological midgut herniation commences at about week 10 of embryonic development.

The duodeno-jejunal flexure (DJF) and jejunum to reduce first and lie to the left. The distal small bowel then follows and lies progressively to the right of the abdominal cavity. The descent of the caecum from its higher position in the right upper quadrant forms the latter part of this complex rotational development; it becomes positioned in the right lower abdomen. The ascending colon then assumes a retroperitoneal position, also on the right side. The base of the small bowel mesentery subsequently fuses with the posterior peritoneum in a diagonal fashion, from the ligament of Treitz at the DJF to the caecum, completing the whole process at about the eleventh week of fetal development [1, 4-6].

The failure of the normal physiological rotation of the midgut leads to various degrees of anomaly including

1. Nonrotation: quite common; called "left-sided colon" and generally is asymptomatic, but twisting or volvulus can occur
    a. Midgut does not rotate after it enters the abdomen, the caudal limb enters before the cranial limb
    b. Small intestines lie on the right side and the entire large intestines on left. May cause obstruction of vessels and gut if kinking or twisting occurs
2. Volvulus and mixed rotation: cecum lies below the pylorus is fixed to the posterior abdominal wall by peritoneal bands that cross over the duodenum
    a. Usually causes duodenal obstruction
    b. Due to a failure of the midgut loop to complete final 90 degrees of rotation, thus, terminal ileum enters the abdominal cavity first
3. Reversed rotation: rare, clockwise rotation (not counterclockwise)
    a. Duodenum lies in front of the superior mesenteric artery and transverse colon behind it, which may ob-

struct the latter due to pressure from the artery

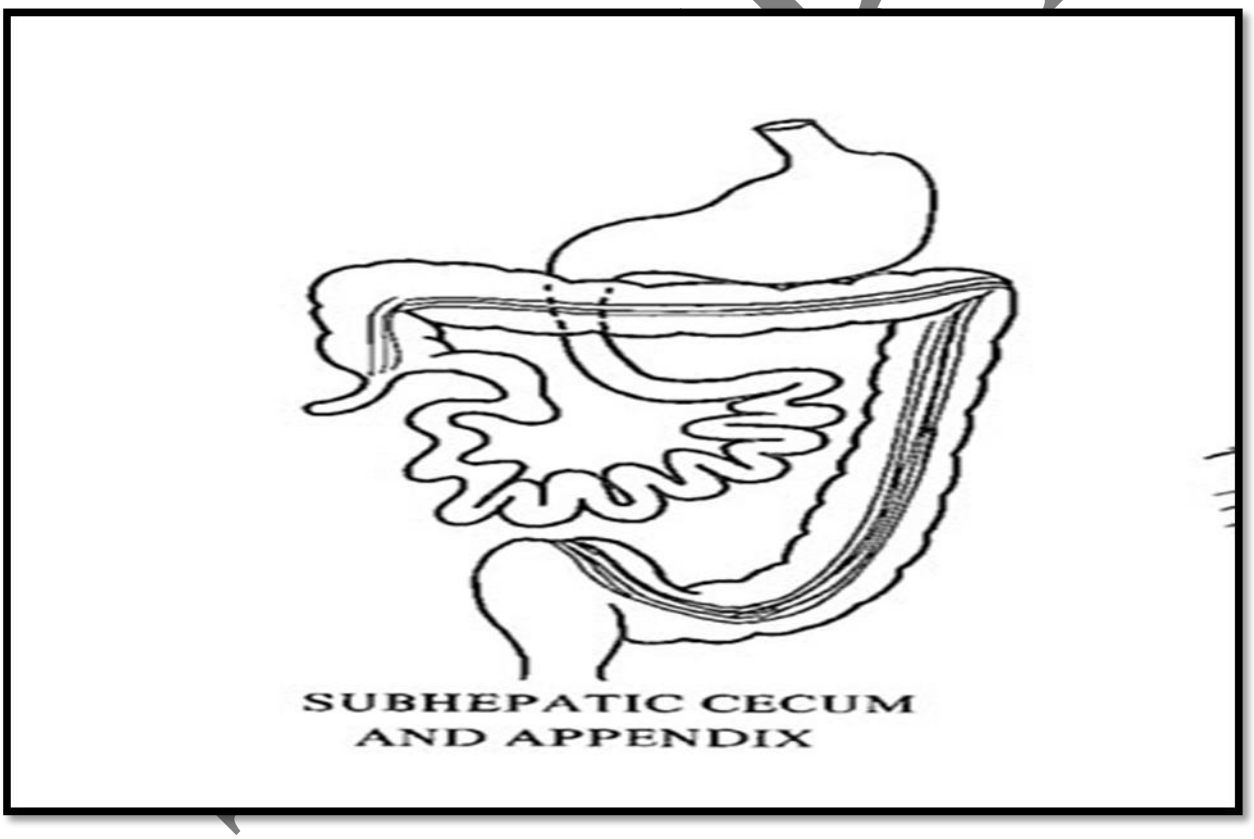
b. Small intestines lie on the left; large intestines lie on the right, and cecum is found in the center

4. Anomalies of position

1. Subhepatic cecum: failure of the proximal colon to elongate during stage 3 of rotation, thus, cecum ends up near the liver as found in our case

2. Mobile cecum: due to incomplete fixation of the ascending colon. Results also in a mobile and variable appendix and even volvulus of the cecum

