Bifid appendix with appendicoumbilical fistula

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ABSTRACT

Umbilical drainage of gas in a newborn in the form of gas or intestinal contents is concerning for patency of the omphalomesenteric duct (POMD) or gastrointestinal fistulas. A three-day old female presented with feculent umbilical drainage caused by a Y-shaped appendix with fusion of one limb to the umbilicus. This appendicoumbilical fistula was repaired via transumbilical excision. To the best of our knowledge, this is the first report of an appendicoumbilical fistula involving a bifid appendix.

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Umbilical drainage of gas or intestinal contents in a newborn represents the hallmark finding for a patent omphalomesenteric (or vitelline) duct (POMD). Failure of the omphalomesenteric duct to obliterate during early fetal development results in this entity.

Other causes for feculent drainage are fistulous connections between the gastrointestinal tract and the umbilicus, not classified as POMD. Reports of connections between the appendix and the umbilicus are exceedingly rare. In a review of 50,000 specimens, Collins et al. reported only three cases of the human vermiform appendix (0.006%), but did not describe their etiology as congenital [1,2].

Anatomic variants of the appendix, especially a bifid appendix, is an extremely rare entity, with an incidence of 2 in 50,000 live births [3,4]. We present a case of an appendicoumbilical fistula arising from a bifid appendix.

1. Case report

A three-day old female infant, born at 38 2/7 weeks gestational age, was noted to drain meconium-like material and gas from a pinpoint opening at the inferior aspect of the umbilical stalk. This opening was probed with a 24F angiocath confirming a connection with the gastrointestinal tract (Fig. 1). The patient was taken to the operating room. We debrided the umbilicus from the dried umbilical stump and found a large patent segment of bowel fused to the umbilical ring (Fig. 2). The obliterated umbilical vessels were found at the cephalad aspect of the intestinal lumen. We used sharp and blunt dissection to circumferentially free the adhesions from the bowel to the fascial umbilical ring and entered the abdominal cavity. Following the ductal bowel structure we reached the appendix and subsequently exteriorized the cecum which was found to be mobile. Distal small bowel was also delivered through the umbilical ring (Fig. 3). We noticed a very unusual malformation with a long Y-shaped appendix with one limb extending to the umbilicus, forming an appendicoumbilical fistula (Fig. 3). We examined the entire small bowel and did not encounter any other malformation. The appendix, including the extension to the umbilicus, was tied off at its base with two 4-0 absorbable braided ties and amputated. The umbilical ring was closed with a figure-of-eight absorbable braided suture. A vacuum dressing was applied [5].

A postoperative upper gastrointestinal imaging series (UGI) ruled out malrotation. The patient was discharged home on postoperative day six in good condition. Gross examination of the specimen revealed a Y-shaped specimen, which showed bifurcation of the main lumen into two separate tubular structures (Y-shaped specimen). Histologically, the cross section of the stem of the Y-shaped specimen resembled an appendiceal wall with normal...
colonic crypts and submucosal lymphoid aggregates (Fig. 4A and B). The same mucosal lining was present within the two tubular structures as well, consistent with a diagnosis of bifid appendix. The serosal aspect of the specimen showed fibroinflammatory reaction and adhesions.

2. Discussion

We present the unusual case of an appendicoumbilical fistula from a bifid vermiform appendix. To our knowledge the combination of these two scarce anatomical findings has not been described and needs to be differentiated from an open POMD.

A patent omphalomesenteric duct (POMD) is present in 2% of population, found with equal frequency between gender and results from incomplete retraction and failure of obliteration of the connection between the yolk sac and midgut of the developing fetus during the fifth to ninth gestational week. This failure of duct involution results in various omphalomesenteric duct remnants. These remnants can further be classified according to the patency of the duct. If the duct remains patent an omphalomesenteric fistula is formed, also known as open omphalomesenteric duct [6].
A persistent sinus tract with patency of the intermediate portion but closure at both ends leaves an omphalomesenteric cyst, also known as closed omphalomesenteric duct [6,7]. POMD arising from the small intestine is common whereas the colon or the appendix is rarely involved. Other causes include perforation or iatrogenic causes like drainage of periumbilical abscesses, from the small intestine is common whereas the colon or the appendix in contrast to a small bowel fistula arising from the base of a solitary appendix. The appendiceal anomaly of a bifid vermiform appendix per se is an extremely rare finding with a reported incidence of 0.004—0.009% [3].

In contrast to a POMD which usually shows small bowel mucosa, the pathology in the current specimen showed colonic mucosa and thus helps to differentiate this appendiceal anomaly from a persistent omphalomesenteric duct arising from the appendix [1].

The etiology of appendicocutaneous fistulas is unknown. An incomplete retraction during the embryological development and defective fixation of the appendix to the umbilicus has been described as possible etiology [13,14]. The striking feature in this report is the finding of a true bifid appendix with fusion of one limb to the umbilicus resembling a POMD. We hypothesize that the duplicate appendix with its anchor-shaped anatomy impaired complete retraction due to increased mechanical resistance, leading to umbilical adhesions and formation of an appendicocutaneous fistula. In this context, the mobile cecum is the consequence of the appendiceal attachment to the umbilicus which suspended the cecum from the peritoneum, preventing the development of peritoneal attachments. In accordance with this theory, Fujikschot et al. associated a mobile cecum with an umbilical appendix [10].

Pal and Singh mention inflammatory processes, associated tumors, or iatrogenic causes like drainage of periumbilical abscesses, as possible etiologies for appendicocutaneous fistulas, the latter usually occurring later in life [2].

3. Conclusion

In this report we present a previously unknown combination of two exceedingly rare findings: an appendicocutaneous fistula, arising from a bifid appendix. Surgical exploration and treatment via transumbilical approach yielded the diagnosis with a virtually scarless result.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Conflict of interest statement

Alfredo D. Guerron, Deepa T. Patil and Federico G. Seifarth declare no conflict of interest, and disclose any financial and personal relationships with other people or organizations that could inappropriately influence their work.

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