

One Case of Appendiceal Duplication (The Rarest Type)

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Abstract: Since 1892, after the first report of the first case of appendicitis duplication, fewer than 100 cases have been reported worldwide with an incidence reported at 0.004. The first classification of appendicular duplications was developed in 1936 by Cave and then modified in 1962 by Walbridge. Since then, a number of authors have made some modifications, which has led to the classification by Cave-Walbridge, which is now the most used and offers three types: Type A corresponds to a complete or partial duplication of which only the base is common, Type B is the most common (60%) and has two subgroups; type B1 where the two appendages are symmetrically arranged with respect to the Bauhin valve; Type B2 where the appendix is in the usual laterocecal position and the second hypoplastic located on a colic strip at a distance from the first, Type C corresponds to a caecal duplication where each cecum carries a proper appendage. It is a condition that occurs most often in the first years of life and its discovery is often fortuitous on the occasion of a laparotomy or laparoscopy for another pathology as the case of our patient who presented in an occlusion chart and in whom surgical exploration found a Caecoappendicular type C duplication and this is the rarest type in appendicular duplications. However, before any abdominal surgery, the surgeon must therefore examine the cecum so as not to miss a caecoappendicular duplication because it is a rare pathology certainly, but it can cause many problems if it is not diagnosed at the time of surgery.

Keywords: Appendicular duplication, caeco-appendicular duplication, digestiv duplication.

INTRODUCTION

Digestive duplications are rare congenital malformations. Appendiceal duplication is exceptional, with a reported incidence of 0.004. It is a condition that occurs most often in the first years of life, sometimes some forms can remain asymptomatic and do not express themselves until adulthood.

Very little research has been reported on this issue. Thus, we report the observation of a newborn who presented a clinical and radiological picture of abdominal occlusion and in whom the surgical exploration highlighted a caecal appendix duplication which is the rarest form of appendicular duplications.

MATERIALS & METHODS

A 17-day-old neonate born of a well-attended pregnancy carried out, vaginal delivery assisted with a 10/10 Apgar at birth, the meconium has been emitted within 24 hours after birth and which presented at 2 weeks of life an abdominal occlusion with bilious vomiting and abdominal bloating, the whole evolving in a context of apyrexia, an abdomen without

preparation was asked which objectified hydro-aeric levels of grelic type with pelvic vacuity, the abdominal ultrasound showed an agglutination of the loops at the level of the right iliac fossa without visualization of the appendix, and the index to the gastrograffin was without particularity.

The newborn was then admitted to the operating room; the surgical exploration found a caecal duplication where each caecum was carrying a clean appendix; one of the two appendages was hail-related causing gastrointestinal occlusion without signs of digestive distress (*Figure 1-2*), then we proceeded to the appendectomy of the 2 appendices, thus liberating the hail leaving in place the 2 caecums because they were communicating.

The operative sequences were simple with a favorable evolution. The newborn was declared outgoing three days after the procedure, he was seen again at the consultation one month after his intervention he was well and presented no complications.



Fig-1: Image of a caecoappendicular duplication discovered intraoperatively



Fig-2: Image showing two appendages from two different caecums

DISCUSSION

Intestinal duplications are rare and poorly described congenital anomalies in the literature, from childhood to adulthood, but two-thirds occur in the first year. They present themselves classically in the intestine, on the mesenteric side, with whom they share the same musculature and the same vascularization [1]. Caecal location is not the most observed; it is described in less than 1% of cases [2, 3]. Less than 100 cases have been reported since its first description in 1892 by Picolo [4-5]. Collins reported four cases (0.0008%) in a study of 50 000 specimens of the human vermiform appendix [6]. F. Calotã *et al.* reported a case in a 45-year-old patient [7].

The causes of this anomaly is unclear due to its rarity and lack of consensus of opinion on this subject. Four main theories are advanced to try to explain the occurrence of these intestinal duplications.

The first one is the notochordal split theory via adhesion between ectoderm and endoderm in the embryonic phase, which explains the association with vertebral anomalies in 15% of cases. The second suggests a lack of regression of the embryonic diverticulum, which would make it possible to understand the occurrence of this anomaly on different segments of the intestine. The third one evokes the formation of a medial septum which induces a flattening of the adjacent intestinal wall by extrinsic compression which would cause adhesion and fusion and, subsequently, splitting of light. The fourth and last

one evokes the parallel presence of two functional structures with little or no external malformation.

However, none of these theories alone explains the origin of the duplications [8]. Wallbridge has therefore proposed a classification specific to appendicular duplications that distinguishes three types [9].

Type A corresponds to a complete or partial duplication of which only the base is common, the partial cystic forms comparable to our observation being less frequent than the complete tubular forms.

Type B is the most common (60%) and has two subgroups: - type B1 where the two appendages are arranged symmetrically with respect to the Bauhin valve (bird-like type of Anglo-Saxon) - and type B2 where the appendix is in the usual laterocecal position and the second hypoplastic is localized on a colon strip at a greater or less distance from the first (Taenia coli type).

Type C corresponds to a caecal duplication where each cecum carries a proper appendage. Types B1 and C are frequently found in the context of genitourinary, digestive or vertebral tail malformations.

The usual clinical presentation of caecal duplications is by acute intestinal obstruction accompanied by vague pain. A mass is palpated at the level of the abdomen, which can however disappear

during the clinical examination according to the degree of communication with the intestinal lumen. This mass is found in only 20% of cases, most often with tubular duplications.

Ultrasound is an excellent diagnostic method, it shows the typical appearance of a duplication in the form of a triple layer. Intestinal duplication can also be diagnosed by CT-scan and NMR [1]. However, the correct preoperative diagnosis is made in less than 25% of cases.

Error of misdiagnosis most commonly include diagnosis of appendicitis, intussusception, omental cyst, cystic lymphangioma, dermoid cyst, ovarian, renal, adrenal cyst and intestinal malrotation [1].

Duplications are usually benign lesions; however, in adults, malignant degeneration has been reported [10]. No review of the literature has made it possible to assess this risk quantitatively on this subject.

The treatment of choice is the surgery which should not be more radical than necessary and which is indicated for 3 reasons: the lifting of symptoms, the prevention of the permanent risk of perforation and haemorrhage, and the risk of cancerization [11]. It consists of the appendectomy of the two appendages and the excision of the caecal duplication if it is not communicating.

CONCLUSION

The appendix caeco duplications are most often discovered during an intraoperative occlusive syndrome by the surgeon performing the procedure who is considered lucky surgeon, lucky by Cave.

Thus, the surgeon must be attentive to the presence of a second vermiform appendix or to the presence of a second cecum, the anatomopathological study of any appendectomy specimen with conservation of the results is of interest here, because if omitted, it may cause complications or medico-legal problems.

Here, as in many other cases, caecal duplication has been misdiagnosed clinically, since it has been interpreted as a grealous occlusion.

Although caecal duplications are rare, they must enter as a differential diagnosis in cases of acute or subacute intestinal obstruction, especially in children. Surgery is the treatment of choice, even for asymptomatic forms and fortuitous discovery.

CONFLICT OF INTEREST

The authors do not declare any conflict of interest and all contributed to the redaction of this publication.

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