Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Gastroenterology

Pelvic Retroperitoneal Epidermoid Cyst: A Rare Case

T. Adioui[1], S. Rokhsi[1*], H. Igorman[1], F. Nejjari[1], S. Berrag[1], M. Tamzaourte[1]

[1]Department of Gastroenterology, Mohammed V Military Training Hospital, Rabat, Morocco

DOI: 10.36347/sjmcr.2022.v10i12.016 | **Received:** 04.11.2022 | **Accepted:** 10.12.2022 | **Published:** 18.12.2022

***Corresponding author:** S. Rokhsi
Department of Gastroenterology, Mohammed V Military Training Hospital, Rabat, Morocco

Abstract **Case Report**

Epidermoid retrorectal cystic tumors are rare and uncommon lesions that are generally asymptomatic, the appearance of symptoms such as pain or neurological disorders should raise suspicion of degeneration. We report the case of a 35 year old woman who was found to have a renitent, painless lump compressing the posterior wall of the rectum while having a history of painful discomfort while sitting. The diagnosis was confirmed after discovering a well-circumscribed presacral mass on computed tomography (CT) scan and MRI. She underwent a posterioc laparoscopy and the excision of the encapsulated cystic mass was later confirmed to being an epidermoid cystic tumor. Finally, due to the risk of infection and malignant transformation, these tumors must be managed with a multidisciplinary approach to maximize successful treatment.

Keywords: neurological disorders, computed tomography (CT) scan, presacral tumors, developmental cyst.

INTRODUCTION

Retrorectal or presacral tumors are extremely rare and uncommon lesions that present with nonspecific signs and symptoms, which lead to difficult diagnoses. With advances in imaging modalities and increased clinicians' awareness, the diagnosis of a retrorectal tumor has been improving over the years allowing an appropiate surgical approach [1].

We are reporting the case of an epidermoid cystic tumor in which surgical excision was performed.

CASE REPORT

A healthy 35 year old woman with a good general condition presents with history of painful discomfort when sitting for two years, and recent symptoms of terminal constipation.

Rectal inspection found a para-anal swelling and digital rectal examination reveals a renitent, painless lump compressing the posterior wall of the rectum and its inferior side touches the anus. Anuscopy had noted posterior rectal bulge with normal rectal mucosa.

Pelvic CT (Fig. 1) and MRI (Fig. 2) had shown a well-limited homogeneous hypodense cystic mass of the presacral retro-rectal space, pushing the rectum and the bladder forward.

The patient undergone surgery by retroanorectal perineal approach allowing the resection of an encapsulated cystic mass (Fig. 3) which was confirmed to be an epidermoid cyst after anatomopathological analysis. The postoperative course was uneventful, and the patient was discharged on day 7.

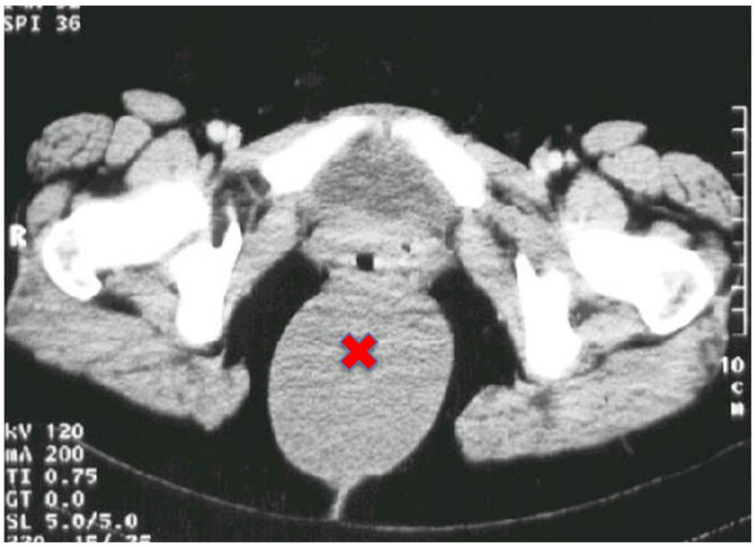

Fig. 1: Contrast-enhanced computed tomography image. A cystic mass (76 × 70 mm) is seen on the left and posterior side of the lower rectum. The wall of the cystic mass is slightly enhanced in the late phase, and the cystic mass is pushing the rectum and bladder forward. Cross: cystic mass

