Case Report

Surgery and Clinical Practice

Retrorectal Cystic Hamartoma - A Rare Case Report

Seif Nuru[1], Denis Katyali[1], Emmanuel Kajana[1], Lucas Mgimwa[1] and Peter Mbelle[2*]

[1]*Department of Surgery, St. Gaspar Referral and Teaching Hospital, Tanzania.*

[2]*Department of surgery, Dodoma referral hospital, Tanzania.*

[*]Correspondence:

Peter James mbelle, Department of surgery, Dodoma referral hospital, Tanzania.

Received: 04 Jun 2024; **Accepted:** 30 Jul 2024; **Published:** 05 Aug 2024

Citation: Seif Nuru, Denis Katyali, Emmanuel Kajana, et al. Retrorectal Cystic Hamartoma - A Rare Case Report. Surg Clin Prac. 2024; 1(1): 1-4.

ABSTRACT

Retrorectal cystic hamartomas or tailgut cysts are rare congenital lesions which result from the non-regression of embryo remnants of the hindgut. In this case report we present a 4-year old female who was referred to our facility with a history of change in stool habit for 2 years characterized by pain during defecation and passage of flat shaped stool. Abdomino-pelvic CT scan suggested retrorectal cystic hamartoma which was successfully surgically excised through posterior sagittal approach. The final pathological diagnosis was retrorectal cystic hamartoma (tailgut cyst) with no evidence of malignancy.

Keywords

Retrorectal cystic hamartoma, Tailgut cyst, Retrorectal space.

Abbreviations

CT: Computer tomography, MRI: Magnetic Resonance Imaging, DRE: Digital rectal Examination, US: Utrasonography.

Introduction

The retrorectal or presacral space is bounded anteriorly by the rectum, posteriorly by the sacrum, superiorly by the peritoneal reflection, inferiorly by the pelvic diaphragm muscles (levator ani and coccygeus), and laterally by the ureters and iliac arteries and vein. Retrorectal cystic hamartomas or tailgut cysts are rare congenital lesions that develop in the retrorectal or presacral space when embryologic tailgut fails to regress [1]. For most cases, tailgut cysts are found in adult female, but also occur extremely rarely in neonates [2].

They can be symptomatic due to their mass effect, but their clinical presentation is non-specific and often the diagnosis is missed [3]. CT scans and MRI are useful in the diagnosis of these retrorectal masses, and surgical resection is the definitive treatment [4].

Worldwide (up to February 2021) there were about 144 articles published about retrorectal cystic hamartoma, very few studies from Africa [5]. To the best of our knowledge, there is no even a single case of retrorectal cystic hamartoma which has been reported in our country, Tanzania.

Case Presentation

We report a case of a 4-year old female who was referred to our facility with a 2 year history of change in stool habit characterized by pain during defecation and passage of flat shaped stool. This was associated with dull pain in the perianal region and buttocks when sitting. There was no associated history of fever, constipation, per-rectal bleeding, or any urinary complaints and had no history of abdominal pain, distention, weight loss or other constitutional symptoms.

She was first treated at her nearby health facilities with oral medications over an extended period of time after receiving a diagnosis of gluteal abscess and had 2 surgical procedures of incision and drainage but symptoms recurred after 3 to 4 month of treatment.

On physical examination had an incision scar on the right gluteal region (Figure 1). On DRE had normal anal verge and sphincter tone. A mass was palpated on the posterior rectal wall compressing the rectum, which was rubbery, non-tender and not mobile. The margins of the mass were regular with a smooth surface and

the overlying skin was not attached to the mass. Upon removal of the gloved finger there was no active bleed or discharge. Other systemic examinations were unremarkable and laboratory investigations were within normal limits.

