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ABSTRACT

Aim: to delineate the symptomatic features and to emphasize the necessity of early diagnosis and complete surgical excision of rectal duplications.

Method. We undertook a retrospective and contemporary review of all patients. Clinical recordings, preoperative evaluations, intraoperative and histological findings, and current patients' condition were studied.

Results. Age of the six patients ranged from new-born to 13 years. There was a broad spectrum of clinical presentation: two children were seen after previous therapy elsewhere with a mistaken diagnosis of perianal fistula, respectively undefined abdominal pain; two presented with exstrophic duplication of the rectum; one neonate was seen with an anal cleft and one infant with rectal bleeding and retrorectal palpable tumour. Paraclinical investigations established preoperative diagnosis in one patient, aided it in two others, and detected associated anomalies in two further patients. All duplications were "in toto" removed using laparotomy (n = 1), transanal (n = 1), or perineal sagittal approach(n = 4). All duplications had contact with the rectum. Smooth muscle coat and intestinal epithelial layer were histological demonstrated in each case.

Conclusions. Rectal duplications are rare anomalies. Clinical manifestations may include abdominal pain, obstipation, rectal bleeding, urinary or bowel obstruction, rectal polyp, perianal fistula, perineal abscess, and pelvic, abdominal, retroperitoneal or perineal mass. Early diagnosis avoids prolonged symptomatic treatment and unnecessary operative procedures. Complete excision is curative.

Key words: rectal duplication; exstrophic duplication of the rectum

INTRODUCTION

Congenital duplications of the alimentary tract are rare but potentially dangerous anomalies. There is no sex predominance. Any segment of the intestinal tract may be concerned, but small bowel is more involved. Among the 764 cases of Daudet [1], 490 (64%) were small bowel duplications (57% jejunum and ileum, 7% duodenum), and 38 (about 5%) were duplications of the rectum.

Duplications are cystic or tubular structures located usually adjacent to the mesenteric border, but other locations were also reported [2, 3, 4]. Rectal duplication may have diverse presentations, which include bowel or urinary obstruction, haemorrhage, infection, perforation, chronic obstipation, perianal fistula, perineal abscess, tumour of the labia major, exophytic tumour of the perineum, asymptomatic mass, pelvic floor hernia [5, 6, 7, 8, 9, 10, 11, 12, 13, 14]. Therefore the diagnostic is often delayed or incorrect. The early complete excision is the choice therapy of the alimentary tract duplications. That is particularly important in rectal duplications because of the risk of late malignant changes [15, 16, 17].

MATERIAL AND METHODS

This review encompasses 6 patients with clinically different manifestations, 4 of them diagnosed and cured by first admission, while 2 have been treated elsewhere over a long time period for perineal abscess, respectively undefined abdominal pain. All patients were diagnosed and treated in our department from September 1992 to march 1996. This study used the patients charts, preoperative investigations, intraoperative findings and histological examinations. All patients underwent clinical follow-up 12 - 22 y (mean 17 y) postoperative.

Case 1 (Surgery 09/1992). An 11-month-old boy was brought to our clinic after a 5 months history of perineal abscess. He was twice operated but symptoms did not disappeared. At admission he presented an inflamed retroanal fistula and had painful defecation. Putrid secretion flowed throughout fistula. Sonography findings were compatible with a retrorectal cystic tumour. After 7 days of antibiotic therapy and local betajodine bath the inflammation ceased. By a posterior sagittal approach the retrorectal cystic tumour was removed. Histological examination revealed colonic structures.