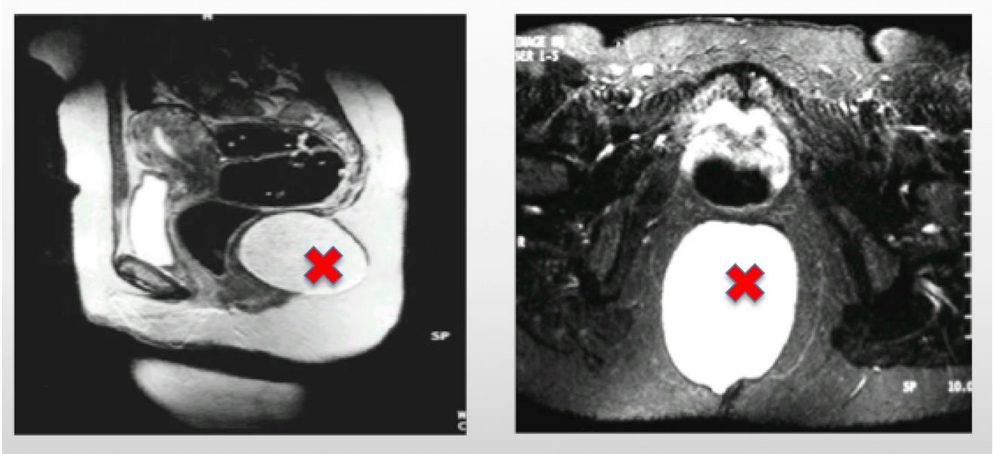

Fig. 2: T1 and T2 weighted magnetic resonance images. Cross: Cystic mass

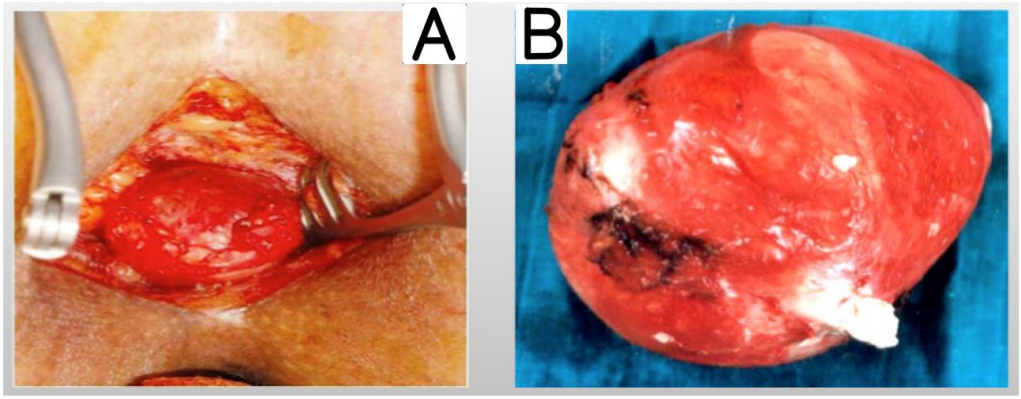

Fig. 3: (A) Intraoperative photograph showing a cystic formation compressing the posterior wall of the rectum. (B) Surgical specimen after surgical resection

DISCUSSION

The term "developmental cyst" signifies a cystic tumor occurring in the presacral space. Developmental cysts include dermoid, epidermoid, and mucus-secreting cysts, and their presence is due to a certain developmental error during the formation of the embryo [2]. The generally accepted definition in describing an epidermoid cyst is as follows: an epidermoid cyst has stratified squamous epithelium with keratohyaline granules, but no skin appendages [3].

Retrorectal tumors are a rare congenital lesions of the presacral space.

The boundaries of this space are the presacral fascia (Waldeyer fascia) posteriorly, fascia propria of the rectum and mesorectum anteriorly, levator ani muscles inferiorly, the peritoneal reflection superiorly, and the iliac vessels and ureters laterally [4].

The majority of lesions are benign, and malignancy is found in 15% of the cases and mostly in children. (Table 1) They are generally asymptomatic with a predominance of females [6] but can also appear with a symptomatology dominated by perineal pain and constipation resulting from the mass effect applied by the tumor which was similar to our patient presentation.

Table 1: Modified classification of retrorectal tumors described by Uhlig and Johnson [6]

	Congenital	Neurogenic	Osseous	Miscellaneous	Inflammatory
Benign	Epideromoid cyst Dermoid cyst Enterogenous cyst Cystic hamartomas Anterior sacral meningocele Teratoma	Neurofibroma Neurilemoma (schwannoma) Ganglioneuroma	Giant-cell tumor Osteoblastoma Aneurysmal bone cyst	Lipoma Fibroma Leiomyoma Hemangioma Endothelioma Desmoid	Perineal or pelvirectal abscess Endometriosis Foreign body granuloma Infectious granulomas Diverticulitis Crohn's disease
Mali-gnant	Chordoma Adrenal rest tumor	Neuroblastoma Ganglio-neuroblastoma Ependymoma Malignant peripheral nerve sheath tumours (Neurofibrosarcoma Malignant schwannoma)	Osteogenic sarcoma Ewing sarcoma Myeloma Chondrosarcoma	Liposarcoma Fibrosarcoma Malignant histiocytoma Leiomyosarcoma Hemangio-pericytoma Metastatic adenocarcinoma	

Retrorectal tumors can frequently present with infection, and patients whose tumors are infected might also experience pain. An infection within the tumor may present as an abscess, a draining sinus, or a fistula tract. Therefore, these tumors may be initially misdiagnosed as fistulas, perirectal abscesses, or pilonidal diseases. Singer *et al.*, reported that patients underwent an average of 4.1 surgical procedures before the correct diagnosis of a retrorectal lesion was made [7].

