

Retrorectal Tailgut Cyst: A Rare Cause of Constipation Carcinoma: Case Report

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1. Abstract

Retrorectal tailgut cyst is located in the presacral space. They are congenital malformations that result from inborn errors of tissue development. These congenital cysts are more common in women and sometimes causes defecation problems in proportion to its size. They are generally benign lesions but malign transformation of the cyst has also been reported. In this case report, we present a 45 year-old woman who was admitted to the hospital with the abdominal pain and constipation for ten years. The tailgut cyst diagnosis was established with the magnetic resonance imaging. She relieved from her complaints with surgical excision.

2. Keywords:

Retrorectal Tailgut Cyst; Retrorectal Cystic Hamartoma; Kraske Operation

3. Introduction

Retrorectal cystic hamartoma, more commonly known as the retrorectal tailgut cyst, is located in the presacral space. They are congenital malformations that result from tissue development errors. These malformations are characterized by abnormal mixtures of mature tissue indigenous to that area. Hamartomas may be derived from any of the three primordial germ layers: ectoderm, mesoderm, and endoderm.

They sometimes causes defecation problems with the proportional to its size. They have no connection with the rectal lumen as long as it does not fistulized to the rectum because of the infection. Most of the time careful digital physical examination is enough for the diagnosis of the rectal submucosal mass. Pelvic magnetic resonance imaging (MRI) or computerized tomography scan is helpful in the differentiation of a solid or cystic mass. Surgical excision is the definitive treatment.

4. Case Presentation

A 45 year old female patient was complaining about abdominal pain and discomfortness from his bottom for ten years. She also had experiencing difficult defecation from time to time. Physical examination in the lithotomy position revealed a submucosal mass occupies right-posterior half the circumference of the lumen in the lower part of the rectum. There was no blood or fistula opening except for mucosal bulging in the rectal lumen colonoscopically. Laboratory findings were normal. MRI confirmed a 7x5cm cystic lesion with multiple thin septa between the posterior mesorectum and the coccyx (Figure 1).



Figure 1: Retrorectal tailgut cyst in a 45 year-old woman. On sagittal T2-weighted image shows a large cystic lesion with multiple thin septa between the rectum and the coccyx (arrow).

The operation was performed in the prone jackknife position (Kraske Operation) under the spinal anesthesia. The coccyx was excised to reveal the lesion entirely (Figure 2).

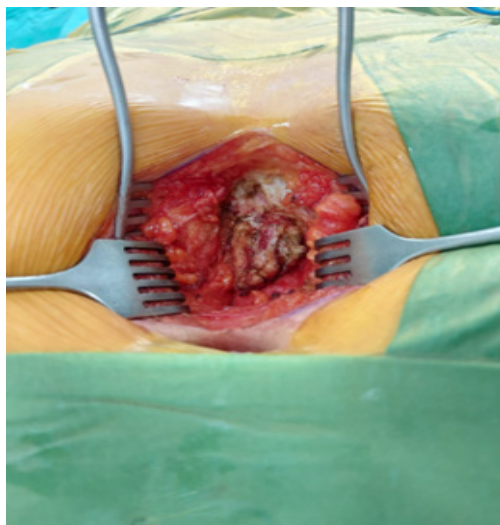


Figure 2: Surgical field of the patient in prone-jackknife position.

The cystic lesion was removed from the posterior aspect of the rectum without any damage to the fascia propria (Figure 3).

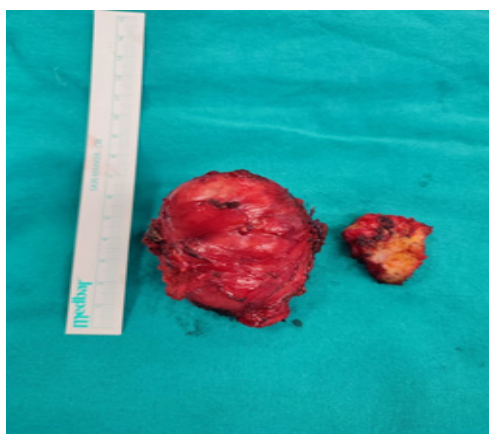


Figure 3: Retrorectal tailgut cyst and the coccyx

The wound was closed primarily with the help of a prolene mesh and a suction drain catheter was placed to the presacral area. The patient was discharged postoperative day five without any problem.

