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Case Report

Midgut Malrotation: Subhepatic Caecum and Appendix

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ABSTRACT

The midgut portion of alimentary canal displays large variety of variations. Developmental variations of the caecum and vermiform appendix are relatively rare and scant attention is given to midgut malrotation as a cause of symptoms in adults. Subhepatic location of caecum and appendix was noticed during the dissection of a male cadaver about 50 -55 years of age. The diagnosis of acute appendicitis remains an enigmatic challenge and the subhepatic position of the caecum and appendix might add to further confusion and difficulty in diagnosing acute appendicitis at any age, including older adults. Also, failure to recognize the nature and characteristic features of these misplacements may lead to grave errors in the procedure, or injurious prolongation of the operation. Hence, knowledge of these variations may be of use to the radiologists and surgeons.

Keywords: Midgut, Mal rotation, Subhepatic caecum, Subhepatic appendix, Appendicitis,

INTRODUCTION

Large intestine extends from the ileocaecal valve to the anus. Caecum is the initial part of the large intestine that develops from the enlargement of the postarterial segment of the midgut loop. Usually, caecum lies on the peritoneal floor of the right iliac fossa and its lower end reaches the pelvic brim. [1] During the course of development of midgut loop, with the relative diminution in size of right lobe of liver, followed by sequential rotation and fixation, the caecum descends to occupy its adult position in the right iliac fossa. Its average length is 6 cm and breadth about 7.5 cm and it is covered by peritoneum on all sides. [2] Caecum receives the opening of ilium on its left wall and opening of the vermiform appendix on its posteromedial wall. The vermiform appendix is a diverticulum of caecum that shows variability in its position in relation to the caecum. The caecum continues as the ascending colon which passes upwards in the right lumbar region and right hypochondriac region to the inferior aspect of liver where it bends to the left forming hepatic flexure and continues as the transverse colon. Anatomical and topographical variations of the caecum are known to occur.

CASE REPORT

During regular dissection classes for first year medical students, variations were noted in the position of caecum in a male

cadaver aged approximately 50-55 years. The caecum and appendix were sub-hepatic in position. The right iliac fossa was empty. The length of the caecum was measured from a horizontal line at the level of ileocaecal orifice to its lowest point and width taken at midregion. It was quadrate in shape and measured 7.2 cm in length and 8.6 cm in width. The retrocaecal vermiform appendix was attached to its posterior wall and located in the subhepatic region in the hepatorenal pouch (Figures 1, 2, 3). The ascending colon was absent.

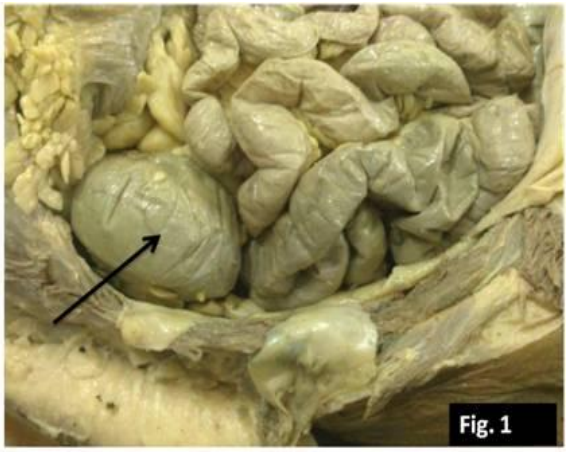


Fig. 1 Showing Subhepatic Caecum

Fig. 2 Showing Retrocaecal Subhepatic Appendix

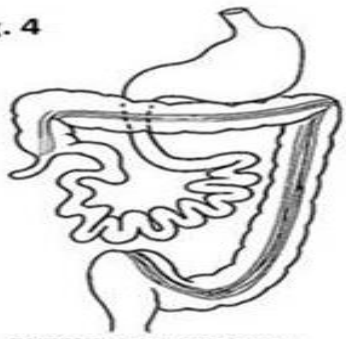
Fig. 3 Showing empty Right Iliac Fossa

DISCUSSION

Malrotation is a consequence of alteration in embryonic development of the midgut. Reported incidence of intestinal rotational disorders is as high as 1% of the total population. [3] Vermiform appendix and caecum develop from the caecal bud of postarterial segment of midgut loop. In early stages of development they are of same diameter but later the appendix narrows down due to the faster growth of the proximal part of the caecal bud. Three general types of caeca according to shapes (a) the infantile; (b) that prevailing in early childhood; and (c) the adult, may be regarded as a developmental sequence. Development of midgut is characterized by rapid elongation of the gut and its mesentery, resulting in formation of the Primary intestinal loop. The primary intestinal loop undergoes rapid elongation and due to the temporary inability of the abdominal cavity to accommodate it, the loop enters the extraembryonic cavity in the umbilical cord during the sixth week of development (physiological umbilical herniation). The increase in length of the loop is accompanied by simultaneous rotation around the axis formed by superior mesenteric artery. During the 10th week, herniated intestinal loops begin to return to

