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## International Journal of Surgery Case Reports

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## Retrorectal tumors: Case report and review of literature

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## ARTICLE INFO

## Article history:

Received 2 November 2020

Accepted 14 November 2020

Available online 23 November 2020

## Keywords:

Retrorectal tumors

Retrorectal epidermoid cyst

Kraske technique

## ABSTRACT

**BACKGROUND:** Retrorectal tumors are rare diseases and they can be challenging to diagnose and to manage. Usually they have a slow growth and they are asymptomatic. When present, symptoms depends on the dimensions of the tumor and their position. Inside the retrorectal space may develop a wide variety of benign and malignant masses.

**CASE REPORT:** A 70-years-old, obese, female patient was admitted to our hospital referring pelvic and lower-back pain for six months. The retrorectal mass was incidentally detected on imaging, and treated with a modified Kraske procedure. Pathological examination revealed a rare retrorectal epidermoid cyst. We also reviewed the informations present in the scientific literature about the incidence, diagnosis and treatment options of retrorectal tumors.

**CONCLUSIONS:** Even though the rarity and heterogeneity of these tumors, we agree with literature that their surgical management is mandatory in order to achieve a definitive diagnosis and to avoid complications including malignant transformation. The surgical approach should be tailored for each patient and according to tumor's features.

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## 1. Introduction

Retrorectal tumors are rare lesions but they include a wide range of histological differentiation. Their incidence is expected to be 1 in every 40.000 patients with a female predominance and an average age of 30 years [1,2]; however, their occurrence may be lower affecting 1 per 63.000 patients [3]. These tumors are formed in the retrorectal space, also known as presacral space, bounded by the fascia propria of the rectum anteriorly, the Waldeyer fascia (presacral fascia) posteriorly, the peritoneal reflection superiorly, the levator ani muscles inferiorly, and the iliac vessels and ureters laterally [4]. Inside this anatomical space may develop a wide variety of benign and malignant conditions, which most commonly present as masses. These lesions can be congenital, inflammatory, embryological remnants, neurogenic or osseous [5]. We present a successful resection of a retrorectal tumor located in the antero-lateral presacral space in an elderly woman. The tumor was diagnosed as an epidermoid cyst which are rarely found in the retrorectal region. A literature review was also performed to discuss the importance of a proper diagnosis and the relevance of the surgical approach of these rare lesions.

## 2. Case report

This case report has been reported in line with the SCARE criteria [6].

A 71 years-old female with a medical history of pelvic and lower-back pain for about six months was admitted to our hospital. She didn't report history of dysuria or tenesmus.

Abdominal Ultrasound (US) and digital rectal examination were found not diagnostic.

contrast enhanced computed tomography (CECT) of abdomen and pelvis showed an oval-shaped hypodense solid mass of 27 × 38 × 19 mm (CC × AP × LL), close to the rectum. It was described as tailgut cyst (Fig. 1). The patient also underwent a contrast magnetic resonance imaging (MRI) of abdomen and pelvis which revealed a well-defined lesion in the presacral space with doubt communication with the rectum which was displaced laterally. (Fig. 2).

Pancolonoscopy showed diverticulosis and excluded the intestinal origin of the tumor, while rectal endoscopic ultrasound (EUS) showed a 3 cm cystic-lesion placed behind the rectum, without communication with intestinal wall. In accord with literature a CT-guided fine needle aspiration wasn't performed in order to avoid the risk of tumor seeding [7]. The surgical procedure was performed also to provide a definitive diagnosis. The tumor was below the coccygeal level, so we opted for a posterior approach. The patient was placed in prone position (jackknife-position) and operated under general anesthesia. The gluteal region were pulled laterally with adhesive drapes and pre-operative incisional planning was performed

