


Intracecal appendix: an extremely rare anatomical variation. A case report and review of literature

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Abstract Appendicitis is the most common surgical abdominal emergency in the developed world. Most of the surprises encountered during an appendectomy are usually due to the various positions of the appendix tip. Anatomical variations are an extremely rare phenomenon, with only a few case reports scattered among volumes of literature on the vermiform appendix. A new variation is described in which the appendix was intracecal and cause for surprise during surgery. A review of literature of rare anatomical variations of the appendix is also presented.

Keywords Intracecal appendix · Intramural appendix · Anatomical variations of appendix

Introduction

The vermiform appendix, although vestigial, is a significant source of morbidity, and occasionally mortality, when inflamed. In spite of being one of the most common emergency surgical procedures performed by the surgeon, appendectomy is fraught with pitfalls for the unwary. Beginning from the first drawings of the appendix by Leonardo da Vinci in 1492, the study of the anatomy and embryology of the appendix had captured the attention of the surgical fraternity right up to the mid-twentieth century. The interest, thereafter, has somewhat waned, and, with the advent of laparoscopy and cross-sectional imaging, publications related to the anatomy per se have all but dried up. The appendix has a remarkably constant anatomy,

classically being located at the base of the cecum at the junction of the three taenia with the tip in the retrocecal position. Almost all the variations seen in surgery are those of the positions of the tip. Other variations like absence, ectopia, and duplication are extremely rare.

A new variation is described in which an intracecal appendix was detected during open appendectomy for acute appendicitis. To the best of our knowledge, this is only the third report of such a case after 1972 [7] and 1983 [1].

Case report

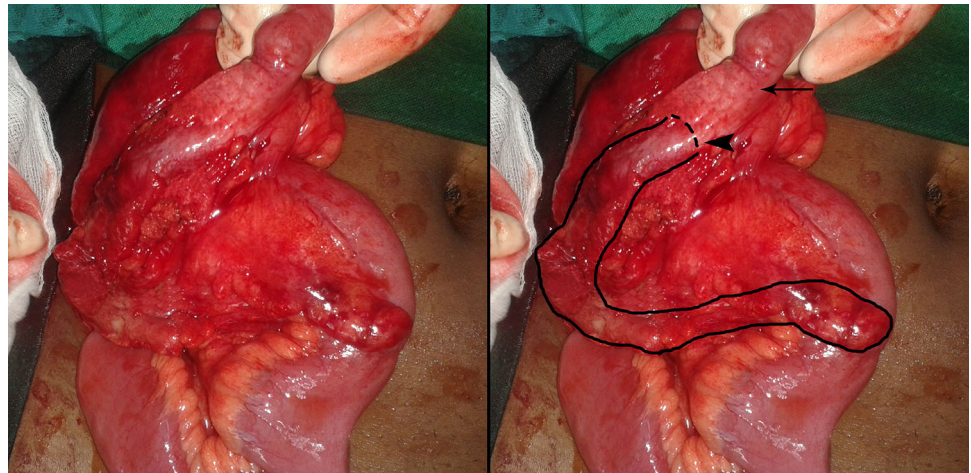
A 28-year-old male presented with the classical history of acute appendicitis in the form of pain in the right iliac fossa for 1 day, anorexia, and one episode of vomiting. On examination, the right iliac fossa was tender with the point of maximum tenderness being slightly above the McBurney's point. Routine investigations revealed only a mildly elevated leukocyte count of 12,500/mm³. Ultrasonography of the abdomen revealed a blind-ending, aperistaltic bowel loop in the right iliac fossa with associated inflammation of the cecal base. Due to a strong clinical suspicion, decision was taken to perform an emergency appendectomy.

The abdomen was entered via a muscle splitting Lanz incision. The anterior wall of the cecum was delivered into the wound, but the appendix was not visualised. On palpation of the region, the appendix was felt in a retrocecal position. Suspecting adhesions of the tip, the entire cecum, terminal ileum, and part of the ascending colon were delivered into the wound. To our surprise, the appendix was still not visible, but could be felt along the posterior cecal wall, extending superiorly and medially where the only tip was visible on the mesentery of the terminal ileum

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Fig. 1 Appendix embedded within the wall of the cecum. *Left* image as seen on the operating table. *Right* outline of appendix (*black line*), base of appendix (*arrow head*), and cecum (*solid arrow*)



(Fig. 1). The landmarks were again confirmed and a diagnosis of intracecal or intramural appendix was made. The appendix was dissected in a retrograde manner, using right-angled clamps to dissect the serosa and part of the muscularis of the cecum covering the appendix and ligating all vessels encountered. The resultant defect in the serosa and muscular layer of the posterior cecal wall measured about 12×3 cm (Fig. 2). The appendix was removed after transfixing the base and the defect in the cecal wall was repaired with a continuous layer seromuscular sutures. The appendix measured about 16 cm in length and was about 15 mm in external diameter. The post-operative period was uneventful and histopathology was consistent with acute appendicitis.



Fig. 2 Defect in the cecum after dissection of appendix

Discussion

Acute appendicitis is the most common surgical abdominal emergency. Surprisingly, the incidence is ten times lower in developing countries as compared to the developed countries. Males are more at risk than females with a sex ratio of 1.4:1 [5]. Most of the variations encountered during an appendectomy are due to the position of the tip which can depend upon a number of factors: the degree of cecal descent and peritoneal fixation, the configuration of the cecum, appendiceal length, associated adhesions, and the habitus of the person [3]. In spite of being the most commonly excised abdominal organ as an emergency, true surprises on the operating table owing to anatomical variations are extremely rare.

