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Horseshoe appendix: a extremely rare anomaly

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Rezumat

Apendice în potcoavă: anomalie extrem de rară

Anomaliile apendicelui vermiform sunt malformații extrem de rare. Prezentăm cazul unui pacient de 43 de ani care a fost operat în urgență de ocluzie intestinală. Am descoperit intraoperator un apendice "în potcoavă" pe care l-am operat. După trecerea în revistă a literaturii de specialitate am încadrat această variantă morfologică de apendice într-o clasificare completă a anomaliilor apendiculare.

Cuvinte cheie: anomalii apendiculare, apendice în potcoavă

Abstract

Appendiceal anomalies are extremely rare malformations. We presented the case of a patient 43-year-old who had undergone emergency surgery for bowel occlusion. Incidentally we have found an "horseshoe-shaped" appendix which removed. After review of the literature we have introduced this appendiceal variant in a complete classification of appendiceal anomalies.

Key words: appendiceal anomalies, horseshoe appendix

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Introduction

Appendiceal anomalies are extremely rare malformations. There have been some cases of true absence of the vermiform appendix reported, and average 100 cases of duplication have been reported to date. Malformations of appendix may be associated with other visceral anomalies.We review the surgical literature and we found only two cases of a "horseshoe"- shaped appendix. Most encountred anomalies was appendiceal duplication.

Case report

We presented the case of a patient 43-year-old who went through surgery 11 months earlier, for a generalized peritonitis, caused by a perforated ulcer on the anterior wall of the superior part of duodenum, just near the pylorus. At this second admittance, through the emergency room, the symptoms were abdominal cramps, constipation, vomiting, which all leaded to diagnose of an intestinal occlusion.

At the local examination we found a supraumbilical scar, a medium distension of the abdomen, with visible bowel movements and by combining the percussion and the auscultation we notified the presence of hydroaeric sounds. At the rectal exploration, the rectal ampoule was empty. The abdominal radiography showed hydroaeric levels in the small intestine.

After the general anesthesia was induced, we made a median incision, both of supra- and subumbilical level. We found multiple adhesions between bowel coils and between the abdominal wall and the intestinal wall, which were removed by adhesiolysis. Of note was a cecum moderately distended and in a proper location but with the finding of a horseshoe appendix with two bases, each communicating with the cecum. At this time we found a horseshoe appendix, of 13/1 cm, almost

sagittally disposed, with a central mesoappendix, which has a vascular trunk with some nutritive arterial branches. By palpation we discovered few coprolites, the biggest of them blocking one of the distal part of appendicular horseshoe. We proceeded to the ligature and section of mesoappendix, (Fig. 1 A,B,C) and of the two caecal insertion areas, which one of 4-5 mm. diameter. After that, we controlled the hemostasis and the peritoneal viscera. Anatomical closure of abdominal wall. The evolution was normal after surgery.

There were no associated other congenital anomalies. Microscopic sections were typical for those of an appendix, with a branching mucosa, submucosa, bilayered muscular wall and serosa, moderate inflammatory infiltrate in submucosa with many lymphoid folicles of the appendicular ends (in the area were stercholite was found).

Discussion

Anomalies of the appendix are extremely rare. There have been reported both absence and duplication of the appendix. Absence of the appendix have been described with a normal cecum, whithout other visceral anomalies. Collins reported four cases (0,0008%) of congenital agenesis in a study of 50 000 specimens of the human vermiform appendix (1). The causes of this anomaly is unclear because of its rarity and lack of consensus of opinions about this. Phylogenetically, in several species, *Felis* (tiger, lion, leopard) and *Canis* (dog, fox, wolf) the appendix are lacking (2). More frequently is reported the duplication of appendix with an incidence of 0,004% and may be associated with other anomalies (3). These anomalies are thought to result from the persistence of a normally transient embryologic second cecal appendix. Cave and Wallbridge (4) have classified the duplication of appendix into three types.

The classification system for appendiceal duplications was first developed by Cave in 1936, then modified by Wallbridge in 1963, and finally by Biermann in 1993 (rep. by 5) This system classifies three types of appendiceal anomalies: A, B, and C.

