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Case report

A rare case of duplication of the descending colon in a pregnant woman: case report and literature review

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Abstract. *Introduction:* Gastrointestinal duplications are uncommon congenital abnormalities that can occur anywhere throughout the intestinal tract. The small bowel is more interested than the large one. Duplications are schematically classified as spherical and tubular, respectively representing 80% and 20% of cases, with different relationships and communications with the native intestinal wall. Although typically diagnosed during infancy and early childhood, tubular colonic sub-type stays frequently hidden for several years until a complication occurs. *Case presentation:* we report the case of a T-shaped tubular duplication in a 20-year-old woman at the 30th week of gestation, who underwent an urgent exploratory laparotomy for intestinal occlusion, treated with the resection of the aberrant large bowel. The patient was notable for a long history of constipation and chronic pain. Diagnostic possibilities were limited by the on-going pregnancy. *Conclusion:* Intestinal duplications are uncommon malformations, and, of these, the T-shaped subtype of the colon is among the rarest ones. In the adulthood, diagnosis is usually established in the operating room during urgent or even emergency surgery performed for abdominal complications. A duplication of the descending colon is extremely rare, and this is, to our knowledge, the only article describing a case found in advanced state of pregnancy. (www.actabiomedica.it)

Key words: intestinal duplication, colonic duplication, acute abdomen, pregnancy, caesarean birth

Introduction

Gastrointestinal duplications represent a rare congenital malformation, which may occur in any site of the gut with a greater prevalence in the ileum, accounting for up to 60% of cases. Localization in the large bowel is reported in only 6-7% of cases (1) while the rectal region is affected in 5% of cases (2). There is a strong female predominance with a female to male ratio of 2:1. Most frequently gastrointestinal duplications are diagnosed in childhood, in 80% of cases before 2 year-age (2; 3). They may be isolated or associated with other congenital malformations, usually involving the skeletal, genital, or urinary systems, which could allow an earlier diagnosis (4). The patients suffering from tubular colonic duplication alone are usually asymptomatic or may just refer chronic constipation or recurrent abdominal pain. Radiological investigations show low sensitivity in the detection of such anatomic anomalies, which may therefore remain misdiagnosed for many years. The diagnosis of duplication is seldom established in adulthood, where more often it is an incidental finding, and extremely rarely in the gestational period so that there are very few cases reported in literature.

Herein we report a case of a 20-year-old pregnant woman who developed an intestinal occlusion due to a T-shaped colonic duplication complicated by volvulus

with bowel infarction. Due to the rarity of the disease, we conducted a systematic review of all the cases reported in the literature focusing on the classification and pathogenesis.

Case report

A 20-year-old woman at the 30th week of gestation underwent an urgent laparotomy due to intestinal occlusion of unknown origin with initial hemodynamic instability. Her clinical history was notable for chronic and severe constipation that developed progressively since her 10th year of age. Evacuation was referred to occur with an average frequency of 2-3 times per week, obtained in most cases only with enema administration and associated with chronic abdominal pain and asymmetric distension in the abdominal right lower quadrant. Due to the relapsing symptoms the patient was repeatedly referred to the emergency department but no cause could ever be established. Abdominal X-ray and ultrasound (US) examination only demonstrated the distension of the sigma with fecal deposits. Even two computed tomography (CT) scans could not identify any lesions other than the aforementioned findings, so that she was simply referred to the gastroenterology department.

A colonoscopy detected only a mild stenosis of the sigma with proximal distension and accumulation of solid feces without any lesions of the intestinal wall. Based on these findings, the patient was offered surgery to resect the stenotic sigmoid tract, which was refused. Subsequently she performed an X-ray intestinal transit study, which identified a slow-down of the colonic transit, with the persistence of markers in the probable sigmoid sling after 84 hours. Owing to the clinical suspicion of Hirschsprung disease or intestinal neuronal dysplasia a rectal manometry and biopsy were performed although with negative response. Laboratory tests excluded IBD. Clinical examination demonstrated a motor incoordination of the pelvic floor and abdominal muscles; therefore, a rehabilitative toilet training was started with partial improvement of the symptoms.

At the age of 17 she underwent a laparoscopic appendectomy due to acute phlegmonous appendicitis. No other lesions were evident during the laparoscopic abdominal exploration. The postoperative course was uneventful. After one year she suffered from an acute episode of adnexitis, conservatively treated with antibiotics. Thereafter chronic constipation persisted but the frequency and intensity of the episodes of abdominal pain decreased.

