Retrorectal Cystic Hamartoma a case report and review of the literature

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ABSTRACT

Hamartoma cysts are rare congenital abnormalities in the retrocecal/presacral region. Clinical diagnosis is difficult and delayed and they can present in childhood and adult life with a variety of clinical symptoms and complications. Differential diagnoses include, rectal duplication cysts, cystic teratoma, epidermal cyst, anal gland cyst and anal gland carcinoma. Magnetic resonance imaging has recently become the modality of choice to image these cysts. Although Tailgut cysts rarely undergo malignant transformation, early surgical resection is presently considered the treatment of choice. Here we report the case of a 47-year-old gentleman with a Tailgut cyst, and review of the literature. We believe that the high incidence of complications associated with operations in the presacral region should be weighed against the generally benign course of these lesions, especially with the quality of modern imaging technology.

Keywords: *Hamartoma cyst, retro-rectal cystic hamartoma, surgical excision, Mauritania*

INTRODUCTION

Hamartoma cysts, also known as retrorectal cystic hamartomas or mucin-secreting cysts, are congenital lesions arising from the remnants of the embryological tailgut [1]. They arise mainly in the presacral space, a potential space defined posteriorly by Waldeyer's fascia, anteriorly by the fascia propria of the rectum, laterally by the ureters and the lateral ligaments of the rectum, inferiorly by the elevator ani and coccygeus muscles and superiorly by the peritoneal reflection between the second and third sacral segments [2,3]. Cases have also been described anterior to the rectum, perianally or even in the perirenal region [4–6]. Tailgut cysts can be the source of chronic perirectal and perianal symptoms and are therefore often unrecognized, misdiagnosed and undertreated, leading to the patient having multiple operations before the final diagnosis is made. The aim of this study was to report one surgeon's experience with tail- gut cysts which presented as a mass located in the right gluteal region, limiting dorsal decubitus positions, without skin disorders, not very sensitive and evolving for several months.

CASE PRESENTATION

Patient aged 47 presented to the surgical consultation at the Hamad Ben Khalifa hospital in Boutilimit for the presence of a solid mass located in the right gluteal region limiting dorsal decubitus positions, without skin disorders, painless and progressive growing for several months.

The clinical and biological investigations without notable abnormalities, Computerized tomography (CT) of the abdomen and pelvis revealed the same mass and characterized it as multiseptate (Fig. 1). It was not seen to invade any of the adjacent structures. No other pelvic or abdominal abnormalities were identified. (figure 1), and the search for tumor markers was negative.

The patient underwent surgery via a posterior approach using the KRASKE technique, and the surgical specimen (figure 2) sent to the anatomopathologist concluded a tumor consisted of a pink-tan 8 x 7 x 1.9 cm multi cystic mass (Fig. 2). The thin-walled cysts ranged in size from 0.3 to 3.5 cm in diameter, and a few of them were collapsed. Many cysts contained a thin yellow fluid and were partially lined by thin wrinkled bright tissue reminiscent of skin. Other cysts were lined by a pale smooth surface.

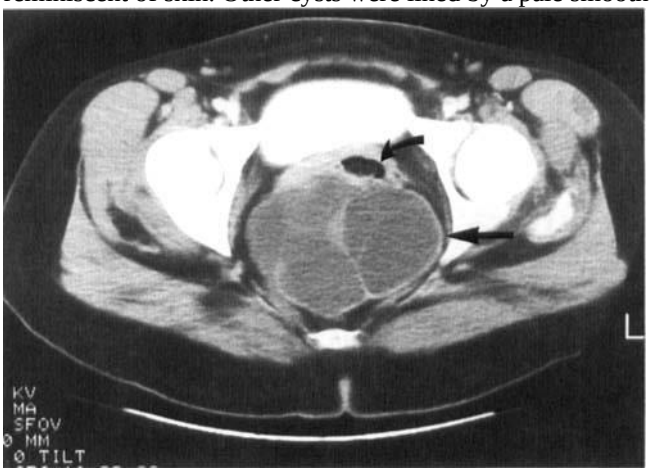

Fig. 1. CT scan of the pelvis demonstrating a large multicyclic mass (straight arrow) seen behind rectum (curved arrow).

