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CASE REPORT

CONGENITAL ABSENCE OF THE VERMIFORM APPENDIX

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Abbreviations: β-HCG, β-human chorionic gonadotropin.

INTRODUCTION

Congenital absence of the vermiform appendix in humans is rare. Only a handful of reports on this topic are to be found in published work, which suggests that several criteria need to be met before a surgeon can confidently state that the appendix is indeed absent. We report a new case of congenital absence of the appendix in a young woman and discuss the criteria that must be met before this diagnosis can be made.

CASE REPORT

A 25-year-old woman presented to the Emergency Department at Royal Prince Alfred Hospital, Sydney, complaining of symptoms consistent with acute appendicitis. She described 48 h of vague central abdominal pain that she had attempted to ignore. However, she was woken from sleep in the early hours of the morning on the day of presentation with more severe abdominal pain that had become localized in the right iliac fossa. She had had no vomiting, no alteration of bowel habit, and no symptoms that could be attributed to the genitourinary system.

Her only past medical history was a recent injury to the skin of the left upper quadrant of her anterior abdominal wall as a result of accidental spillage of some boiling water. She also had been diagnosed with hyperprolactinaemia. She had not undergone any previous surgery.

On examination she was in obvious discomfort, with tenderness, guarding, percussion tenderness and rebound tenderness in the right iliac fossa. There was erythema from her recent burn, but no scar indicating any previous abdominal surgery. A gynaecological examination was normal as was urinalysis. She was afebrile and her white cell count was 9.3 g/dL (normal range, 3.5–11.0 g/dL). Serum β-human chorionic gonadotropin (β-HCG) was 4 lU/L (normal range, 0–5 IU/L).

A provisional diagnosis of acute appendicitis was made, and the patient was taken to the operating theatre. A standard approach was taken with incision over McBurney's point. The caecum was identified and the taeniae coli were followed to their confluence. When this revealed no appendix, the caecum and ascending colon were more thoroughly mobilized, but still the appendix could not be identified (Fig. 1). The entire ileocaecal region was then palpated to exclude any thickening of the bowel wall or intraluminal mass that could represent an appendiceal intussusception. Despite all these manoeuvres it was not possible to locate the appendix. There was no Meckel's diverticulum, but there was a right-sided ovarian cyst, which was presumably responsible for the patient's symptoms.

The abdomen was closed and the patient made an uneventful recovery.

DISCUSSION

Morgagni described congenital absence of the appendix in 1718.[1] Since that time other cases have been reported, but the incidence of absence of the appendix remains very low. It is easy to attribute apparent absence of the appendix to a failure to find it, but there are a few authenticated cases of complete absence of the appendix. The majority of these were operative diagnoses, but some have been incidental findings at post mortem examination.

Less than 100 cases of agenesis of the appendix have been reported since Morgagni's first description. Host *et al.* state that the congenital absence of the appendix is found in 0.006% of bodies undergoing autopsy, and 0.25% of bodies undergoing anatomical prosection.[2] Chevre *et al.* observed that the incidence of vermiform appendix agenesis in living people is estimated at 1 in 100 000 laparotomies for appendicitis.[3]

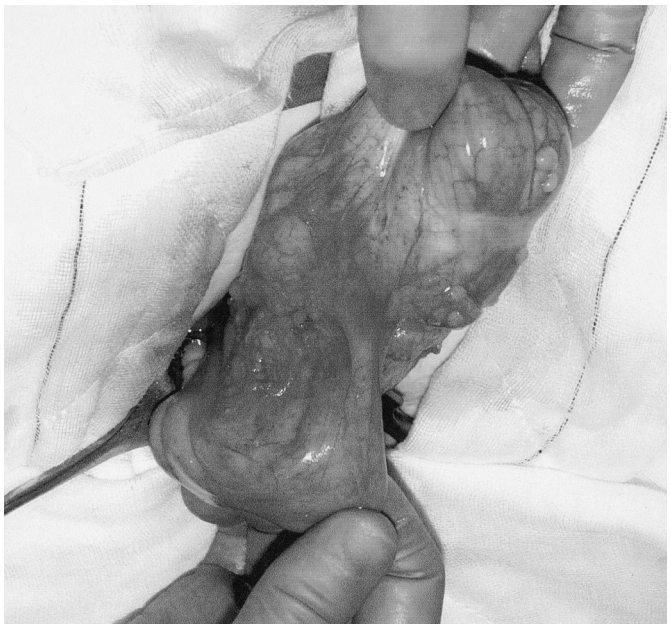

Fig. 1. Mobilized caecum demonstrating absent appendix.

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In two children, agenesis of the appendix has also been attributed to a secondary effect of the maternal ingestion of thalidomide during pregnancy.[4,5]

The appendix is the apex of the embryonic caecum, arising from its posteromedial aspect, approximately 1.5–2.0 cm below the ileocaecal junction. On average it is 9 cm long and 6 mm in diameter, although wide variations occur.[6] The appendix arises from the caecum during the eighth week of foetal development, shifting from its original lateral position on the caecum to a more posteromedial position.[3] The arrest of this movement can occur at any point and it is this that is responsible for the wide variety of positions that the appendix can occupy. The various positions that the appendix can occupy are well described elsewhere.[6] It is worth noting that a very mobile caecum or malrotation of the gut can be responsible for the appendix being abnormally located within the abdomen. Cases have been reported where the appendix has been found in the thoracic cavity as a result of malrotation and diaphragmatic herniation, as well as in the lumbar area.[7,8] Intussusception of the appendix and intramural appendicies have been described, both possibly being confused with a congenitally absent appendix.[9]

When difficulty is encountered in locating the appendix at surgery, the taeniae coli should be followed to their confluence. If this reveals no appendix, the caecum and ascending colon should be mobilized and a thorough search carried out. The entire ileocaecal region should then be palpated to exclude any intraluminal or intramural masses indicative of either appendiceal intussusception or an intramural appendix. A diagnosis of congenital absence of the appendix can be made once it can be established that there has been no previous abdominal surgery (including laparoscopy). If the diagnosis is seriously suspected, it is important to reaffirm this history postoperatively. All old hospital notes should be closely scrutinized and parental information should be sought on paediatric surgical procedures (if any).

Collins described a classification system for classifying abnormalities of the caecum and appendix.[10] Class I is for cases in which the caecum and appendix are both absent. Class II comprise cases where there is a rudimentary caecum with no appendix. In Class III there is a normal caecum and no appendix. In Class IV there is a normal caecum with a rudimentary appendix, and in Class V there is a giant caecum with no appendix. The case in this report would belong to Collins Class III, which is also the most common class.

CONCLUSION

This report describes a rare instance of a young woman with congenital absence of the appendix. The steps a surgeon should undertake when experiencing difficulty locating the appendix are outlined, as are the criteria that must be met before a diagnosis of congenital absence of the appendix can be made.

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