Case 2 (Surgery 03/1993). An 11-days-old male was admitted with a mucosal-lined skin defect and an exophytic mass left perineal. The mass in contiguity with the rectum had a separate perineal opening (fig. 1). Clinical examination revealed hemihypertrophy with asymmetry of the pelvis, hip luxation, left thigh hypotrophy, and clubfoot on the left side. The following investigations were performed: plain x-ray and sonography of the pelvis and abdo-

men, echocardiography, micturating cystourethrogram, diuretic nephroscintigram, cystoscopy with retrograde ureteropyelography, magnetic resonance imaging (MRI), and urodynamic examination. These investigations showed the absence of musculus gluteus maximus and musculus piriformis, absence of sacrotuberous and sacrospinosus ligament, no foramen ischiadicum majus and minus. Instead of the last two there was a defect where through part of the colon and left kidney herniated subcutaneously in the gluteal region. Additionally, the patient had an aortic isthmus stenosis, PDA, bilateral vesicoureteral reflux, caudal regression syndrome, tethered cord, lumbosacral lipoma, At 3-months the perineal mass was excised and histologicaly identified as colonic structure. The left kidney was relocated in the pelvis using a vicryl-net. Two years later the left kidney herniated again. A plasty with prolene-net was achieved. Twenty two years postoperatively the patient has regular bowel movements, normal renal function, no urinary infections, and good function of extremities. He is on medical therapy for hypertension.

Case 3 (Surgery 09/1993). A 3-days-old female infant with a birth weight of 3210g was brought to our department with an anal cleft at "3 o'clock" (with patient in supination). Pelvic sonography showed no pathologic findings. A contrast enema was carried out: there was a diverticular structure communicating with the rectum. A transanal resection followed when the child was 3 weeks old. The postoperative course was uneventful. Histological examination diagnosed colonic structures.

Case 4 (Surgery 07/1994). A 6-year-old female presented with an exophytic mass (7x5cm) of the labia minora (fig. 2). The mass covered by epithelium, had a lumen with an opening onto the vulva which through a probe was easy introduced. No other anomalies of outer genitalia, meatus urethrae or anus were observed. Paraclinical investigations detected a left ureteral duplication with ureteric ectopia and upper pole dysplasia, and vesicoureteral reflux of the lower pole. By a paramedian anterior sagittal approach the exophitic mass was excised. Intraoperatively a contact between the mass and the rectal wall was found. The histological diagnosis was rectal duplication covered by colonic and ectopic gastric mucosa. The dysplastic upper pole of the left kidney was removed by a subcostal incision.

Case 5 (Surgery 02/1996). A 13 ½ -yr-old boy was brought to our department after being treated over a long period for undefined abdominal pain. No pathological findings at physical examination were found. The sonography showed a precaval, subhepatic cyst with a diameter of 3 cm. Nuclear magnetic resonance scans (NMR) demonstrated the cyst located in the retroperitoneum. The cyst ended in the right side of the rectal wall and was filled with grey fluidly-mucous content (Fig. 3). The excision was carried out through a right supra-umbilical transverse laparotomy. A tailgut cyst lined by epithelium with gastric mucosa ectopy was demonstrated by histological examination.

Case 6 (Surgery 03/1996). A 3-month-old female infant was admitted for rectal bleeding. Rectal examination revealed walnut size tumour on the posterior wall of the rectum. Sonography showed a 3 x 2 cm cystic structure between sacrum and rectum. This tumour was removed by a posterior sagittal approach. The rectum and duplication shared a muscular layer. Six days after the operation a small dehiscence of the wound occurred. This closed spontaneously 10 days later. Histopathological exam identified a tailgut cyst with included ectopic gastric mucosa.