Pathological examination was revealed multiloculated cystic lesion lined with squamous, transitional, and mucinous epithelium in the sections (Figure 4).

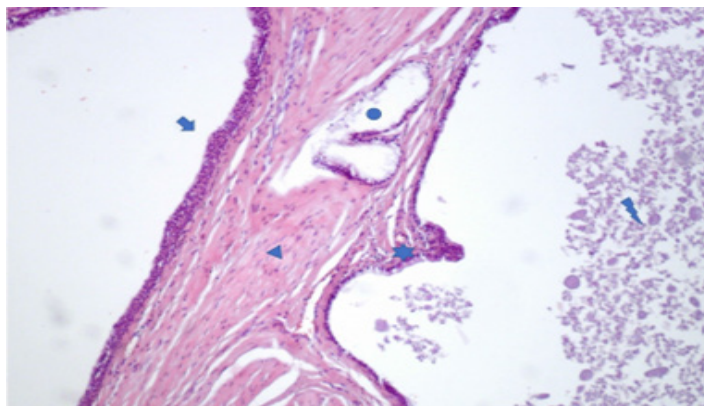


Figure 4: Histopathological photo of the multiloculated tailgut cyst. Cyst lined with transitional epithelium (arrow), smooth muscle fibers (triangle), mucinous glands (circle), cyst lined with

mucinous epithelium (star), organized proteinaceous material (lightning) inside the cyst are seen.(Hemotoxylin eosin x100)

Disorganized smooth muscle bundles were commonly noted adjacent to the lesion wall. While glandular structures are observed in the wall of cyst, some of them have keratinous material. The final diagnosis of the lesion was tailgut cyst and the malignancy was rule out. The patient provided written consent for the publication.

5. Discussion

Retrorectal tailgut cysts are congenital lesions that are derived from the hindgut germinal layers, and any type of intestinal tissue may predominate without a definite muscular or serous coat. Other retrorectal cysts may be confused with tailgut cysts. For instance, dermoid cysts are characterized by only a squamous type of epithelial lining. Duplication cysts have a well-defined muscular wall with a myenteric plexus. Tailgut cyst term includes either actual malformation with tissue excess present at birth or inborn tissue anomaly that manifests itself by excessive growth continuing until puberty. Therefore, it can occur at any age including childhood. According to the literature, Middledorff described the first presacral mass in a one-year old girl in 1885 [1]. The largest case series of tailgut cysts was reported by Hjernstad and Helwig in 1988 [2]. They had found female dominance in 53 patients and half of them were symptomatic with the most common symptoms being rectal pain and constipation.

Patients may be asymptomatic but perirectal pain, rectal fullness, tenesmus, and defecation problems are common symptoms if the mass is large enough. They are usually found to have an extrarectal mass as an incidental finding on rectal examination. Colonoscopic examination is unrewarding. Magnetic resonance imaging, computed tomography, and endorectal ultrasonography is useful for the differential diagnosis. Preoperative biopsy is unnecessary and should not be performed because of the perforation risk. Moreover, histopathological differentiation of the cyst from the other cystic lesions can be difficult with a biopsy sample.

Complete surgical excision of the tailgut cyst is the recommended treatment of choice because of the complication risks. The best known complication is the malignant transformation of the cyst which is rare. The first malignant tailgut cyst was described by Ballantyne in 1932 [3]. Adenocarcinoma of the cyst is the most common type of malignant tailgut cysts [4-7]. Mucinous carcinoma, neuroendocrine tumor, and carcinoid tumor have also been reported [8-10]. MRI may help the diagnosis of malignant transformation of these inborn retrorectal cysts [11]. Other complications are included infection, abscess, rectal or perianal fistula formation [12]. The type of surgery to be performed is determined by the location of the lesion. If the cyst is located above the sacral three level, abdominal open or laparoscopic surgery is done [13,14]. Robotic

surgery has been applied in recent years [15]. A combined abdominal and sacral approach should be performed when malignant transformation is suspected. Below the coccygeal level or levator muscle a posterior sacral approach (Kraske Operation) without entering the abdomen is appropriate, as was done in this case. Transanal endoscopic microsurgery is also feasible [16]. Non-surgical methods has no place for the treatment of the tailgut cysts.