- A: partial duplication of the appendix on a single cecum *(Fig. 2)*;
- B: two completely separate appendices on a single cecum with two subtypes:
  - B_1: "bird-like appendix" or "avian type": two appendices symmetrically placed on either side of the ileo-cecal valve. This type is found normally in birds. In humans it is found associated with intestinal and/or genitourinary anomalies *(Fig. 3)*.
  - B_2: "taenia-coli type": one appendix arises from the usual site on the cecum with another rudimentary arising from the cecum along the tenia of the cecum. This type and the followings have no other associated anomalies *(Fig. 4)*.
  - B_3: the second appendix is located along the tenia of the hepatic flexure of the colon.
  - B_4: the second appendix is located along the tenia of the splenic flexure.

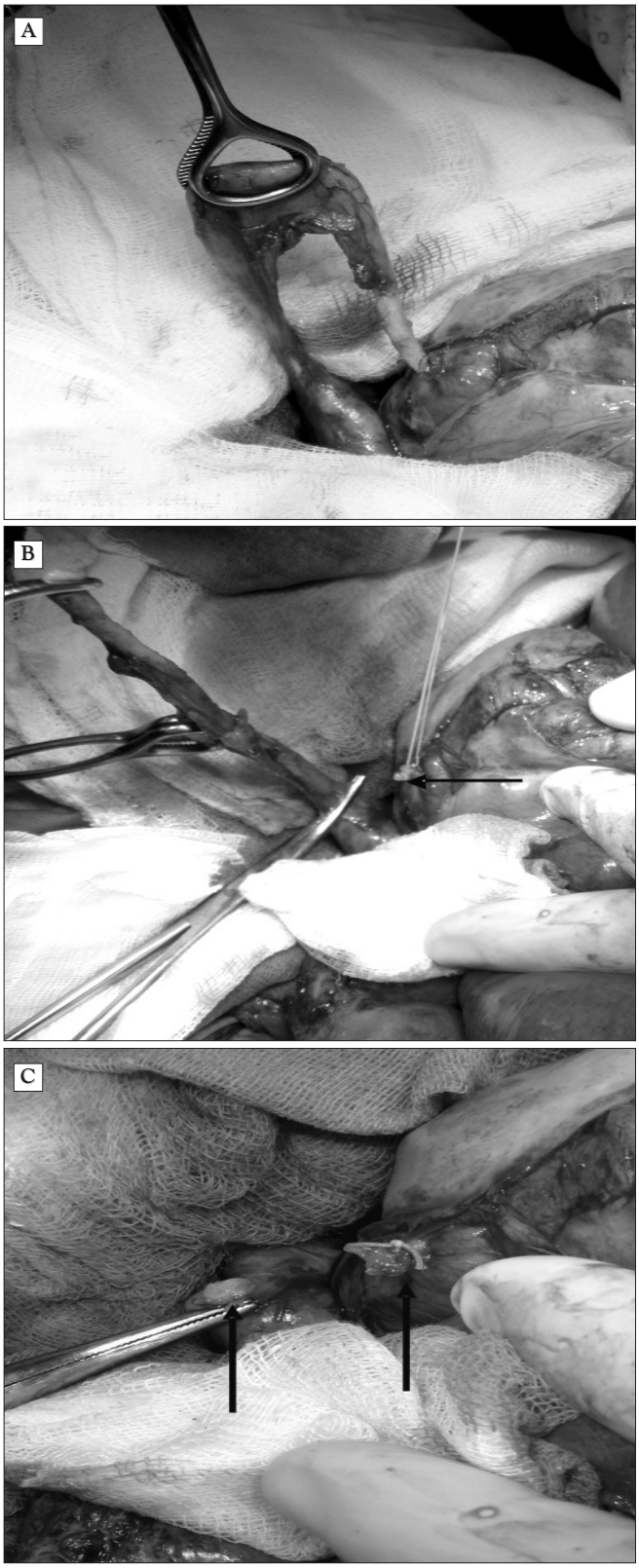


Figure 1. ***(A)*** *Horseshoe-shaped appendix with sagital disposal - intraoperative aspects.* ***(B)*** *Horseshoe-shaped appendix with sagital disposal - intraoperative aspects - distal caecal implantation was ligated.* ***(C)*** *Horseshoe-shaped appendix with sagital disposal - intraoperative aspects - both caecal implantations are ligated*



