

Retrorectal tumor: Mature teratoma clinical case and review of the literature

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ABSTRACT

Introduction: Retrorectal tumors constitute a rare and heterogeneous group of neoplasms, which are characterized by being located in the so-called retrorectal space. In most cases these are benign lesions. The main imaging test for its characterization and planning its therapy is magnetic resonance imaging. Surgery is generally the treatment of choice, often without preoperative anatomopathological studies, given the potential risk of malignancy of these tumors and the appearance of compressive symptoms during the course.

Clinical case: The clinical case of a 34-year-old patient is reported, who consulted for a history of pain in the dorso-lumbar region of months of evolution, accompanied by rectal pain. Magnetic resonance imaging of the abdomen and pelvis was requested, which revealed a retrorectal tumor with imaging characteristics suggestive of teratoma. The resolution was a surgical procedure through a perineal approach, with the diagnosis being confirmed by deferred pathological anatomy.

Conclusion: Retrorectal tumors are rare lesions in adults that represent a diagnostic and therapeutic challenge. The surgical approach depends above all on the tumor size and the relationship with the S3 sacral vertebra. In the case presented, a mature retrorectal teratoma, the perineal approach is a valid therapeutic option that allowed correct exposure and tumor resection.

Keywords: teratoma, retrorectal tumor, posterior approach

INTRODUCTION

Retrorectal tumors constitute a heterogeneous group of neoplasms, which are characterized by being located in the so-called retrorectal or presacral space. Anatomically this is included between the sacrum behind, the rectum in front; above by the peritoneal reflection, below by the levator ani and coccygeus muscles, and by the retrosacral fascia or Waldeyer's fascia. Laterally bordered by the lateral rectal ligaments, the ureters and the iliac vessels [1, 2].

They are rare injuries that even predominate in the pediatric age, for this reason, the evaluation of these patients by the adult surgeon is unusual [3]. The true incidence in the general population is difficult to determine, with an estimate of 1 in every 40,000 hospital admissions [4, 5].

These are tumors with a wide range of differential diagnoses and of diverse histological origin since this region during embryonic development is the meeting point of the nervous, skeletal and digestive systems [3, 6]. Most cases are benign lesions, 70% although in 20% to 50% of cases can be malignant [7, 8].

According to their etiology, they can be divided into congenital, inflammatory, neurogenic, bone and miscellaneous, the most frequent being congenital with 60% of the total and within these, the dermoid cyst and the Sacrococcygeal Teratoma. From a histopathological point of view, they are divided into congenital: benign or malignant, and acquired: benign or malignant [9, 10].

Clinically, the majority are asymptomatic lesions and are discovered incidentally by a routine rectal examination or by other pathology. If symptoms occur, they are due to the compressive effect of the tumor on adjacent structures, especially nerves and the rectum, which may cause pain with defecation, straining and tenesmus. On some occasions, the involvement of pelvic nerves can cause sexual impotence, or urinary and digestive incontinence. Although complications are not common, they occur secondary to abscessation or rupture of the tumor, which can lead to persistent or recurrent abscesses and anal fistulas [11].

In short, retrorectal tumors are characterized by: their scarce and non-specific symptoms that constitute a diagnostic challenge, and their low prevalence and heterogeneity make this pathology a therapeutic challenge.

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LITERATURE REVIEW

Aim

To present the clinical case of a patient with a retrorectal tumor recently treated at the Hospital de Clinicas de Montevideo.

Based on the clinical case presented, discuss diagnostic guidelines and therapeutic tactics.

Clinical case

Female patient, 34 years old, with a history of kidney stones. No surgical history. No family history of oncology.

Consultation due to dorsolumbar pain of 8 months duration, which intensifies especially at night and is associated with occasional rectal pain. There is no alternating constipation, diarrhea, digestive bleeding, or general repercussions.

On physical examination, good general condition, normal

colored skin and mucous membranes. Anus-perineal inspection without injuries. On rectal examination, normotonic sphincter, with bulging on the posterior surface of the rectum that begins approximately 3 cm-4 cm from the anal margin without palpating the upper limit, with a firm elastic consistency, painless, without clinical mucosal involvement.

In order to advance the diagnosis, a computed tomography of the abdomen and pelvis (CT) is performed, which shows a tumor measuring 48 mm × 52 mm of homogeneous density, with a liquid-fat level inside, which displaces the rectum anteriorly, with fatty plane of separation. Video Colonoscopy (VCC) showed healthy mucosa without evidence of lesions.