6. Conclusion

Retrorectal tailgut cysts are not rare lesions. They should be considered in the differential diagnosis of rectal fullness and defecation problems. Complete surgical removal is the only effective treatment whether they cause complaints or not.

References

1. Killingsworth C, Gadacz TR. Tailgut cyst (retrorectal cystic hamartoma): report of a case and review of the literature. *Am Surg*. 2005; 71: 666-73.
2. Hjermsstad BM, Helwig EB. Tailgut cysts. Report of 53 cases. *Am J Clin Pathol*. 1988; 89: 139-47.
3. Ballantyne EW. Sacrococcygeal tumors: adenocarcinoma of a cystic congenital embryonal remnant. *Arch Pathol (Chic)*. 1932; 14: 1-9.
4. Sakr A, Kim HS, Han YD, Cho MS, Hur H, Min BS, Lee KY, Kim NK. Single-center experience of 24 cases of tailgut cyst. *Ann Coloproctol*. 2019; 35: 268-74.
5. Li W, Li J, Yu K, Zhang K, Li J. Retrorectal adenocarcinoma arising from tailgut cysts: a rare case report. *BMC Surgery* 2019; 19: 180.
6. Valecha J, Ojha SS, Sharma A, Nilkanthe R. Adenocarcinoma in a tailgut cyst: a rare case report. *J Cancer Res Ther*. 2019; 15: 261-3.
7. Şahin S, Kepil N, Batur Ş, Çetin SE. Adenocarcinoma in a tailgut cyst: a rare case report. *Turk Patololoji Derg*. 2020; 36: 169-72.
8. Tziakou P, Myoteri D, Zizi-Sermpetzoglou A, Dellaportas D. Mucinous carcinoma arising in a tailgut cyst. *Virchows Arch*. 2017; 471: 5319-20.
9. Iwata E, Orosz Z, Teh J, Reynolds J, Whitwell D, Tanaka Y, Athanasou NA. Neuroendocrine tumor arising in a tailgut cyst: a rare presacral tumor. *Int J Surg Pathol*. 2019; 27: 336-42.
10. Song DE, Park JK, Hur B, Ro JY. Carcinoid tumor arising in a tailgut cyst of the anorectal junction with distant metastasis: a case report and review of the literature. *Arch Pathol Lab Med*. 2004; 128: 578-80.
11. Mouloupoulos LA, Katvouni E, Kehagias D, Dimopoulos MA, Gouliamos A, Vlahos L. MR imaging of complex tailgut cysts. *Clin Radiol*. 1999; 54: 118-22.
12. Johnson KN, Young-Fadok TM, Carpentieri D, Acosta JM, Notrica DM. Case report: misdiagnosis of tailgut cyst presenting as recurrent perianal fistula with pelvic abscess. *J Pediatr Surg*. 2013; 48: e33-6.
13. Carpelan-Holmström M, Koskenvuo L, Haapamaki C, Renkonen-Sinisalo L, Lepistö A. Clinical management of 52 consecutive retro-rectal tumors treated at a tertiary referral centre. *Colorectal Dis*. 2020; 22: 1279-85.
14. Hove MG, Gil JM, Rodríguez TS, Lormas AF, Casajus JP, Lopez-Farre A, Guijarro JJ. Laparoscopic approach to tailgut cyst (retrorectal cystic hamartoma). *J Minim Access Surg*. 2019; 15: 262-264.
15. Rompen IF, Scheiwiller A, Winiger A, Metzger J, Gass JM. Robotic-Assisted Laparoscopic Resection of Tailgut Cysts. *JLS* 2021; 25: 1-7.
16. Duek SD, Kluger Y, Grunner S, Weinbroum AA, Khoury W. Transanal endoscopic microsurgery for the resection of submucosal and retrorectal tumors. *Surg Laparosc Endosc Percutan Tech*. 2013; 23: 66-8.