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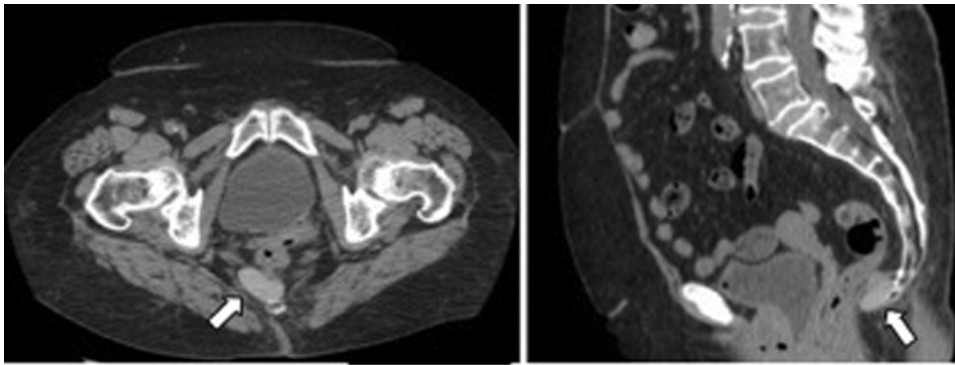


Fig. 1. CECT shows that the retrorectal tumor is located under the third sacral vertebral body.

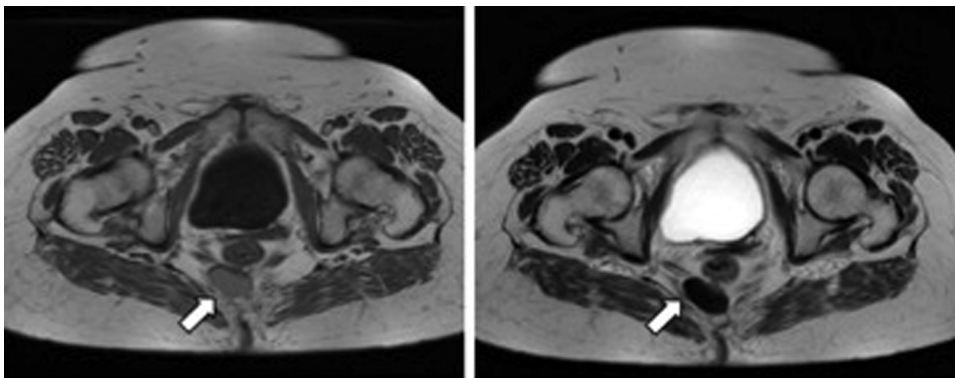


Fig. 2. Contrast-MRI: white arrows indicate the enhancing cystic mass.

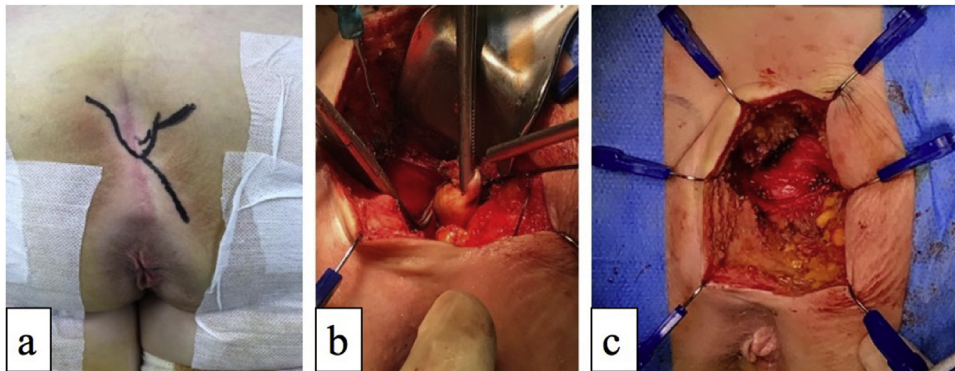


Fig. 3. Images of posterior approach show: a) the gluteal region pulled laterally with adhesive draps and pre-operative incisional planning; b) the surgical exploration of the presacral space confirming the presence of the mass located anterior to S4; c) view of the surgical field after the excision.

(Fig. 3a). Afterwards a right longitudinal incision was made above the sacrococcygeal midline and the exploration of the presacral space confirmed the presence of the mass (Fig. 3b). It was located anterior to S4 and completely removed (Fig. 3c). The communication between the mass and the anal canal or the rectal wall was excluded by the injection of hydrogen peroxide and methylene blue and through a frequent intraoperative digital rectal examination.