Most of the symptoms associated with retrorectal tumors can be vague and nonspecific, thus the most important aspect of the diagnostic process is an accurate physical assessment, which facilitates the identification of the appropriate surgical method. . A careful rectal examination is essential to establish the diagnosis in 90% of patients [8]. Mostly, the lesions can be soft, tender and easily compressible which could be unnoticeable if the physician does not maintain a high index of suspicion.

Although small lesions may not be detected by sigmoidoscopy, flexible sigmoidoscopy and anuscopy can show a more or less important bulge of the posterior face of the rectum covered by a normal mucosa and confirm the level of proximal extension of the tumor [9]. These endoscopic examinations will make it possible to search for a possible intrarectal fistulous orifice that is more or less productive.

Glasgow *et al.*, investigated 34 patients treated for retrorectal tumor and reported the sensitivity of proctoscopy to be 53%. This increased to 100% with the use of transrectal ultrasound and provides information on the size, consistency of the mass, and evidence of local invasion [10]; the accuracy of magnetic resonance imaging vs CT for specific histologic tumor type was 28% vs 18%, respectively. Singer *et al.*, reported a physical examination in combination with CT scanning to result in the correct diagnosis of retrorectal cyst in all their patients. They advocated that it is not advisable to avoid resection based solely on noninvasive studies [7].

As technology has evolved, radiographic imaging has become more sophisticated and plain radiographs have been replaced by computed tomography (CT) scans and magnetic resonance imaging (MRI), as they became the imaging modalities of choice in diagnosing retrorectal tumors. Plain x-rays also elucidate bony destruction and/or calcification of soft tissue secondary to chordomas, sarcomas, and locally aggressive tumors such as giant cell tumor, aneurysmal bone cyst, and neurilemoma [11].

Other imaging techniques that may assist in the diagnosis and management of retrorectal tumors include angiogram and venogram, endorectal ultrasound (ERUS), and fistulograms. An angiogram and venogram can be added to MRI (MR angiogram and venogram) to determine vascular anatomy and involvement if there is distortion secondary to mass effect by the tumor. ERUS has been used to evaluate whether the lesion is cystic or solid, and to evaluate rectal involvement [11].

Fine-needle aspiration biopsy of the lesion has been controversial for a long time. Jao *et al.*, reported their findings for patients who had biopsies prior to tumor resection, and they advocated that preoperative biopsies may elicit tumor spread, abscesses, fecal fistulas, or meningitis, so biopsies should not be performed if the tumors are potentially resectable [9]. But in the other hand, the puncture should be performed only if the lesion appears to be unresectable and if a tissue diagnosis is required to guide adjuvant therapy [9].

An accurate pathological diagnosis with a preoperative biopsy is often very difficult and the biopsy may lead to a misdiagnosis. Also, the histological type of retrorectal tumor does not influence the choice of surgical approach [6]. Therefore, a biopsy prior to an operation is usually not recommended.

Once the diagnosis is established, Complete surgical resection is the cornerstone in the management of retrorectal tumors, even in asymptomatic patients, as many lesions may likely to reccur, or might possess the potential for growth or malignancy, or cause complications, such as infection [8].

The approach depends on the location of the cyst. The laparoscopic approach has been shown to be effective only after careful selection of patients and after eliminating the possibility of a malignant origin of the lesions [11]. Trans-anal resection, which is dangerous when the cyst is degenerated, is reserved for cysts of less than 4 cm [12]. Jao *et al.*, emphasized that once a retrorectal cyst is diagnosed, the first choice is an early surgical resection because a retrorectal cyst may become infected, and once such an infection occurs, the postoperative recurrence rate is high [9]. Moreover, the cystic lesion may be malignant. Most previously reported patients with retrorectal epidermal cysts underwent tumor removal via a posterolateral or transanal approach [13]. Our patient also underwent a complete tumor removal via a posterior approach, and the postoperative course was also uneventful.

CONCLUSION

Epidermoid cyst tumors benign congenital tumors yet rare. Diagnosis of these lesions is accomplished via a thorough physical examination in conjunction with radiological imaging, ultrasonography, and ultrasound endoscopy.

Imaging helps in the diagnosis and in choosing the surgical approach according to the location and extent of the tumor. Biopsy is not recommended, as it exposes the risk of infection, rectal fistula and diffusion in case of malignancy. Surgery is systematic with complete removal of the cyst; it confirms the diagnosis, eliminates a malignant lesion and protects against spontaneous complications. The prognosis of benign tumors is good with rare recurrences.

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