A sigmoidoscopy showed a mass pushing the rectum from posterior side, which was compressible allowing the passage of the scope. It was extending from 5cm to 12 cm and beyond it the colon was normal. CT scan of the pelvis and abdomen showed a large cyst (60.1x62.7x64.5mm) without any solid or abnormal enhancing components behind the rectum in presacral space extending into perianal region with no communication to the spine or the rectum (Figure 2A, 2B).

The treatment of choice was complete surgical excision of the lesion by a posterior sagittal approach. Intraoperatively the patient was kept in prone jackknife position then a trans-sacral incision was made separating muscles by layer. Blunt dissection was used to carry the incision down directly in the midline until the presacral fascia was encountered. Finding was a well capsulated presacral cyst pushing the rectum towards the urinary bladder. Though the lesion was attached to the rectal wall, no communication was found with the rectal lumen. While removing the cyst ruptured with thick milky mucoid material oozing out but complete excision of the cyst was achieved and a drainage was kept in the retrorectal space.

The gross specimen consisted of a translucent cystic structure containing thick mucoid fluid (Figure 4). The specimen was sent for histopathological diagnosis which revealed retrorectal cystic hamartoma (tailgut cyst) with no evidence of malignancy. On day 5 post-operation the drainage was removed with < 15 mls of serosanguinous fluid since the day of operation. The patient was discharge after 6 days and her postoperative course was uneventful. The patient returned to our clinic 3 months later for follow up. She was symptom-free with no evidence of recurrent or residual disease.

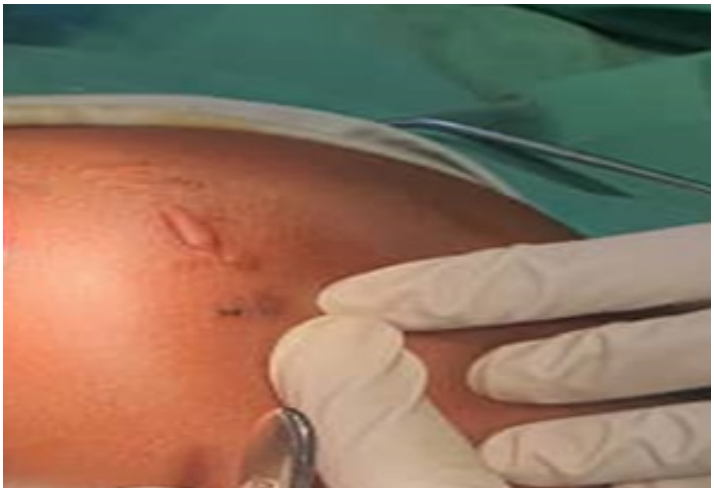

Figure 1: Surgical incisional scar for drainage.