The diagnostic evaluation is complemented with magnetic resonance imaging of the abdomen and pelvis (MRI), which presents a well-defined retrorectal tumor measuring 70 mm × 45 mm × 40 mm (L, T and AP respectively), hyper intense on T2 with fluid-fat level in non-declining sector, without solid sectors. It moves to the rectum anteriorly, with a fatty plane of separation. Figures 1 and 2.



Fig. 1. MRI pelvis cross section in T2. At the pelvis level, a retrorectal presacral tumor is observed slightly lateralized to the right (arrow). The presence of fatty separation planes and the absence of lymph nodes stand out

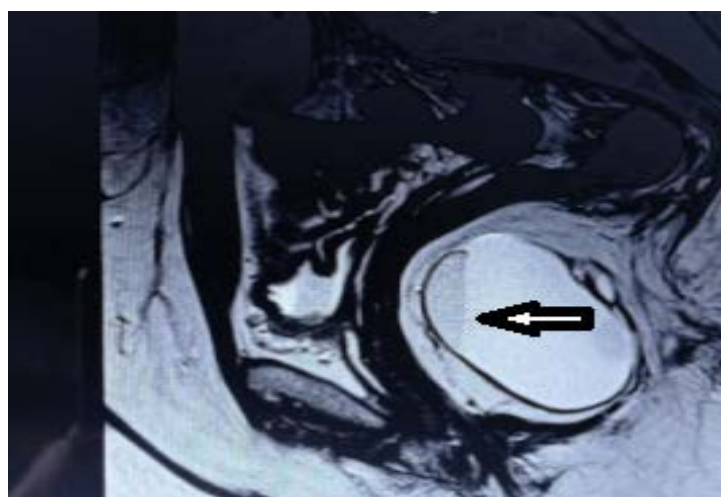


Fig. 2. MRI pelvis sagittal section in T2. In this section, you can see the rectum displaced forward and the proximal extension to the first sacral vertebra. The arrow indicates the presence of fatty liquid level

The clinical case is discussed in an interdisciplinary meeting and a surgical resolution is defined, through a posterior perineal approach, placing the patient in a prone position, Kraske or razor position. A longitudinal incision was made centered on the lesion, the subcutaneous plane was dissected, and the lumbosacral fascia

was exposed. In this way, the retrorectal space was approached; subsequently, careful dissection of the tumor was carried out, trying not to injure the rectum Figure 3 and 4. The specimen is sent for delayed anatomopathological study Figure 5.



Fig. 3. Posterior perineal approach, patient in prone, Kraske or razor position, longitudinal incision

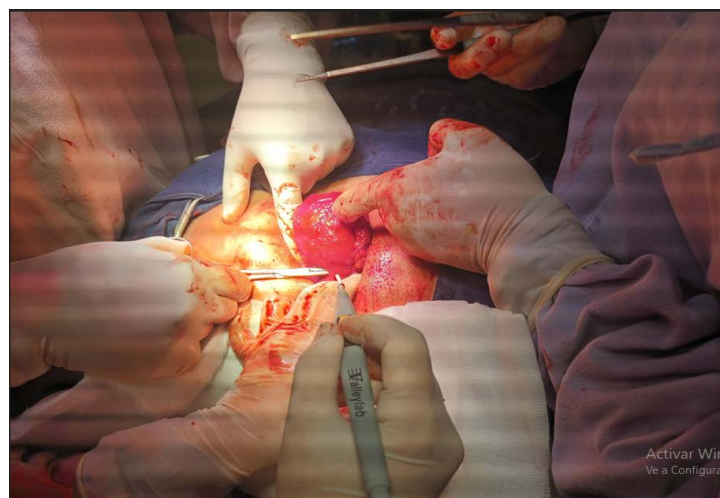


Fig. 4. Complete tumor resection. At this point, the performance of a delicate dissection is highlighted to avoid injury to the rectum

The patient presented a good postoperative evolution, with good pain management under regulated analgesia, and was discharged 48 hours after surgery. The definitive pathological anatomy confirms the presence of a mature teratoma. With a piece measuring 76 mm × 70 mm × 55 mm, weighing 113 grams, with an irregular surface, little adipose tissue and cystic structure, scaly coating, skin annexes and sectors with enteric coating and smooth muscle.



Fig. 5. Piece sent for definitive anatomical pathological study

The patient is currently undergoing clinical and imaging follow-up (12 months), with no clinical or imaging evidence of loco-regional recurrences.