Surgical resection specimen consisted of an irregular ovoidal 3 cm mass with liquid content and histologically the lesion was defined as epidermoid cyst. Post-operative days were uneventful and the drain was removed in second post-operative day. The patient was discharged on 4th day after surgery. A digital rectal examination and a CT scan of abdomen and pelvis performed after one year from surgery have excluded a local recurrence and at the moment the patient doesn't refer symptoms.

### 3. Discussion

This paper is an updated review of diagnostic and therapeutic approach to retrorectal tumors. A PubMed search using the keywords “retrorectal tumors”, “presacral tumors”, and “retrorectal epidermoid cyst” was utilized to identify the latest reported cases and review articles of this kind of tumors. The research was made without any language or publication restrictions. Retrorectal tumors include rare lesions growing into the retrorectal or presacral space which is defined as the space between the mesorectum and the posterior pelvic wall [8]. More precisely this space is located between the fascia propria of the rectum and the presacral fascia [9,10]. This anatomical accuracy also comes from recent understanding of the pelvic anatomy in oncological resection [11]. The heterogeneous range of retrorectal tumors, both benign and malig-

**Table 1**  
Classification of presacral tumors using the Uhlig and Johnson system [5].

Types of presacral tumors
<b>Congenital:</b> developmental (dermoid, epidermoid, hamartomas or tailgut cysts), teratomas, chordomas, rectal duplication cysts.
<b>Inflammatory:</b> abscesses
<b>Neurogenic:</b> schwannoma, ganglioneuroma and neurofibroma.
<b>Osseous:</b> Ewing sarcoma and chondrosarcoma.
<b>Miscellaneous:</b> b-cell NH lymphoma, neuroendocrine carcinoma, gastrointestinal stromal tumor, smooth muscle tumor, pecoma (perivascular epithelioid tumor), fibrous histiosarcoma, fibrosarcoma, myeloliposarcoma, metastasis from Squamous cell carcinoma, myelolipoma, intraductal papillary mucinous neoplasm and hemangiopericytoma.

nant conditions, could be explained by their development from different embryological remnants in the presacral space [12].

During the embryological development, the different tissues of the retrorectal space (neurectodermal and endodermal) undergo remodelling process. This can lead to the formation of masses derived from these vestigial tissues [13]. The most frequent masses are congenital and benign, their impact vary from 55% to 81% in different reported series [1]; they are epidermoid cyst, dermoid cyst, rectal duplication cyst, retro-rectal hamartoma (tailgut cysts) and teratoma [14]. The tailgut cyst represent the most common retrorectal benign tumor, ranging from 8 to 62%, followed by dermoid and epidermoid cysts detectable from 3 to 22% of cases [3,14,15] generally in women during the reproductive period [16]. The histological difference between dermoid and epidermoid cysts is the presence of cutaneous adnexal structures in the first one [17]; they could be post-traumatic lesions or result from closure defects of the ectodermal tube with inclusions of skin [18]. The chordoma is the most common malignant tumor, present in almost one third of cases [19]. A complete list of presacral tumors using the Uhlig and Johnson classification system is shown in Table 1, although the retrorectal cysts were originally classified by Lovelady and Dockerty in 1949 [20]. The presence of a lesion in the presacral space may lead to a wide variety of clinical pictures. Most of these lesions are benign and asymptomatic incidental finding, and therefore they may frequently be clinically unrecognized, diagnosed late and undertreated [13]. The infection of a presacral cyst is observed in one-third of patients. It could be misdiagnosed as ano-rectal abscess, specifically if it form a fistula with the intestinal walls [21]. Sometimes, according to their size and location they may cause compressive symptoms including urological, neurological, and defecation difficulties. Acute urinary retention and constipation have been reported with large tailgut cysts [22]. In these conditions it is important to suspect the presence of a rectorectal mass in order to provide a successful diagnosis and treatment. A digital rectal examination or a US, performed in symptomatic patients, are useful for mass detection. For small lesions, EUS allows visualization of the extension of the tumor, infiltration of adjacent organs, the presence of invasion of the rectal muscularis and allows differentiation between solid or liquid lesions, unilocular or multicystic [23]. Pre-operative right planning is mandatory to select the appropriate surgical approach in order to obtain a road map for optimal resection. CT and MRI are the best pre-operative diagnostic modalities [24]. Although CT is often the first line diagnostic study, it cannot always distinguish the benign or malignant nature of the mass [25]. Therefore, MRI is preferable, due to its major diagnostic accuracy; it is more effectiveness in mass characterization compared to CT [26] and their sensitivity and specificity for malignant disease were 81% and 83%, respectively [27]. The limitations of imaging in order to discriminate between benign and malignant disease, specifically for solid tumors, highlights the need for histological diagnosis [28]. For this reason the definitive diagnosis occurs

