IV. PEDIATRIC SURGERY

A 5 YEARS EXPERIENCE WITH THE DUPLICATION OF THE RECTUM

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Abstract
This paperwork presents several cases of congenital duplication according to localisation of the disease, type, complications, diagnosis and treatment.
Key words: congenital duplications.

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 change (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 period of time 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 patient's charts, preoperative investigations, intraoperative findings and histology. All patients underwent clinical follow-up 1 ½ - 5 y (mean 3 3/12 y) postoperative.

Case 1 (Op. 09/92)

An 11-month-old boy was recovered after a 5 months history of perineal abscess. He was twice operated but symptoms did not disappear. At admission he presented an inflamed retro anal fistula and had painful defecation. Putrid secretion flowed through fistula. Sonography findings were compatible with a retro rectal cystic tumour. After 7 days of antibiotic therapy and local betajodine bath the inflammation ceased. Through a posterior sagittal approach a retro rectal cystic tumour was removed. Histology: colonic structure.

Case 2 (Op. 03/93)

A 11-day-old male neonate was admitted with a left gluteal exophytic mass. He presented an asymmetry of the pelvis, his left thigh was hypotroph, his left foot was deformed (pes adductus). The following investigations were performed: plain x-ray and sonography of the pelvis and abdomen, echocardiography, micturating cystourethrogram, diuretic nephroscintigram, cystoscopy with retrograde ureteropyelography, nuclear magnetic resonance (NMR), urodynamik. These investigations showed the asymmetry and rotation of the sacrum and coccyx, left hip luxation, left kidney ectopy with gluteal herniation, vesicoureteral reflux II-III bilateral, spina bifida occulta with lipomeningocele and tethered cord, partial absence of the left gluteal muscles, cord anomaly, and

the left gluteal mass with continuity with the rectum. When he was 3 ½ months the mass was excised by the paramedian posterior sagittal approach.
Histology: colonic duplication.

Case 3 (Op. 09/93)

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"(pat. 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. Histology: colonic structure.

Case 4 (Op. 07/94)

A 6-year-old girl was examined for an exophytic mass at the lower pole of the left labium minus. No other anomalies of outer genitalia, meatus urethrae or anus were observed.

Through mass (7x5cm), wich was covered by epithelium a probe was easy introduced. Paraclinical investigations detected a left ureteral duplication with ureteric ectopia and upper pole dysplasia, and vesicoureteral reflux of the lower pole. By means of a paramedian anterior sagittal approach the mass was excised. The upper pole nephroureterectomy was achieved by a subcostal incision.

Histology: rectal duplication covered by colonic and ectopic gastric mucosa.

Case 5 (Op. 02/96)

A 13 ½ -year-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 excision was carried out through a right supra-umbilical transverse laparotomy. The cyst ended in the right side of the rectal wall (communication no visible) and was filled with grey fluidly-mucous content.

Histology: tailgut cyst lined by epithelium with gastric mucosa ectopy.

Case 6 (Op. 03/96)

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 demonstrated a 3 x 2 cm cystic structure between sacrum and rectum. This tumour was removed by means of a posterior sagittal approach. The rectum and duplication shared the muscular layer. Six days after the operation a small dehiscence of the wound occurred. This closed spontaneously 10 days later. Histology: tailgut cyst (ectopic gastric mucosa included).

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:

-2 patients presented with extrophied perineal mass: one of them had multiple associated anomalies (AA) (case 2), the other only renal AA (case 4).

-1 neonate female was diagnosed with an anal cleft at "3 o'clock"(case 3).

-1 patient was seen because of rectal bleeding (case 6).

-2 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), retro rectal cystic tumor in 2 (cases 1 and 6), diverticular rectal duplication in 1 (case 3), and retroperitoneal cystic tumor in 1 (case 5).

In three cases the preoperative diagnosis (AA excepted) was by clinical means only (cases: 2, 3, and 4), twice by examination and sonography (cases: 1and 6), once sonography and NMR (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. Follow-up (mean 3 3/12 yr. postoperative): good function, good cosmesis in all cases, no complaints.

Table I. 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. 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 duplication 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 (12,13). Two of our patients had a very special duplication form: the rectum extrophy (cases: 2 and 4). An other 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 NMR investigation to improve diagnosis (case 5). Because high rate of AA, 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 (8,10,11,18).

No patient in this series had duplication of the bladder, urethra or genitalia (19,20,21). Only one patient had an unilateral ureteral duplication (case 4). There were no duplications in our patients communicating with urinary tract or intraspinal space (22,23).

All lesions presented here fulfilled the criteria for alimentary tract duplications as defined by Ladd and Gross (24): 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 (25). Therefore completely surgical excision is required.

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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