At the age of 20 she got pregnant, and the gestation proceeded without complications until the 30th week, when she was admitted to the Obstetric Department due to the acute onset of abdominal pain with diffuse tenderness, hyperpyrexia up to 38°C and inability to pass gas and stool.

US examination detected a marked distension of a likely colonic intestinal sling. No complications for the fetus were evident. Neutrophil leukocytosis (13 x 10^9/L), elevated PCR (7048nmol/L) and increased lactate (2mMol/L) were detected at laboratory tests. The prophylaxis for hyaline membrane disease was started, but it was limited to the first administration since the clinical condition of the patient worsened with the onset of biliary vomit and hemodynamic instability, so that she was submitted to urgent laparotomy. The abdominal exploration revealed a markedly distended left colon compressed by an intestinal sling of probable colonic nature, with a T-shape originating from the lateral side of the sigma, compatible with a congenital colonic duplication. The sling was 30 cm long, filled by a large fecaloma and affected by longitudinal torsion that caused wall infarction with incipient perforation. Since the vital parameters of the fetus worsened, a caesarean delivery was performed. The newborn showed a weight of 1560 g and an Apgar score of three and was consequently transferred to the Neonatology department. Thereafter a resection of the sigmoid tract associated with the origin of the duplicated colon was performed; intestinal continuity was restored with a colorectal latero-terminal mechanic anastomosis. No intraoperative complications occurred. The postoperative course was uneventful, and the mother was discharged on the 7th postoperative day. The newborn was discharged after one month and in good clinical conditions. Histological examination confirmed the diagnosis of congenital duplication of the colon without associated lesions.

Discussion

Intestinal duplications are rare congenital anomalies that can occur in any site of the intestinal tract. Colonic duplications represent 6.8% of the cases (3). Several classifications for colonic duplication have been published, mainly based on macroscopic morphological or topographic criteria. Gross et al. identified four types of duplications according to their tubular, double barreled, cystic, or spherical macroscopic structure. The fourth type is the most common one; it appears as a spherical structure contiguous to some part of the bowel, especially to the ileum (5). Several classifications were proposed over the years; In 1971, Kottra and Dodds (6) modified the previous classification from Smith (7), considering the anatomic localization. They distinguished 2 types: the first one, tubular or spherical in shape, limited to a tract of the colon or rectum; the second one generally involving the colon entirely and frequently associated with duplications of the genital or lower urinary tract or both.

Duplications can be connected with the native intestinal lumen through one or more communications, or they can be completely separated from it; they can also end in a genital, urinary or perineal fistula. The type 2 duplications frequently present fistulas with another hollow viscus or may be associated with an imperforate anus or a separate perianal anus. The lesions without communications are often dilated by the accumulation of secretions produced inside them; they can also protrude inside the lumen of the adjacent native colon causing intussusception. Double-barreled duplications with a good drainage through a distal communication with the colon or through a second anus, on the other hand, remain generally asymptomatic and undiagnosed unless they are associated to others congenital anomalies. Different varieties of duplications may also coexist in the same patient. In addition, intestinal malrotation, situs inversus, Meckel diverticulum, double appendix and other anomalies can be associated with duplications (8).

Some authors (9) recently proposed a simplified classification, based on clinical purposes, that distinguishes duplications in simple cystic (type I), diverticular (type II) and tubular structures (type III). The latter, in the case of colic localization, are further divided in T or Y-shaped according to the angle created with the native intestinal wall (10). T-shaped colonic duplications are very uncommon in the literature; they characteristically present a wall consisting of an own mucosa, muscular layer and serosa and they are endowed by a proper peristalsis (11). They typically arise from the mesenteric border of the bowel, and they may have one or more direct communications with the adjacent part of the bowel across the common septum (12).

Pathogenesis

Several theories attempting to explain the pathogenesis of these anomalies have been proposed, but the exact mechanism is still unknown. Considering the complex variability of intestinal duplications, a singular theory appears not exhaustive and sufficient to explain the implicated mechanisms (13). Among the many theories, aberrant luminal recanalization seems to be the favorite one. During the 6th developmental week, embryonic bowel (hindgut, midgut, low gut) goes through a stage in which an intense epithelial proliferation occludes the lumen determining a condition named “solid stage”. A process, known as vacuolization, follows this step, which lead to the reconstitution of a single lumen in esophagus, intestine and colon, through the coalescence of vacuoles. Duplications seem to be the result of an incomplete vacuolization between the 5th and 7th week of embryogenesis. Instead of a single lumen channel, two or more parallel channels are formed by the coalescing vacuoles with or without communication whit the primitive gut (14). Many others etiological hypotheses have been proposed for duplications involving the large bowel. The embryonic diverticula one (15), among these, could explain some of these anomalies, like ours, but leaves unexplained those with proper circular and longitudinal muscle layers. Blinder supposed that tubular forms, as in our case, can be caused by an alteration of the lateral closure of the embryonic disk for an abnormality of the longitudinal line, while the cystic forms may originate from diverticula later in the evolution (16). Split notochord theory postulates a neural tube traction mechanism that could explain the 15% of cases of enteric duplications associated with vertebral defects (17).

