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Case Report

Surgery Section

Horseshoe Appendix: An Extremely Rare Appendiceal Anomaly

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ABSTRACT

Appendiceal anomalies are extremely rare malformations that are usually found incidentally. Agenesis and duplication of the appendix has been well documented however, the cases of horseshoe appendix reported is very limited, only four cases reported so far. Here, we report a four and half-year-old who underwent interval appendectomy. Intraoperatively both the ends of the appendix were found to be communicating with the cecum with two separate base or stump located at a sagital disposal- the so called “horseshoe appendix”.

Keywords: Appendectomy, Duplex appendix, Transient appendix

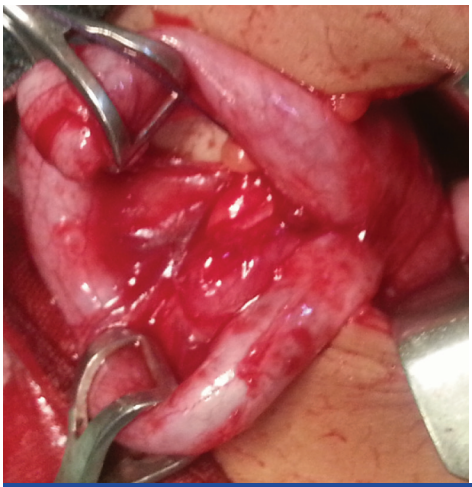
CASE REPORT

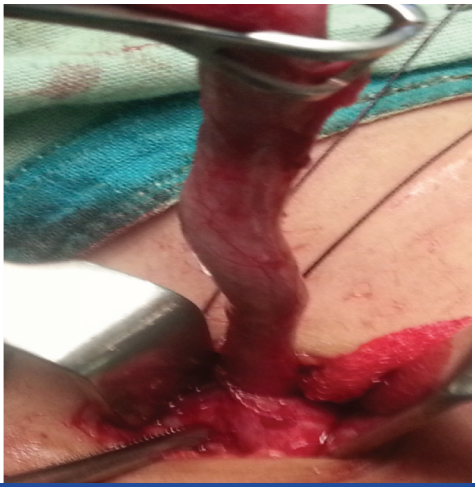
A four and half-year-old male child was admitted with 4 days history of pain in the lower abdomen associated with vomiting and fever for 2 days. On examination the child was febrile and tenderness felt at the right iliac fossa. The Total Leukocyte Count was raised- 15,600cumm. Diagnosis of acute appendicitis was made provisionally and supported by ultrasound finding of the abdomen which showed the appendix measuring 14mm in diameter with adhesions. The child was managed with non operative measures and underwent interval appendectomy 4 months later. A Grid iron incision was made on the right iliac fossa and the cecum was identified. The taenia coli were traced down to its confluence and the base was identified. Attempt was made to release the tip but found out that it was communicating with the cecum by another stump. The two “bases” or “stumps” were positioned frontally on the cecum with a central mesoappendix [Table/Fig-1,2]. Appendectomy was completed [Table/Fig-3]. The postoperative recovery was uneventful and the patient was discharged on the 3rd postoperative day.

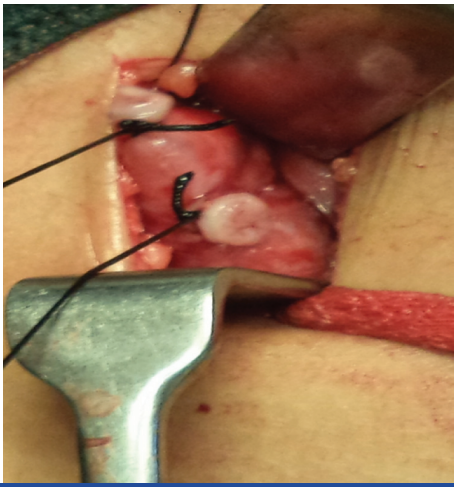
DISCUSSION

Appendiceal anomalies are extremely rare. Collin studied 50,000 appendix specimens and in the study he reported only four cases of agenesis (0.008%) and still rarer, he reported only two cases of duplication (0.004%) [1], however, no case of horseshoe appendix was reported. Cem ORUÇ et al., reported the fourth case in July 2013 after which no new case has been reported as per our knowledge making this the fifth case of reported horseshoe appendix [2].