the abdominal cavity. Although the factors responsible for this return are not precisely known, it is thought that regression of the mesonephric kidney, reduced growth of the liver, and expansion of the abdominal cavity play important roles. [4] When the intestine returns to the abdomen, the caecum and appendix occupy the subhepatic position. Later, in the eleventh week, they descend to the right iliac fossa. The development of midgut undergoes rotation and fixation of parts to assume adult position. The caecal region undergoes changes in the form of descent or caudal migration of caecum with the relative diminution of size of right lobe of liver so that ascending colon, hepatic flexure are defined and the caecum reaches the right iliac fossa. Derangements may occur at any stage, either rotation or fixation. The factors of intrinsic growth and gravity (weight-bearing of caecal contents) are considered in the evolution of the caecum from infantile to adult types. [5] Common varieties of anomalous location of caecum include pelvic caecum, lumbar caecum, or sub hepatic caecum (Fig. 4). [6]

Fig. 4



SUBHEPATIC CECUM AND APPENDIX

In the present case, caecum and appendix were found in the subhepatic region. Literature related to prevalence of sub–hepatic caecum is comparatively scarce. Surgeons have described occasional cases discovered at operation. The sub hepatic caecum has been reported in 6% of population by certain authors. [7] In a cadaveric dissection study on 25 cadavers, caecum and appendix were found in subhepatic position in 4% cases. [8] Palanivelu et al (2007) reported subhepatic appendix with frequency of 0.09%. [9]

Caecum is present in most amniote species, and also in lungfish, but not in any living species of amphibian. In reptiles, it is usually a single median structure, arising from the dorsal side of the large intestine. In humans the caecum and appendix usually lie in the right iliac fossa. Most mammalian herbivores have a relatively large caecum, hosting a large number of bacteria, which aid in the enzymatic breakdown of plant materials such as cellulose; in many species, it is considerably wider than the colon. In contrast, obligatory carnivores, whose diets contain little or no plant material, have a reduced caecum, which is often partially or wholly replaced by the appendix. [10] Mammalian species which do not develop a caecum include raccoons, bears, and the red panda. Over 99% of the bacteria in the gut flora are anaerobes, but in the caecum, aerobic bacteria reach high densities. [11] Variations in anatomical location of caecum and appendix can be either causative or may influence patterns of clinical conditions and is therefore of clinical and surgical importance. Sub–hepatic appendicitis was first described in 1955 by King and since then has been reported infrequently. [12] Our case report is in line with that of other researchers who reported retrocaecal appendix with subhepatic caecum. Subhepatic appendicitis is a rarely reported variant of a common surgical emergency that leads to delayed diagnosis and incurs higher complication rates, including suppuration and perforation. Inflammation of a subhepatic appendix can mimic cholecystitis and perforation of a subhepatic appendix can mimic liver abscess. [13] These developmental anomalies of gut may be kept in mind as an unusual cause of recurrent

abdominal pain in otherwise healthy adults. The subhepatic caecum and appendix situated in the hepatorenal pouch lie closely related to the liver, right kidney and right adrenal. Infections from the appendix if any might spread to the right kidney, suprarenal region, and liver, and could lead to a diagnostic dilemma. [14] Also, a prior knowledge of such a variant position of caecum and appendix may help in diagnosing cases of appendicitis with atypical presentations and in planning proper incisional techniques preoperatively.

CONCLUSIONS

Subhepatic caecum and appendix is a rare anomaly, wherein, the intimate relationship of the right lobe of liver, right kidney and right adrenal to the caecum with its rich bacterial flora may have its own clinical implications. Such an unusual location of these organs adds confusion in diagnosis and further management. With a high incidence of appendicitis in the general population along with the accompanying diagnostic dilemma, knowledge of such rarely reported unusual locations of the appendix may be helpful in preventing diagnostic delays and guide the surgeon for planning appropriate surgery.

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