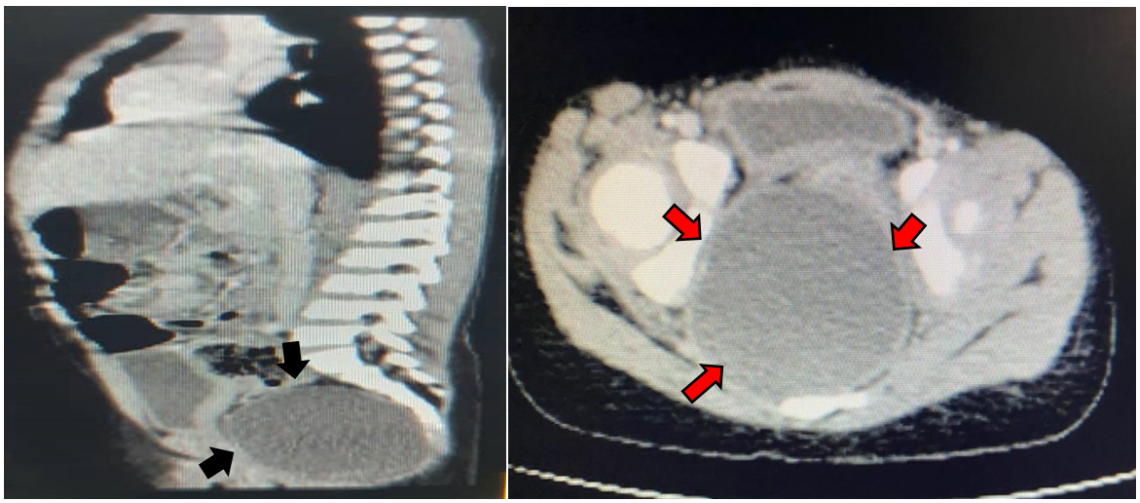

Figure 2B: Axial view of the abdominopelvic CT scan showing cystic lesion (Red arrows).

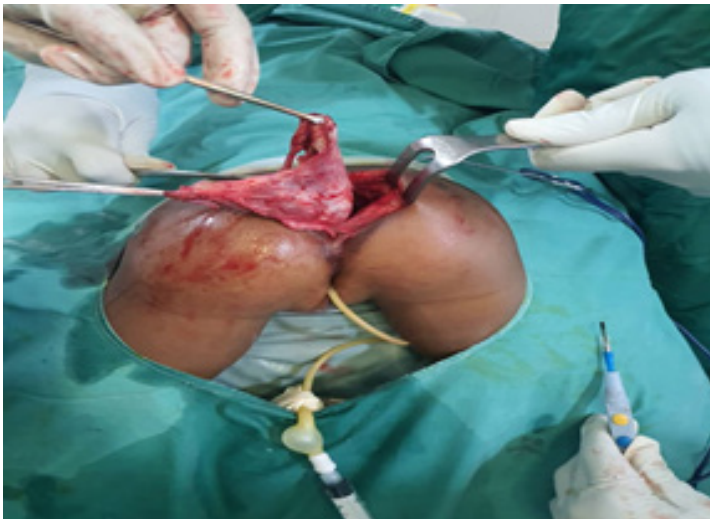

Figure 3: Image of the cyst being excised.

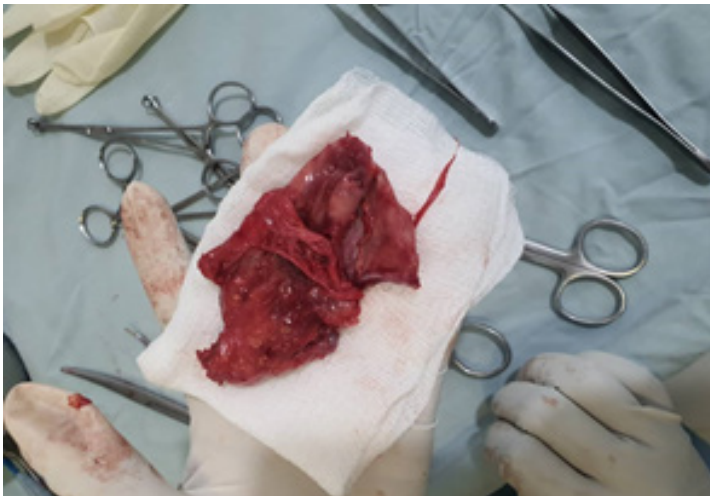

Figure 4: Gross image of the cyst.

Discussion

Retrorectal cystic hamartomas, or tailgut cysts, are rare congenital tumors with an estimated incidence of 1/40,000. They are more commonly seen in adults. Women are more likely (3:1) to have retro rectal cysts than men [1].

Early in its development, the embryo possesses a true tail, which develops maximally the 8-mm stage (35 days -gestational age) and usually completely regresses by the 35-mm stage (56 days-gestational age). The anus is formed cephalad to the tail. Because the primitive gut extends into the tail beyond the point at which the anus develops, it is called the tailgut. When the tailgut fails to regress normally, congenital cysts tend to occur in this region [2,3]. They are often lined with mucin-secreting columnar cells, squamous, transitional, or a combination [6].

