A rare malformation of the alimentary tract, tailgut cyst : a case report

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Abstract

Tailgut cyst (cystic hamartoma) is a rare congenital pathology that arises -from post-natal primitive gut remnants in the retrorectal-presacral space. Because of the rarity of the lesion and the variability of the anatomical position, its diagnosis and surgical treatment are often difficult. Complete surgical excision of the multilocular and multicystic process prevents recurrent draining sinuses and eliminates the possibility of malignant change. We describe a case in which a tailgut cyst localized in the retrorectal and presacral space was characterized by abscess, repeated urinary tract infection, and rectal pain. (Acta gastroenterol. belg., **2018,** 81, **528-530**).

Keywords : Tailgut cyst ; retrorectal cystic hamartoma ; developmental cyst.

Introduction

Alimentary duplication cysts are rare anomalies that occur during the development of the digestive tract from the embryonic foregut, midgut, hindgut, and tailgut (1). Tailgut cysts, which are covered in this study and also known as retrorectal hamartomas, are infrequent congenital lesions that originate from remnants of the embryonic hindgut (2-4). These cysts are characterized by several variants of intestinal epithelial tissue, most often by columnar epithelium (3). They are generally benign, but malignant transformation has been reported on rare occasions (5-8). Tailgut cysts are almost invariably located in retrorectal or presacral space but have also been reported in various locations such as perirenal space, perineal skin, and, very rarely, prerectally (2,3,5,6). They are usually asymptomatic, but in rare cases they may cause local rectal compression, constipation, and urinary symptoms (9-11). However, the main problems associated with tailgut cysts are bleeding, infection, and malignant transformation (12).

We describe a case in which a tailgut cyst localized in retrorectal and presacral space was characterized by abscess, repeated urinary tract infection, and rectal pain, and we review the relevant literature.

Case report

A 6-year-old female was referred to our clinic with a few years' history of progressive constipation, pain at the time of defecation, and perianal pruritus. She was first treated at her regional hospital over an extended period of time after receiving a diagnosis of multiple urinary tract infections and constipation. On physical examination, a draining sinus near the anus was observed (Figure 1), and digital rectal examination revealed a rubbery mass posterior to the rectal wall, compressing the rectum and was not mobile. Additionally, the physical exam showed an anal sphincter with normal tone.

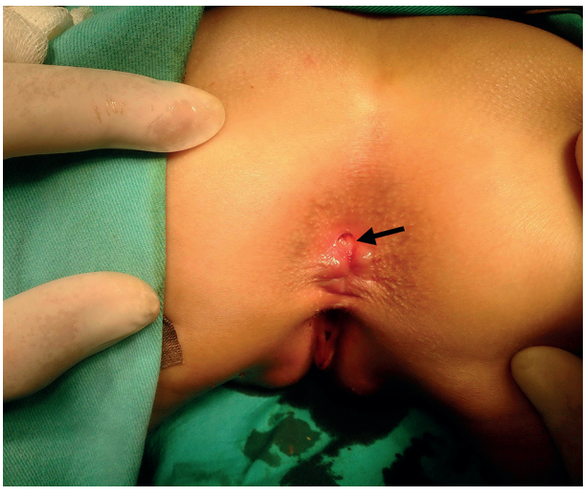

Fig. 1. — Tailgut cyst fistula orifice (arrow).

MRI of the pelvis showed a large retro-rectal multiloculated cystic mass with smooth walls and abscess content and without communication to the spine or the rectum (Figures 2a, b, c, and d).

The treatment of choice was complete excision of the lesion by a posterior approach (Figure 3). The lesion carefully could be separated from the rectum and pelvic wall. No communication was found with the rectal lumen. It was a multiloculated cyst filled with mucoid material. Coccyx was removed to excise any residual tailgut cyst remnants and to allow better visualization. The final pathological diagnosis was retro-rectal cystic hamartoma (tailgut cyst) with no evidence of malignancy. The patient was discharged after 5 days with no postoperative complications. The patient's post-operative course was uneventful, and, at four months post-surgery, our patient is symptom free with no evidence of recurrent or residual disease.

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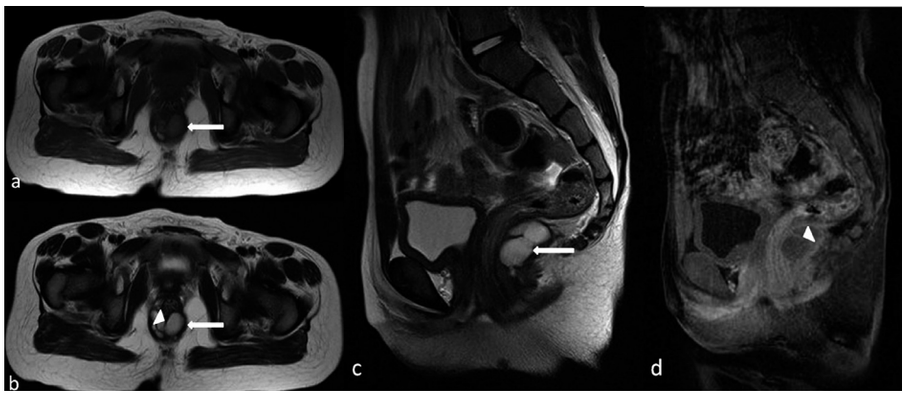

Fig. 2. — Tailgut cyst in 6-year-old female: (a) Axial T1-weighted MR image shows the well-defined, thin-walled cyst with intermediate signal intensity (arrow). (b) Axial T2-weighted MR image shows the well-defined cyst with homogeneous high signal intensity (arrow). Note the thick septum with low signal intensity (arrowhead). (c) Sagital T2-weighted MR image shows the well-defined cyst with homogeneous high signal intensity (arrow) in retrorectal space. Rectum is anteriorly displaced. (d) Contrast-enhanced sagittal T1-weighted MR image shows enhanced thick septa (arrowhead).