Embryologically, the appendix is the terminal portion of the embryonic cecum and becomes distinguishable due to its slower growth compared to the proximal cecum. This differential growth persists into the early childhood. The appendix is usually visible at about the eighth week of gestation. It initially projects from the apex of the cecum and with the growth of the cecum, which predominantly occurs in the lateral wall; the appendiceal origin shifts medially toward the ileocecal valve. The taeniae of the longitudinal muscle coat of the colon originate from the base of the appendix, showing the same displacement. The position of the tip of the appendix varies owing to the mesentery.

The absence of the appendix is extremely rare with a reported incidence of about 1/100,000 laparotomies for acute appendicitis [9]. The earliest reports of absence include those of Morgagni [8] and Hunter [6]. This condition is more commonly diagnosed in adults than children. There are many theories of causation like intra-uterine vascular accident, autoamputation of the appendix, and the rarest, appendiceal atresia.

The ectopic appendix is even rarer with only a few cases being reported. Fawcitt [4] reported an appendix in the thorax associated with malrotation and a diaphragmatic hernia. Babcock [2] removed an appendix from the lumbar area, probably due to an associated hernia. McGladdery [7] and Abramson [1] have reported an intramural or intracecal appendix. To the best of our knowledge, our case is the third report of this variation which occurs when the appendix fuses with part of the developing cecal wall and can be covered with any of the layers of the cecum. This fusion most likely occurs just prior to the fixation of the cecum in the right lower quadrant of the abdomen after rotation of the midgut. This is the time that the appendix is forced into a retrocecal position and is most closely applied to the posteromedial cecal wall. The subsequent growth of the cecum occurs, with the closely abutted appendix getting incorporated into the cecal wall. Thus, the most common position of the appendix in this variation is retrocecal. The hallmark of this variation is the apparent absence of the mesoappendix which is likely the first structure which gets incorporated into the cecal wall. Commonly, only the serosa of the cecum envelops the appendix [1], but rarely, the appendix can lie within the muscular layer, as was seen in our case. There have been unsubstantiated and only anecdotal reports of the elusive ‘submucosal’ appendix. Abramson [1] goes so far as to say that the agenesis of the appendix is actually a missed intracecal appendix.

One point of contention that needs to be addressed here is why we chose to call this an intracecal/intramural appendix and not just a retrocecal appendix adherent to the posterior cecal wall. We, ourselves, were hesitant to call this an intracecal appendix because of its apparent rarity, but were forced to consider it because of the following reasons:

- (a) In a retrocecal appendix, the base of the appendix is generally free and distinguishable from the cecum.
- (b) The adhesions between the appendix and the cecum generally occur to the inflamed part. In this case, the tip was the only inflamed part, seen clearly in Figs. 1 and 2. However, this was the only part of the appendix which was free.
- (c) There was no mesoappendix distinguishable separately.
- (d) The vascular supply seemed to come from the posterior cecal wall, and on separating the appendix, a large arterial spurter was encountered on the cecal wall which had to be controlled using a figure-of-8 stitch.
- (e) After separating the appendix, circular muscle fibres of the cecum could be seen, and in the proximal portion near the base, even the circular muscle was cut and submucosa was visible.

Appendiceal duplication is slightly more common and has generated enough reports in the literature to warrant

attempts on a classification system. The classification system in vogue is that of Wallbridge [11] who built upon the original system of cave. Rarer entities include congenital appendiceal diverticula and heterotopic mucosa which may include pancreatic, gastric, or esophageal types.

Cross-sectional imaging (CT scan) is fast becoming established as the gold standard in the diagnosis of acute appendicitis. Thin slice CT scan may yield information on certain variations of appendix, but it is very difficult to identify positional variations. In fact, most of the positional “variations” in the appendix reported on CT scan are due to abnormal positions of the cecum [10]. The true value of CT scan in detecting an intracecal appendix is unknown as there is no report of it to the best of our knowledge.

A thorough knowledge of anatomy and even the rarest of variations is an invaluable tool to the surgeon. Keeping these variations in mind may help the flummoxed surgeon when he is faced with an “absent” appendix when operating for acute appendicitis. Furthermore, in today’s era of laparoscopic surgery, the lack of tactile feedback makes it all the more necessary to be aware of this entity, whose only clue may be a turgid, tubular structure palpable in the posteromedial wall of the cecum.

Conclusion

Anatomical variations of the appendix are a rare occurrence and probably chance upon a surgeon only once or rarely twice in his career. This is evident by the sparse literature available on this topic. Therefore, we feel that this is an apt case to report as a thorough search of literature from 1930 onwards revealed only two reports thus far. In addition to expanding existing literature, it will also reiterate to surgeons to be mindful of the anatomical variations of the appendix while operating and be more adept at handling them.

Author contributions SC: conception and data acquisition; critical revision of article; final approval; accountability for published work. SA: data acquisition and analysis; drafting of article; final approval; accountability for published work.

Compliance with ethical standards

Conflict of interest The authors declare that they have no competing interests.

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