after surgical excision which allows histological and immunohistological examinations of the specimen [29]. The percutaneous needle biopsy of a presacral tumor is not recommended due to the risk of tumor seeding, especially for cysts and suspected malignant presacral tumors [7]. It should be considered only in unresectable cases or in local, advanced-stage tumors [30] and it should never be performed in patients with epidermoid cysts because it can lead to abscess, fecal fistula, or meningitis [3]. The mass excision is mandatory in order to prevent haemorrhage, infection, compression of adjacent organs, chronic pain, dystocia during delivery, malignant transformation [31] and because it's not possible to characterize the mass by imaging alone. This latter risk varies from 1 to 12%, but can be higher if the retrorectal cyst contains a solid component [21]. The probability that malignant tumors arise from epidermoid cysts is extremely rare, only 0.011%–2.2% [32]. About half of presacral masses in adults are malignant or have areas of malignant change within them [33]. The surgical approach can be done by a perineal, a parasacroccygeal (Kraske procedure), an abdominal or a combined abdomino-perineal approach [13] and it is also related to the tumor localization, having as anatomical landmark the third sacral vertebrae (coccygeal level) [34,35]. In general an abdominal or an abdomino-perineal approach is preferred when the mass is located above this landmark [36]. Most of the tumors were below the coccygeal level, so the posterior approach was usually used [37] and most frequently described [18,38]. In recent times a new surgical approach using transanal minimally invasive surgery (TAMIS) was described for small low-lying lesions [39]; it shows comparable outcomes with the other approaches with the advantage of being less invasive [40,41]. However, enter to the retrorectal region through the rectum means a greater risk of pelvic infection [42]. In a systematic review of 482 papers led by Nicoll et al. the commonest recorded method of surgical excision was by Kraske or a similar approach [11]. The resection should respect the oncological criteria in order to avoid the rupture of the lesions and the dissemination of the contents, which can lead to local recurrences of both benign and malignant tumors. It is necessary to respect adequate margins during surgery if the preoperative diagnosis indicate a malignant tumor [30]. The post-operative complications vary according to the surgical approach [43,44] but it is significantly influenced by the histological type of tumors [31]. The overall rate of post-operative complications was 25%, this includes bleeding, surgical site infection, rectal injury, neurological complications, and urinary incontinence [23,45].

#### 4. Conclusions

We agree with literature that the surgical management of retrorectal tumor is mandatory and it is recommended as firsts line-therapy even for asymptomatic benign mass. They are rare entities in clinical practice but are encountered by general surgeons during their careers. The surgical approach should be tailored for each patient and according to tumor's features.

#### Conflicts of interest

We have no conflicts of interest to disclose.

#### Funding

The work was not supported by any fund/grant.

#### Ethical approval

This study is exempt from ethnical approval in our institution.



## Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Author contribution

All authors contributed to the idea for the article. Material preparation and literature search were performed by [Giorgio La Greca], and [Pietro Conti]. The first draft of the manuscript was written by [Giorgio La Greca] and [Pietro Conti] and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

## Registration of research studies

N/A.

## Guarantor

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## Provenance and peer review

Not commissioned, externally peer-reviewed.

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