Retrorectal cystic hamartomas are frequently misdiagnosed due to the frequent absence or unusual symptoms and their rarity; therefore, the first diagnostic move is simply to include the possibility of such a lesion on the differential diagnosis [7]. They are usually discovered only incidentally, and symptoms often depend on the size and relation to the adjacent structures [8]. Complications from mass effect and infection can be the primary indication of a retrorectal cystic hamartoma [3].

The differential diagnoses of retrorectal cystic hamartomas should include sacrococcygeal teratoma, epidermoid cyst, rectal duplication cyst, anterior meningocele, and inflammatory cysts. The classification of these masses in the retrorectal region is dependent upon the type of epithelium present [9].

Imaging modalities including MRI, CT and USS can help differentiate retrorectal cystic hamartoma from other cystic masses, in terms of the presence of calcification, fat, hemorrhaging and communication with other structures [10]. Retrorectal cystic hamartoma rarely can transform into a tumor such as an adenocarcinoma, carcinoid, neuroendocrine carcinoma, or sarcoma [11]. Therefore, retrorectal cystic hamartoma need surgical excision to prevent (and treat) complications, such as infection, recurrence, and malignant transformation [3] hen a retrorectal

cystic hamartoma is diagnosed complete surgical excision is the treatment of choice. Incisional biopsies are contraindicated because they can lead to fistula formation [12].

Conclusion

Retrorectal cystic hamartomas are rare congenital lesions originating from a remnant of the embryonic postnatal gut tailgut. They often occur in the presacral retrorectal space. Digital rectal exam is an important examination to have a differential diagnosis of retrorectal cystic hamartoma. CT scan and MRI are helpful diagnostic imaging to define the extent of the cystic mass and its relationship to the surrounding structures. Complete surgical excision is the definitive treatment of retrorectal cystic hamartoma.

Declarations

Ethical approval and consents to participate

Written informed consent was obtained from the guardian of the patient

Consent for publication

Consent for publication was obtained from the guardian and St Gaspar Hospital

Acknowledgement

We would like to appreciate the Main the director and surgical team from St. Gaspar hospital who participated full during treatment of this client, may God bless your gifted hands.

References

1. Lorenzo B, Gustavo S, Pereira B, et al. Tailgut Cyst – A Case Report. 2022; 190-192.
2. Hyen J, Yun L, Lee S, et al. Tailgut Cyst in a Neonate: A Case Report. 2016; 315-317.
3. Ozturk H, Dagistan E, Ozturk H. CASE REPORT A rare malformation of the alimentary tract, tailgut cyst : a case report. 2018; LXXXI(December): 2017-2019.
4. Kardoun N, Hadrich Z, Boujelben S, et al. Tailgut cyst : 2 case reports. 2021; 6-10.
5. Mastoraki A, Giannakodimos I, Panagiotou K, et al. Epidemiology, diagnostic approach and therapeutic management of tailgut cysts : a systematic review. 2021; 513863.
6. Li J, Song X, Shi J, et al. Surgical management of tailgut cysts. 2019; 6: 2018-2020.
7. Jang S, Jang K, Song Y, et al. Unusual prerectal location of a tailgut cyst: A case report. 2006; 12: 5081-5083.
8. Letters S. Gastroenterología y Hepatología. 2018; 41:103-105.
9. Gouveia GDC, Okada LY, Paes BP, et al. Tailgut cyst : from differential diagnosis to surgical resection - case report and literature review. 2020; 1-3.
10. Shah N, Edelstein P. Retrorectal Tailgut Cyst : A Case Report. 2022; 14: 7-10.
11. Atiya S, Horn A, Wedel W, et al. Case Report A Rare Case of Ruptured Tailgut Cyst Leading to Carcinomatosis. 2023; 2023.
12. Reports C, Kildu E, Evaldas N. Surgical management of a retro-rectal cystic hamartoma (tailgut cyst) using a trans-rectal approach: a case report and review of the literature. 2014; 11-15.