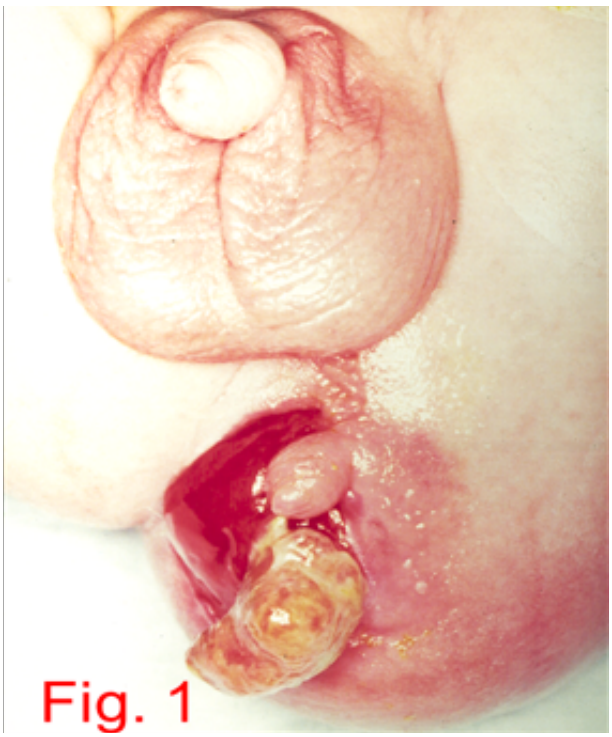


Fig. 1

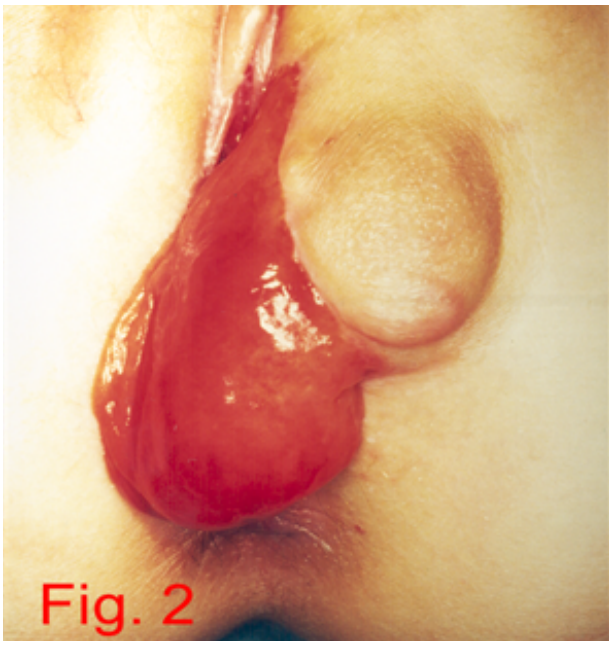
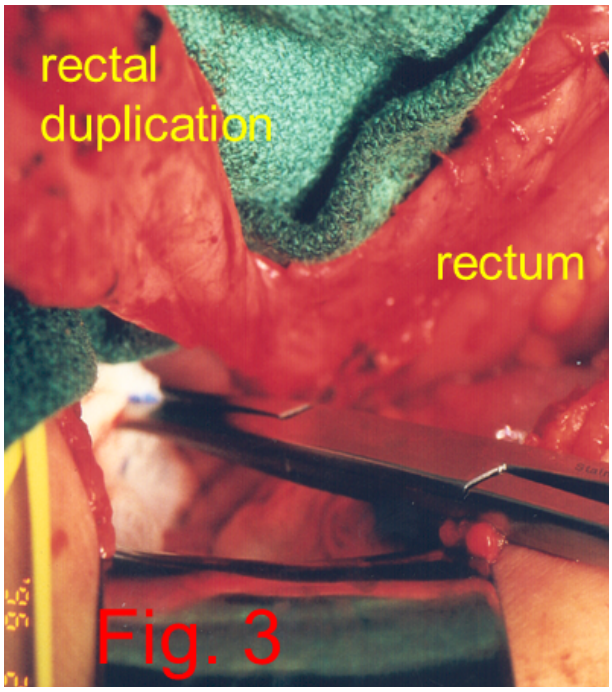


Fig. 2



The explanations of this material are presented in the text (red.)