Figure 2. *Appendiceal duplication (partially) in "Y-shaped"*



Figure 3. *Duplex appendix - "Avian" type*

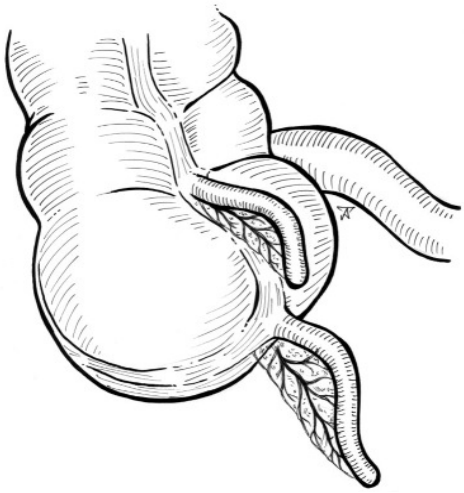


Figure 4. *Duplex appendix - "Taenia-coli cecum" type*

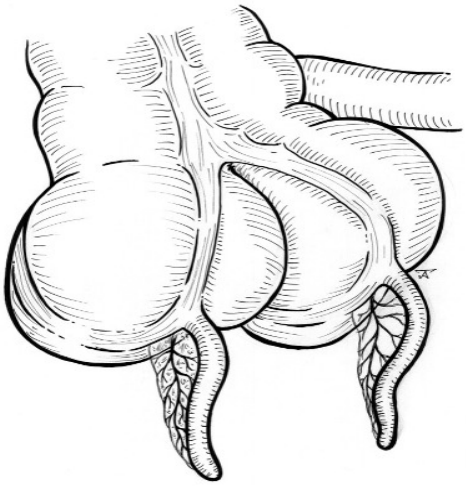


Figure 5. *Duplex appendix on caecal duplication*

- C: two caeca, each bear an appendix. This type occurs in association with hindgut maldevelopment (ileum, colon, anus) and other anomalies of genitourinary tract and lower vertebral column (2) (*Fig. 5*).
  Tinckler (6) and more recently Uriev L et al (7) have described a triplication of appendix, Tinckler in a child with exstrophy of the bladder and Uriev in an adult patient whithout other anomalies. We can it consider another type in previously classification.
- D: three completely separate appendices with or whithout other anomalies.

When the anomalies of vermiform appendix are detected in childhood they are nearly always associated with severe intestinal, genitourinary or bony malformations, seen most often in conjunction with B1 and C duplications (6).

Brown have reported the first description on a barium enema of type C duplicated appendix (8) and Peddu a type B_1 duplicated appendix (9).

Our patient is the third reported case of a horseshoe-shaped appendix after Mesko (2), and DasGupta (10). The appendix communicated with the cecum at both ends in sagital direction along the *taenia coli* (sagital disposal), and was supplied by a single fan-shaped mesentery. The appendix was continuous and its lumen patent with some moving coprolites. This appendiceal anomaly must be the result of some unknown embryologic event. Perhaps during embryologic life the single appendiceal base somehow split in two and became further separated during cecal growth. This might account for just such a double-based, yet single, structure (2). In the case of Mesko et al, the bases were located fairly near on another, neither arising clearly on a tenia (frontal disposal).

The appendiceal anomalies, although rares, should be known to the general surgeon.

In a more completely classification the anomalies of the appendix were:

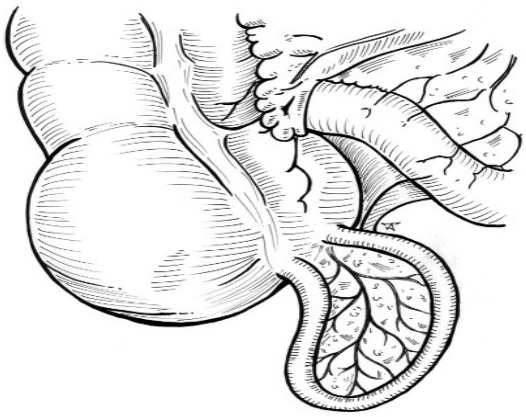


Figure 6. *Horseshoe-shaped appendix with frontal disposal*

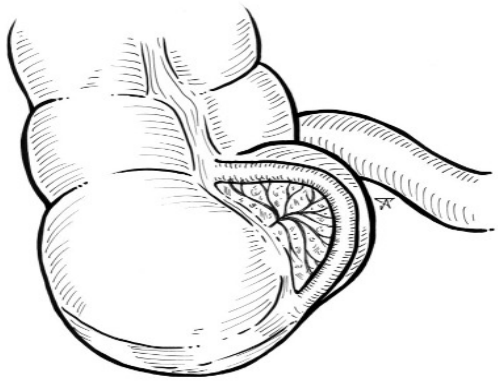


Figure 7. *Horseshoe-shaped appendix with sagital disposal*

Number anomalies

1. Congenital agenesis
2. Multiforious appendices:
   A: appendiceal duplication(partially) in "Y-shaped" *(Fig. 2)*;
   B: duplex appendix on a single cecum:
   - B_1 - "avian type" with intestinal and/or genito-urinary anomalies *(Fig. 3)*;
   - B_2 - "taenia-coli cecum type" *(Fig. 4)*;
   - B_3 - "taenia-coli hepatic flexure type";
   - B_4 - "taenia-coli splenic flexure type".

   C: duplex appendix on a two caeca (each bear an appendix) with hindgut, genitourinary tract, lower vertebral column maldevelopment *(Fig. 5)*.
   D: triplex appendix:
   - "new-born type" ± other congenital anomalies;
   - "adult type"whithout other congenital anomalies.

Shape anomalies:

Horseshoe-shaped appendix:
- with frontal disposal *(Fig. 6)*;
- with sagital disposal *(Fig. 7)*.

These may be found during abdominal surgery for an unrelated problem or during an operation for appendicitis itself. When the appendiceal anomalies (number and shape anomalies) are discovered during surgery, they should be removed if possible. Duplex appendix "avian type" and with two caeca are found in children when they are serious anomalies of the alimentary and genitourinary tracts. In these cases the operation performed is imposed by the most serious anomaly present (2).

Anomalies of the appendix may have serious medico-legal significance. Maizels (11) points out that this is particularly true for the B_2 type of duplication. He cited a legal case of a child who had appendectomies performed twice during a 5-month period.

In conclusion, although rare, anomalies of the appendix may occur. In this paper we report a third case of horseshoe appendix mentioned in surgical literature. In our case, sagital disposal along the *taenia coli* is peculiary. This patient has no other associated anomalies.

References

1. Collins DC: A study of 50 000 specimens of the human vermiform appendix. Surg Gynecol Obstet. 1955;101(4):437-45.
2. Mesko TW, Lugo R, Breitholtz T: Horseshoe anomaly of the appendix: a previosly undescribed entity. Surgery. 1989; 106(3):563-6.
3. Chew DK, Borromeo JM, Gabriel YA, Holgersen LO. Duplication of the vermiform appendix. J Pediatric Surg 2000; 35:617-618.
4. Wallbridge PH. Double appendix. Br J Surg. 1962;50:346-347
5. Bender Elizabeth, Schmidt S. Type B2 appendiceal duplication with appendicitis: a case report. Internet J Surg. 2002;3:2.
6. Tinckler LF. Triple appendix vermiformis: a unique case. Br J Surg. 1968;55:79-8.
7. Uriev L, Maslovsky I, Monouskin Y, Ben-Dor D. Triple barreled type of appendiceal triplication. Ann Diagn Pathol. 2000;10(3): 160-161.
8. Brown RB. Duplication of the cecum and appendix. Radiology. 1957;69:266-267.
9. Peddu P, Sidhu PS: Case report: Appearance of a Type B duplex appendix on a barium enema. The British Journal of Radiology. 2004;77:248-249.
10. DasGupta R, Reber PU, Patel AG: Horseshoe appendicitis. Case report. Eur J Surg. 1999;165(11):1095-6.
11. Maizels G: Duplication of the vermiform appendix. S Afr Med J. 196624;40(46):1123-5.