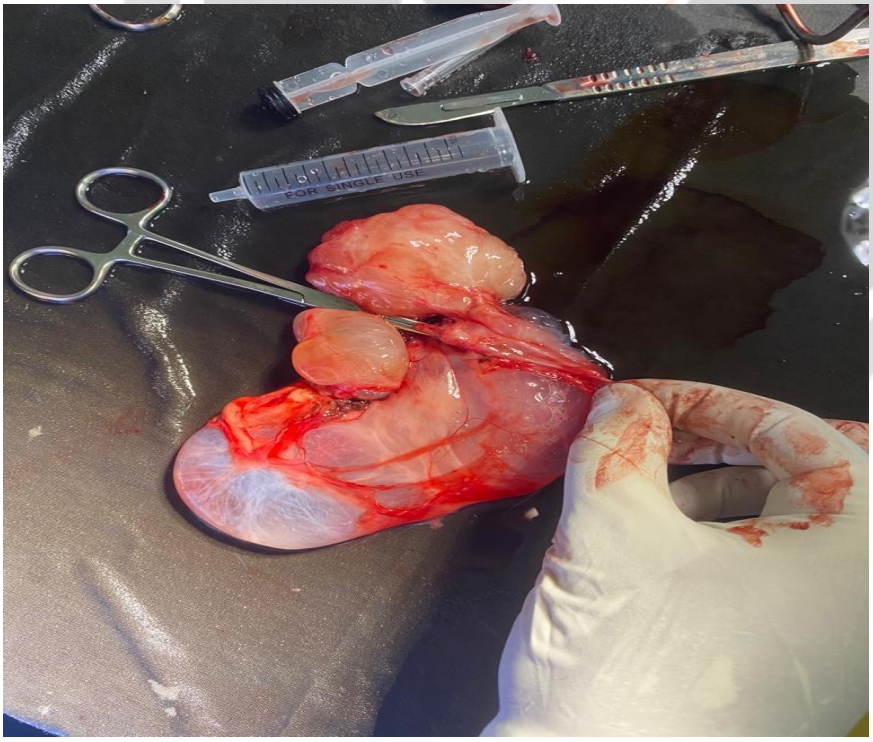

Fig. 2. Resection specimen consisting of a large multiloculated cyst.

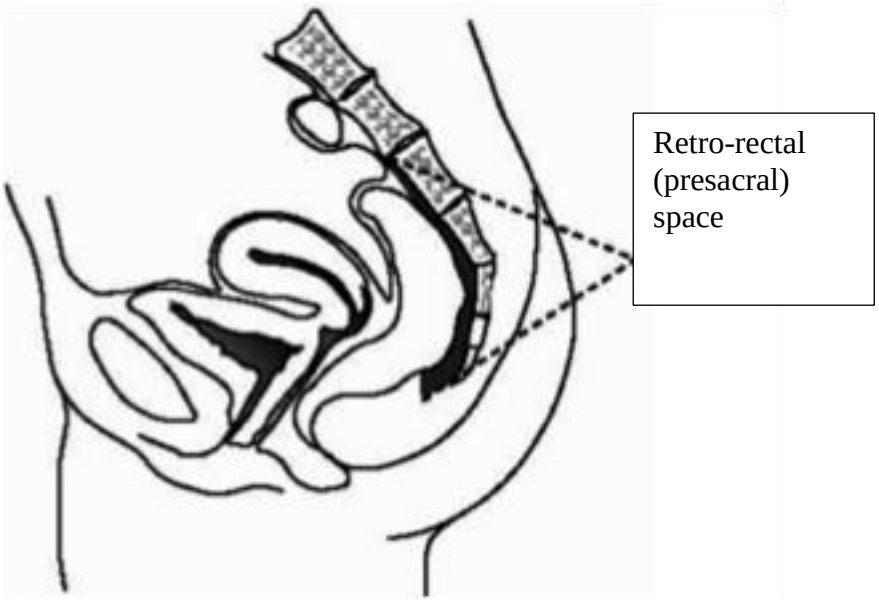

Figure 3 Anatomy of retro sacral or pre- sacral space.