"Abortive twinning theory" (18), "intrauterine vascular occlusion" (19) and recanalization defects are other significant theories attempting to explain the origin of intestinal duplications; hypoxia, trauma and environmental factors also may play an important role in determining this anomaly.

Clinical manifestations and diagnosis

Clinical history and examination as well as radiologic images can be nonspecific, putting this anomaly in differential diagnosis with several diseases of organic and functional nature. Clinical picture varies according to location, size, presence, and number of communications with adjacent bowel, compression of adjacent organs and presence of ectopic gastric mucosa within the duplication. Bowel endometriosis can also frequently be misdiagnosed and should be ruled out as a potential cause of chronic pelvic pain in women of reproductive age, particularly in patients presenting prolonged, less specific bowel-related symptoms. (20, 21)

Intestinal obstruction and volvulus are common clinical manifestations, while bleeding and perforation are rarer (22); they may also be asymptomatic or misunderstood until adult age or exhibit only minor symptoms such as constipation. T shaped colonic duplications possessing a proper muscular layer and a proper peristalsis that allow the emptying of secretions, may not give any symptom at all. Anyway, a long history of constipation (more than 10 years), abdominal pain and cramps may constitute typical symptoms, as our patient was notable for.

Traditional radiology may be a helpful diagnostic tool, but sometimes the interpretation of the images could be challenging. Diagnostics rely on US, barium enema, CT scans and magnetic resonance images (MRI). Pre-operative diagnosis has changed in the last 25 years because of the overwhelming use of abdominal ultrasound. In cystic type duplications, for instance, US scans typically show a hypoechoic mass with strong posterior wall echoes due to the fluid content, although sometimes hemorrhage or tissue thickening may give rise to more echogenic aspects. If the typical inner hyper-echogenic mucosal layer and the outer hypo-echoic muscle layer are detected on US, the diagnosis of duplication can be established (23), but when necrosis or alterations of the mucosa occur, this sign may not be present, as in our case. Moreover, ultrasound is not so much accurate in the detection of the tubular subtype and may be limited by the presence of high quantity of gas. Abdomen X-Ray can show a collection of gas in apposition with the colon or gas filled structures (3). After contrast enema administration, duplications may appear as extrinsic mass with respect to the intestinal tract or, in case of communication with the lumen, they can be opacified by the penetration of contrast medium inside (4). CT scans may depict the location and the extent of the duplication as well as the relationship with the neighboring organs. Moreover, it can also demonstrate the presence of vertebral malformations or other associated anatomic anomalies (24). Duplication cysts can be recognized on CT as smoothly rounded, fluid-filled cysts or tubular structures with thin, slightly enhanced walls, inside or adjacent to the wall of part of the alimentary tract. In our case, X-ray and tomography were not performed due to the ongoing pregnancy. MRI provides, in most cases, the same information as CT, but it is relatively expensive and less accessible (25). However, MRI can be done in pregnant patients without contrast medium and it is more appropriate in childhood. In addition, Technetium-99m pertechnetate scintigraphy can be helpful for suspected esophageal, duodenal, and small-bowel lesions that contain ectopic gastric mucosa, especially when findings in the previous instrumental examinations are highly suggestive for the diagnosis of duplication (22).

Treatment

The treatment of choice in colonic duplications is surgical resection. Type and extension of surgery depend on the size and location of the duplication as well as on the number of communications with the native colon. A segmental colon resection could be performed in simple and little spherical or tubular duplications. However, it is difficult to have an accurate and complete diagnosis before surgery, so the choice is often taken in the operative theatre. Urgent surgical interventions are often performed for bleeding, acute abdomen, volvulus, intussusception, gangrene, perforation, and peritonitis. Resection is also indicated in

case of diagnosis of ectopic gastric mucosa to prevent potential complications, such as perforation, and to eliminate the risk of malignant transformation (26). The resection must always include the origin of the duplication and, in case of inflammation of the duplicated tract, it must be pushed at least 2 cm far from the origin itself to prevent an anastomotic stenosis caused by post inflammatory fibrosis.