3. Midgut volvulus: mesenteries fail to undergo normal fixation, and the intestines twist with incomplete rotation of the mid-gut loop. [7]



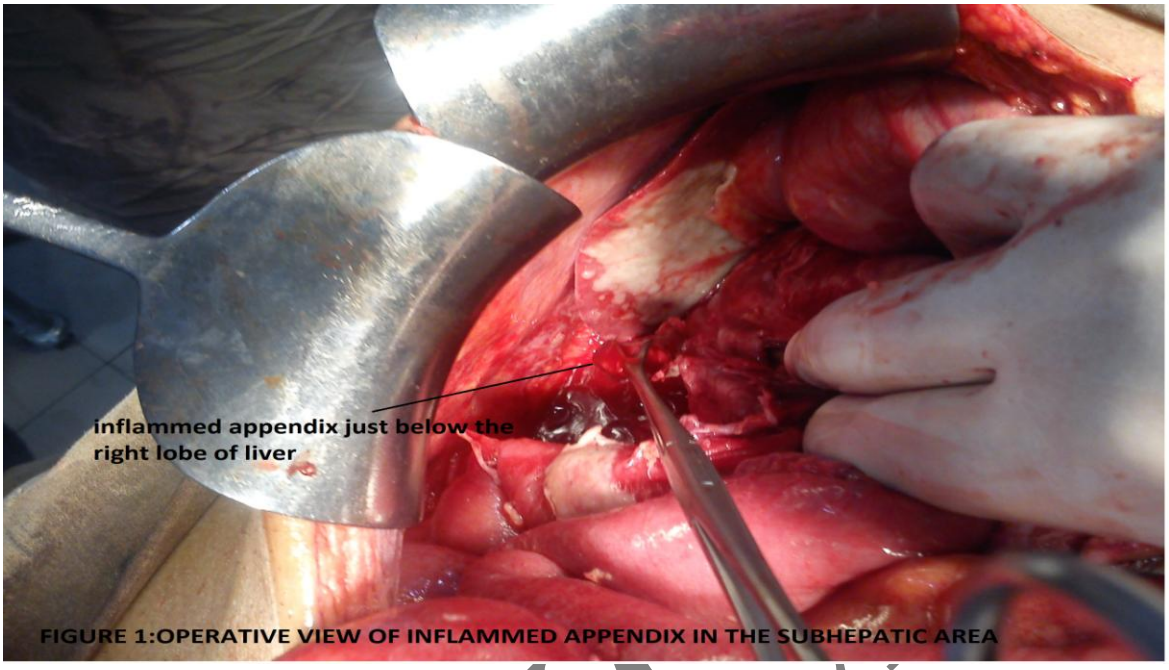


FIGURE 1:OPERATIVE VIEW OF INFLAMMED APPENDIX IN THE SUBHEPATIC AREA

FIGURE 2:ANOTHER OPERATIVE VIEW SHOWING THE APPENDIX IN SUBHEAPTIC AREA

Not all patients with malrotation present with symptoms. Indeed, most adult patients are asymptomatic and incidentally discovered later either at surgery for other conditions or at autopsy [9]. However, some may present with chronic symptoms of intermittent bowel obstruction or vague abdominal complaints. Even fewer may report acute episodes of agonizing abdominal pain [10]. Symptoms can arise from acute or chronic intestinal obstruction that may be caused by the presence of the Ladd bands and/or a volvulus. There is no typical set of symptoms that are ascribed to this clinical syndrome. The location of the pain may vary from epigastric to left upper abdominal quadrant and it may be described as either intermittent cramping or persistent aching pain. It most often occurs post-prandially and may last several minutes to an hour. Others have described severe abdominal cramping followed by diarrhea suggestive of chronic volvulus [11]. Vomiting may or may not be bilious and it is variable in duration and frequency. Another well-described presentation is a malabsorption pattern associated with diarrhea, nutritional deficiencies and failure to thrive [11, 12]. Some authors postulate that diarrhea and mal-absorption may be caused by bowel lymph edema resulting from lymphatic obstruction by chronic volvulus and resulting in loss of proteins into the bowel lumen [13]. Rare presentations of chronic volvulus include cases of obstructive jaundice by mechanical compression of the biliary tract [9, 13], chylous ascites and superior mesenteric vein thrombosis, secondary to long-standing lymphatic and venous obstruction [14]. Lymphatic hypertension and disruption have been postulated to occur secondary to torsion of the small bowel mesentery. Other reported symptoms include constipation, solid food intolerance and gastro-esophageal reflux [11, 12, 15, 16].

The diagnosis of malrotation in adulthood is often delayed, because of the wider and more obscure constellation of clinical symptoms observed in adult patients, who leads clinicians and patients to attribute symptoms to the wrong diagnosis. All too often, such patients undergo numerous investigational tests and carry diagnostic labels such as irritable bowel syndrome, peptic ulcer disease or psychogenic disorder [8,17].

Conclusion

Intestinal malrotation is a rare condition but is considered an important cause of bowel obstruction in adults. The diagnosis of

malrotation after childhood is difficult and usually not readily considered as the cause of intra-abdominal symptoms. The presentation is usually nonspecific and this often leads to diagnostic and treatment delay with possible bowel ischemia and necrosis. Acute appendicitis at times can be very challenging to operate in this case.

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