DISCUSSION

The first report of a cystic mass in the retrorectal space was by Middeldorpf, 1885 [7], who described his findings in a one-year-old girl, most likely presenting with a rectal duplication cyst. Tailgut cysts (TGCs) are congenital developmental anomalies in the retrorectal space. The retrorectal (presacral) space is the potential space bounded anteriorly by the rectum, sacrum posteriorly, superiorly by the peritoneal reflection, inferiorly by the levator ani and the coccygeus muscles. The lateral boundary is formed by the ureters and iliac vessels (Fig.3). There has been some controversy about which regions these congenital cysts originate, and two embryological structures, the tail gut and the neurenteric canal, have been suggested as the origins of these cysts [8–16].

TGCs are more commonly seen in females [1,11,13,18] but the reason for this tendency is unclear. In the largest series of TGCs to date, the authors described 53 patients with an average age of 36 years and range of 4 days to 73 years (Hjermstad and Helwig [1]). It is not known why these congenital abnormalities should exhibit delayed presentation. There is no correlation between patient age and lesion size. TGCs are often an incidental finding, but can present symptomatically with rectal fullness, rectal pain, rectal bleeding, painful bowel movements, change in stool calibre, urinary frequency and urinary obstruction. All those symptoms are most likely related to the physical size of the cyst.

There are reports in the literature of TGCs being associated with a postanal dimple [14,17,18]. Post anal dimple (fovea coccygea), which is a short blind pit found in the skin beneath the tip of the coccyx, is regarded as an indicator of an underlying TGC. In one series of 12 cases, a post anal dimple was present in all subjects [19]. This is most likely due to the pull of the filum terminale on the skin during growth and development. There is an association between TGC and sacrococcygeal abnormalities. Presence of a retrorectal cystic abnormality in a mother and daughter has also been reported [14].

Clinical diagnosis is difficult and may be delayed, which could be attributed to unfamiliarity on the part of the clinician or the extraluminal location of the cyst. TGCs are multi cystic and multilocular and do have a muscular coat, which enables differentiation from rectal duplication cysts. Other differential diagnoses include cystic teratoma, epidermal cyst, anal gland cyst and anal gland carcinoma. Endosonography, computed tomography and MR imaging have all been applied to the differential diagnosis. Recently, MR has become the modality of choice to image these cysts, firstly because it is able to image in surgically relevant planes (notably coronally and sagittally), and secondly it is also able to characterize cyst content, depending on the sequences used. For example, T1-weighted sequences will image fat and fat-suppression techniques, such as the STIR sequences employed in our case; it will help eliminate dermoid from the differential diagnosis. Multi-planar imaging is helpful in confirming that the cyst is remote from both the anal canal- excluding anal gland cysts and the spinal canal- excluding meningocele and chordoma. In contrast to duplication cysts, tailgut cysts do not contain a defined muscle layer and the absence of this on MR imaging supports the diagnosis of tailgut cyst [20].

CONCLUSION

Hamartoma cyst of retrorectal space in an elderly man is extremely rare. It can be taken for an abscess and fistula of the buttock. CT scan and MRI are very important examinations, which allow the diagnosis. The basic treatment method is radical resection of the lesion.

CONSENT

A written informed consent was signed by the patient prior to publication of this paper.

CONFLICTS OF INTEREST

None.

AUTHOR CONTRIBUTION

YT and BR have designed, conceptualized the study, and wrote the first draft. BR operated upon the patient. MK, AMI, MJSB contributed to the drafting of the paper. All authors have reviewed and approved the final manuscript.

ABBREVIATION

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