The aetiology of double appendix or horseshoe appendix is still unclear because of its rarity and the extremely limited cases that are reported so far. A “transient appendix” which differs from the normal appendix has been observed in human embryos [3]. Some theories that has been put forward is that the horseshoe appendix may develop from the fusion of the tip of the normal appendix with that of the variant appendix which is at the abnormal site thereby giving rise to the so called horseshoe anomaly or, the other possible explanation could be that the horseshoe appendix develops from the fusion of the tip of the normal appendix with another part of the caecum which later on become the second base. However, these theories doesn’t seem to explain why the appendix was supplied by a single blood vessel in the mesoappendix with its tributaries spreading out rather than forming an arcade on the inner aspect of the horseshoe as seen in the cases reported which also stands true in our case too. Thus the most likely explanation could be that it is due to some abnormality arising during the embryologic period, which could be perhaps during the embryologic life, the base of the appendix somehow split in two, and during the course of development and cecal growth gets separated further leading to a double-based, yet single structure [4,5]. However, more cases are needed to be studied to support the claim in establishing the cause for horseshoe appendix.







[Table/Fig-1]: (1) The two bases of the appendix seen at frontal disposal. **[Table/Fig-2]:** The divided and undivided stumps. **[Table/Fig-3]:** Completed appendectomy showing the two divided bases

In 1963 Wallbridge updated and modified Cave's classification [6]. However, horseshoe appendix doesn't appear in this classification system. Wallbridge classified appendiceal duplication into;

Type A: A single appendix base with various degrees of duplication of the tip.

Type B: Two separate appendixes from one caecum.

B_1 - Two appendixes on either side of the ileocaecal valve, similar to the arrangement found in birds "Bird like". This type of appendiceal abnormality is usually associated with other gastrointestinal (Including hindgut) mal development (ileum, colon, anus) and urinary malformation.

B_2 - There are two appendixes, one at the normal site, and the other usually on the taenia in the caecum at a varying distance from the first, also known as "Taenia-coli type". This type of anomaly is not usually associated with other congenital anomalies.

Type C: Two caecums, each with its own appendix.

Biermann in 1993 classified appendiceal anomalies into [7]:

- A: Partial duplication of the appendix on a single cecum;
- B: Two completely separate appendices on a single cecum with two subtypes:
  - B1: "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.
  - B2: "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.
  - B3: The second appendix is located along the tenia of the hepatic flexure of the colon.
  - B4: The second appendix is located along the tenia of the splenic flexure

The later three (B2, B3 and B4) are usually not associated with other congenital anomalies.

- C: Two caecum, each bearing an appendix. This type occurs in association with hindgut mal development (ileum, colon, anus) and other anomalies of genitourinary tract and lower vertebral column.
- D: Three completely separate appendices with or without other anomalies.

Calota F et al., classified appendiceal anomaly into "Number anomalies" and "Shape Anomalies" [8].

Number Anomalies

1. Congenital Agenesis
2. Multiforious appendices

A: Appendiceal duplication(Partially) - "Y shaped"

B: Duplex Appendix on a single cecum:

B_1- "Avain type" with intestinal and/or genitourinary anomalies

B_2- "Tenia-coli cecum type"

B_3- "Tenia coli hepatic flexure type".

B_4- "Tenia coli splenic flexure type".

C: Duplex Appendix on two cecum (with hindgut, genitourinary tract, lower vertebral column maldevelopment.

D: Triplex appendix:

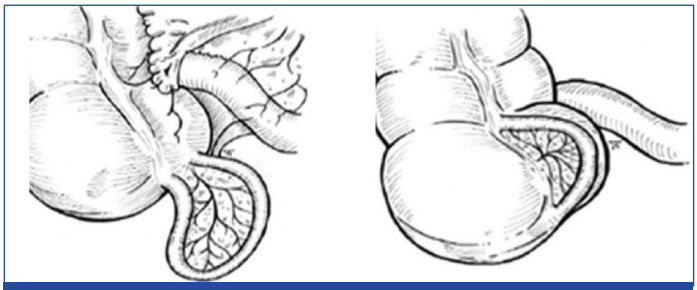
– "Newborn type" with/without other congenital anomalies.

– "Adult type" without other congenital anomalies.

Shape Anomalies

Horseshoe shaped appendix:

- With Frontal disposal [Table/Fig-4].
- With Sagital disposal [Table/Fig-5].



[Table/Fig-4]: With frontal disposal [8]. **[Table/Fig-5]:** With Sagital disposal [8]

The appendiceal anomaly can be associated with other congenital anomalies, which is more common in type B1 and type C. In our patient the anomaly was a horseshoe type with sagital location and there was no associated congenital anomaly. Radiology has been of not much help in the diagnosis of appendiceal anomalies preoperatively as was evident in our case.

CONCLUSION

Although rare the appendiceal anomalies may be encountered during any abdominal surgeries related to the appendix or otherwise. It is important for a surgeon to be aware of the possibility of anatomical anomalies or mal positions as legal case may arise. This case is being reported due to its particular rarity and also to enlighten the surgeons about the appendiceal anomalies as it is unlikely that most surgeons will ever encounter a case of horseshoe appendix in their whole life of practice as a surgeon.

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