Reported cases of colonic duplication

Only few cases of T-shaped colonic duplications are reported in the literature. In our case radiological investigations were limited by the on-going pregnancy, so we were unable to identify the duplication pre-operatively. The review of the literature anyway shows that it is quite usual and many patients with unspecific symptoms perform surgery without an accurate preoperative diagnosis (12; 27). In our pregnant patient the duplication originated from the descending colon, which was dislocated medially from the gravid uterus in the 30th gestational week. We conducted a review of the cases of intestinal duplications in pregnant women, which are very few (table 1).

A.K. Cook et al. reported a case like ours; they described a retroperitoneal T-shaped colonic duplication presenting in a pregnant woman, which originated from the descending colon (28) and was not associated with genitourinary or skeletal anomalies or with the presence of ectopic gastric mucosa (29). When associated to these anomalies (60-75%), duplications are usually detected earlier, rarely remaining unknown until adulthood (10).

We noticed that in all four cases found in literature, including ours, the patients were not Caucasian; three of them were of African origins (28, 30) and one was from South America (31). We haven't found other similarities in terms of maternal age, gestational week, site, and size of the duplication.

Returning to our case, we decided to perform an urgent exploratory laparotomy, although an accurate preoperative diagnosis could not be established due to the progressive clinical deterioration of the patient. We noticed that the increased volume of the uterus during pregnancy created a situation in which feces were facilitated to enter inside the duplication, dilating it gradually. The intraluminal pressure increased progressively leading to inflammation and pain. We performed a resection of the tract of sigma comprising the origin of the duplicated colon, similarly to what is reported in other cases in literature. However, some authors suggest that T-shaped colonic duplications have to be removed without sacrifice of the native bowel, limiting the resection to the originating tract on the native colon (29). Laparoscopy may represent an option if a proper preoperative assessment is performed, with a barium enema, and if size and localization allow it (32). In our case the open approach was justified by the rapid clinical deterioration of the patients, joined to the ongoing pregnancy for which the possibility to perform an abdomen RX or CT was excluded, making it difficult to formulate a correct diagnosis.

Conclusions

Intestinal duplications, especially colonic, represent a condition where diagnosis remains a real challenge often not achievable prior to surgical exploration. In pregnant women, the diagnosis is even more difficult due to the complex general clinical picture in which an acute abdomen could develop requiring instrumental investigations such as abdomen CT, generally contraindicated in these patients. In conclusion

Table 1. Review of intestinal duplications in pregnant patients.

Reference	Age	Race	Localization	Length	Week's gestation
Omar Ragab	31	Sud Americana	Total genitourinary and rectal duplication	-	34th
George A. Radich	39	African-American	Small-bowel	14.8	24th
A.K. Cook	27	African	Slenic flessure/ retroperitoneal	30	10th
Present case	20	African	sigmoid	35	30th

in front of a history of abdominal pain and chronic constipation, with inconclusive laboratory and instrumental tests, we should not forget the possible diagnosis of intestinal duplication, pathology that, although rare, can give rise to life threatening complications, if belatedly diagnosed.