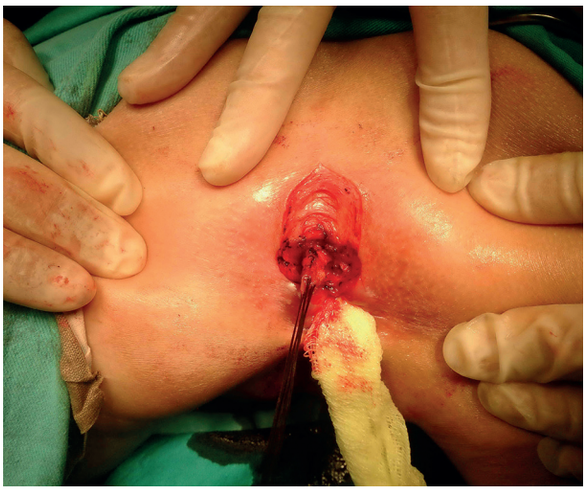

Fig. 3. — To remove tailgut cyst, identify of the plane between the cyst and surrounding tissues using blunt and sharp dissection. Intraoperative appearance of sinus tract and the cyst partly free from the surrounding tissues (black arrow). The white arrow shows the anus.

Discussion

The retrorectal area is a potential space surrounded by the rectum anteriorly, the sacrum and coccyx posteriorly, and the peritoneal reflection superiorly. A space-occupying lesion, such as a tailgut cyst, in this region can lead to different symptoms and a variety of clinical signs. (13,14). In the process of embryologic development of the tailgut or postanal gut, the embryo has a true tail, which reaches its largest diameter on the 35th day of gestation. Hereafter, the anus develops above the tail on the 56th day of gestation, by which time the latter has completely regressed (3,15). Tailgut cysts are a rare congenital lesion thought to arise from the embryonic postanal gut, well described by Hjermstad et al. in a series of 53 cases (6). They are often lined with mucin-secreting columnar cells, squamous, transitional, or a combination histology (12). Tailgut cysts may also be located in perirenal and subcutaneous areas (3,16).

They are usually discovered only incidentally, and symptoms often depend on the size and relation to the adjacent structures (6). Infection development is the most frequent complication (occurring in 40-50% of cases). Additionally, tailgut cysts are detected as asymptomatic masses but may be found in patients who present with abdominal pain, rectal bleeding, local abscess, and rectal fullness or constipation (15). There can also be a communication between tailgut cyst and skin; primary infection may progress to post-inflammatory fibrosis if left untreated (10-12). MRI is a suitable examination vehicle for preoperative evaluation of tailgut cyst. MRI improves tissue characterization because of its high contrast resolution between different tissue compartments. It also allows detection of mucin and blood in a cystic lesion (17). Needle biopsy of cystic presacral lesions is not recommended as a diagnostic tool because of the risk of life-threatening infections (18).

A tailgut cyst can transform into a tumor such as an adenocarcinoma, carcinoid, neuroendocrine carcinoma, or sarcoma (15). Hjermstad and Helwig (6) reported on one case (2%) of poorly differentiated carcinoma in a series of 53 tailgut cysts. Prasad et al. (7) reported on two cases (40%) of malignancy in a series of five tailgut cysts. Therefore, tailgut cyst need complete surgical excision to prevent (and treat) complications, such as infection, recurrence, and malignant transformation (19).

When a tailgut cyst is diagnosed, surgical excision is necessary because of the undesired, life-threatening complications in long-standing cases (17). Incisional biopsy provides only limited material for diagnosis, and subsequent follow-up excision always should be performed (6). Surgical approach depends on the

location within the pre-sacral space, size of the lesion, and the presence of complications. For most tailgut cysts, a posterior approach with or without excision of the coccyx bone will allow a good surgical view and removal of the multiloculated cyst (6,20,21). For high lesions that occupy most of the pre-sacral space, a trans-abdominal approach is usually required for treatment. For the treatment of large external tumors, a combined approach may be required (5).

The differential diagnosis of tailgut cysts should include sacrococcygeal teratoma, epidermoid cyst, rectal duplication cyst, anterior meningocele, and inflammatory cysts (22). In the differential diagnosis of presacral masses, the unilocular or multilocular characteristic is important. Tailgut cyst and lymphangioma are usually multicystic. The MR imaging may be useful for the differentiation of unilocular and multilocular masses (23).

In this case, the patient had received long-term treatment due to complaints of constipation, urinary tract infections, and perianal pruritus. However, upon careful examination of the perianal region, an orifice of the tailgut cyst opening to the outside at 6 o'clock in the lithotomy-position was seen. Additionally, we learned that there was occasional pus discharge from the orifice of the cyst. MRI examination detected septal cysts in the retrorectal field, and the tailgut cyst was removed by surgical excision.

In summary, tailgut cysts are rare congenital lesions originating from a remnant of the embryonic postanal gut tailgut. They often occur in the presacral retrorectal space, and MRI is a helpful technique to define the extent of the cystic mass and its relationship to the surrounding structures. When diagnosed this cystic lesions should be done definitive resection because of the risk of malignant transformation.

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