RESULTS

The age at presentation of the 6 patients ranged from new-born to 13 ½ years (mean 3 4/12 yr.). The female: male ratio was 4 : 2. There was a broad spectrum of clinical presentation:

- two patients presented with extrophied perineal mass: one of them had multiple associated anomalies (case 2), the other only renal associated anomalies (case 4).
- one neonate female was diagnosed with an anal cleft at "3 o'clock" (case 3).
- one patient was seen because of rectal bleeding (case 6).
- two patients came to us after previous therapy elsewhere: the first with perineal swelling was twice operated erroneously for perianal fistula (case 1), the second treated for chronic abdominal pain (case 5) with medications.

The preoperative diagnosis was extrophy of the rectum in 2 patients (cases 2 and 4), retrorectal cystic tumour in 2 (cases 1 and 6), diverticular rectal duplication in 1 (case 3), and retroperioneal cystic tumour in 1 (case 5).

In three cases the preoperative diagnosis (associated anomalies excepted) was by clinical means only (cases: 2, 3, and 4), twice by clinical examination and sonography (cases:1and 6), once by sonography and MRI (case 5).

The surgical approach was perineal sagittal in 4 patient (posterior median in 2, posterior paramedian in 1, anterior paramedian in 1), transanal in 1, and laparotomy in 1.

Complete excision of the tumour was accomplished in each patient. All patients had intraoperative and postoperative antibiotic therapy, and were drained for 2 - 5 days postoperatively. Recovery was uneventful in all patients, except for a small wound dehiscence (case 6). Histological anatomy is shown in table I. The follow-up (mean 17 years postoperative) shows good function, good cosmesis in all cases, without complaints due to rectal duplication.

Table 1. Histological anatomy of the excised structures

>Small muscle coat	all
>Intestinal mucosa* - including crypts of Lieberkühn	all
>Gastric mucosa heterotopy	n = 3

**Taylgut cyst mucosa: cylindrical, transitional and squamous epithel, crypts of Lieberkühn*

DISCUSSION

The embryogenesis of these abnormalities is uncertain [9, 18]. The most satisfactory theories of alimentary tract duplications are the partial twining theory and that relating to the residua of the neurenteric canal. The dorsal anatomic location of most duplications is supportive of this last theory [9]. However more duplications have been found in other sites on the bowel circumference [2, 3, 4]. Perineal exophytic mass or tumour of the labia majora are other possible presentation forms of rectal duplications [19, 20, 21, 22, 23]. Two of our patients had a very special duplication form: the rectum extrophy (cases: 2 and 4). Another one has a retroperitoneal, prerenal cystic duplication with the caudal end in the lateral wall of the rectum (case 5).

Clinic examination and sonography in the case of 5 patients provided enough information to submit the patients for surgery. A patient needed supplementary MRI investigation to improve diagnosis (case 5). Because high rate of associated anomalies, all patients with rectal duplications will be thoroughly clinicaly and, in doubt, paraclinically examined.

Differential diagnosis of rectal duplications enclose all pelvic, and some abdominal and perineal tumours. Rectal duplications can be confused with rectal polyps, haemorrhoids, anal fistula (case1), and perirectal abscess [8, 10, 11, 24]. No patient in this series had duplication of the bladder, urethra or genitalia [25, 26, 27]. Only one patient had a unilateral ureteral duplication (case 4). There were no duplications in our patients communicating with urinary tract or intraspinal space [28, 29]. All lesions presented here fulfilled the criteria for alimentary tract duplications as defined by Ladd and Gross (30): a) contiguity with and strong adherence to same part of the alimentary tract; b) a smooth muscle coat; c) a mucosal lining consisting of one or more types of cells normally observed in the alimentary tract.

Presence of heterotopic gastric mucosa may be a source of rectal bleeding [7]. Malignant degeneration in rectal duplication in adults age is possible [15, 16, 17]. Carcinoid tumour in a rectal duplication in children have been also reported [31]. Therefore completely surgical excision is required.

COMCLUSIONS

These observations showed that the child with rectal duplication is a good candidate for surgical procedures planed to cure completely the child's suffering. Early diagnosis avoids prolonged symptomatic treatment and unnecessary surgical procedures.

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