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References

1. Puligandla PS, Nguyen LT, St-Vil D, Flageole H, Bensoussan AL, Nguyen VH et al. Gastrointestinal duplications. J Pediatric Surg. 2003; 38: 740-744.
2. Kekez T, Augustin G, Hrstic I, Smud D, Majerovic M, Jelincic Z et al. Colonic duplication in an adult who presented with chronic constipation attributed to hypothyroidism. Worl J Gastroenterol. 2008; 14: 644-6.
3. Mourra N, Chafai N, Bessoud B, Reveri V, Werbrouck A, Tiret E. Colorectal duplication in adults: report of seven cases. J Clin Pathol. 2010; 63: 1080-3.
4. Kaur N, Nagpal K, Sodhi P, Minocha VR. Hindgut duplication-case report and literature review. Pediatr Surg Int. 2004; 20: 640-2.
5. Gross RE, Holocomb GW Jr, Farber S. Duplications of the alimentary tract. Pediatrics. Apr 1952; 9 (4): 448-68.
6. Kottra J, Dodds WJ. Duplication of large bowel. AJR Am J Roentenol. 1971; 113: 310-5.
7. Smith ED. Duplication of the anus and genitourinary tract. Surgery. 1969; 66: 909-21.
8. Shah KR, Joshi A. Complete Genitourinary and Colonic Duplication. J Ultrasound Med. 2006; 25: 407-11.
9. McPherson AG, Trapnell JE, Airth GR. Duplication of the. Duplication of the colon. Br J Surg. 1969; 56: 138-42.
10. Correia-Pinto J, Romero R, Carvalho JL, Silva G,. Neonatal perforation of a Y-shaped sigmoid duplication. J Pediatric Surgery. 2001, Vol. 36, p. 1422-4.
11. Trotovsek B, Hribernik M, Gvardijancic D. Giant T-shaped duplication of the transverse colon. A case report. J Pediatr Surg. 2006; 41: E59-61.
12. Holcomb GW, Gheissari A, O'Neill JA et al. Surgical management of alimentary tract duplications. Ann Surg. 1989; 209: 167-74.
13. Iyer CP, Mahour GH. Duplications of the alimentary tract in infants and children. J Pediatr Surg. 1995; 30: 1267-70.
14. Bremer JL. Diverticula and duplications of the intestinal tract. Arch Pathol. 1944; 38: 132-40.
15. Lewis FT, Thyng FW. Regular occurrence of intestinal diverticula in embryos of pig, rabbit, and man. Am J Anat. 1908; 7: 505-19.
16. Blinder G, Hiller N, Adler SN. A double stomach in an adult. Am J Gastroenterol. 1999; 94: 1100-1102.
17. Bentley JF, Smith JR. Developmental posterior enteric remnants and spinal malformations: the split notochord syndrome. Arch Dis Child. 1960; 35: 76-86.
18. Edwards, H. Congenital diverticula of the intestine: With the report of a case exhibiting heterotopia. Brit. J. Surg. 1929; 17: 7-21.
19. Favara B.E, Franciosi R.A, Akers D.R. Enteric duplications: thirty seven cases. A vascular theory of pathogenesis. Am J. Dis Child. 1971; 122: 501-506.
20. Barra F, Biscaldi E, Scala C, et al. A Prospective Study Comparing Three-Dimensional Rectal Water Contrast Transvaginal Ultrasonography and Computed Tomographic Colonography in the Diagnosis of Rectosigmoid Endometriosis. Diagnostics (Basel). 2020;10(4):252.
21. Raffaelli R, Garzon S, Baggio S, et al. Mesenteric vascular and nerve sparing surgery in laparoscopic segmental intestinal resection for deep infiltrating endometriosis. Eur J Obstet Gynecol Reprod Biol. 2018;231:214-219.
22. Brown RI, Azizkhan RG. Gastrointestinal bleeding in infants and children. Semin Pediatr Surg. 1999; 8 (4): 202-9.
23. Patiño Mayer J, Bettolli M. Alimentary tract duplications in newborns and children: diagnostic aspects and the role of laparoscopic treatment. World J Gastroenterol. 2014 Oct 21; 20(39):14263-71.
24. Di Serafino M, Mercogliano C, Vallone GF. Ultrasound evaluation of the enteric duplication cyst: the gut signature. J Ultrasound. 2015 Nov 23;19(2):131-3.
25. Abdur-Rahman LO, Abdulkadir AY, Nasir AA, Ibrahim OOK, Adeniran JO, Adesiyun OAM. Gastrointestinal Duplications: Experience in Seven Children and a Review of the Literature. Saudi J Gastroenterol. 2010; 16(2): 105–9.
26. Inoue Y, Nakamura H. Adenocarcinoma arising in colonic duplication cysts with calcification: CT findings of two cases. Abdom Imaging. 1998; 23: 135-7.
27. Hsu CF, Huang FC, Ko SF et al. Duplication of transverse colon: report of one case. Acta Paediatr Taiwan. 2003; 44: 47-9.
28. Cook AK, Dorman G, Redman CW. A duplication of the descending colon presenting in pregnancy. Journal of Obstetrics and Gynaecology. 1999; 19 (4): 423.
29. Chang YT, Lee JY, Liao YM, Chiou SS. Laparoscopic resection of a giant retroperitoneal T-shaped duplication of descending colon. Journal of Pediatric Surgery. 2008; 43: 401-4.

30. Radich GA, Altinok D, Adsay NV, Soulen RL. Papillary adenocarcinoma in a small-bowel duplication in a pregnant woman. AJR Am J Roentgenol. 2006 Mar; 186(3): 895-7.
31. Ragab O, Landay M, Shriki J. Complete cloacal duplication imaged before and during pregnancy. J Radiol Case Rep. 2009; 3(11): 24-8.
32. Kiu V, Liang JT. Laparoscopic resection of Y-shaped tubular duplication of the sigmoid colon: report of a case. Dis Colon Rectum. 2010; 53 (6): 949-52.

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