DISCUSSION

Teratomas are tumors that are made up of the 3 embryological layers and are classified according to the tissue they contain; 1- mature: when it contains well-differentiated tissue of a benign course and 2-immature: when it is composed of immature tissue and embryonic structures of a more aggressive nature [12]. When they develop outside the ovaries or testes, they are called extragonadal teratomas. Within the abdomen and pelvis, the most common location is the retrorectal region. They usually arise in places close to the midline and their frequency varies depending on age, being more common in childhood; in adults, they occur more frequently in middle-aged women [13, 14].

They can be solid, cystic or solid-cystic. Although these are benign lesions, they present no less risk (10%) of malignancy, which in some series reaches up to 40%. Mature cystic teratomas are also called dermoid cysts since they characteristically present structures related to the skin [15].

Retrorectal teratomas, like other tumors located in this topography, are a true diagnostic and therapeutic challenge for the general surgeon who must evaluate these patients. Beyond the definitive nature of the anatomopathological study, the diagnostic and therapeutic strategic assessment is broadly common in all retrorectal masses. The main diagnostic studies and therapeutic strategies in the approach to retrorectal tumors are presented below.

The symptoms of these tumors are nonspecific and depend more on the tumor size than on the nature of the lesion. Careful rectal examination is one of the most important pillars to establish the diagnosis of retrorectal mass. Despite this, trained surgeons are required, as well as maintaining a high level of suspicion since these injuries can go unnoticed [5].

The first-line studies for diagnosis are CT and MRI of the abdomen and pelvis, with great sensitivity and specificity in the morphological characterization of the lesion [16].

CT of the abdomen and pelvis is usually the first study to be performed in non-specific abdominal pain. It establishes the characteristics of the tumor (solid, cystic or mixed), anatomical relationships, especially bone, possible complications, and allows us to presume etiology and plan therapy. MRI, for its part, can better evaluate the involvement of soft tissues and adjacent structures. It allows correct planning of the extent of surgery (local or en bloc resection) and the approach (abdominal or perineal) [17]. The ability of MRI to correctly delineate the soft, vascular and nervous planes, define tumors of neural and bone origin, and determining the relationships or infiltration of the mass with the rest of the structures, make MRI the modality of choice for the study of retrorectal lesions [18].

Based on these considerations, the role of the imaging specialist is fundamental in identifying specific characteristics that allow limiting differential diagnoses, as well as describing relevant findings that influence the therapeutic strategy.

Video colonoscopy is an important study to evaluate the rectal mucosa and exclude primary rectal cancer. Transrectal ultrasonography may be useful in the evaluation of local invasion [11].

Preoperative biopsy continues to be controversial; although it allows a more precise diagnostic approach regarding the nature of the lesion, it is not without risks. Among the disadvantages is the possibility of an erroneous diagnosis in 44% of cases, the risk of infection and tumor seeding [9, 10]. However, in solid, heterogeneous lesions, especially with imaging suspicion of malignancy, preoperative biopsy can be beneficial in the therapeutic strategy since there are some types of tumors that improve survival with neoadjuvant therapy, such as Ewing and osteogenic sarcomas [19, 20]. If a biopsy is performed, a perineal or presacral route by TC 18 should guide it.

Regarding treatment, retrorectal tumors require surgery for two fundamental reasons: the risk of malignancy and the possibility of developing symptoms. Surgery allows, even in asymptomatic patients, to confirm the diagnosis, avoid complications and eliminate the risk of malignancy. Only in selected cases of cystic lesions, but especially due to contraindications for surgery, can MRI follow-up be chosen [21].

Treatment consists of complete resection of the lesion in benign tumors and en bloc resection with compromised structures in malignant lesions. For this, a fundamental point is the correct exposure that allows the complete removal of the tumor. The approach to consider is perineal, abdominal and combined. To define the approach, it is necessary to take into account the topography of the tumor. If the equator of the tumor is below the 3rd sacral vertebra or it is palpable on digital rectal examination, a perineal approach can be performed. The rest require an abdominal approach (laparoscopic or open) or a combined approach (abdominal and perineal) [22]. The Kraske or jackknife position is a variant of the prone position that has shown less recurrence and complications than the other forms of approach [23].

Regardless of the approach, we use, as in any surgical procedure, adequate exposure and complete resection of the tumor will be key, which has a direct relationship with the risk of tumor recurrence, knowing that if a complete resection is achieved, in a tumor that is benign, the risk of recurrence is almost zero.

The long-term prognosis of these tumors depends on the histology of the lesion and the quality of the resection [18].

CONCLUSION

Retrorectal tumors are rare lesions in adults that represent a diagnostic and therapeutic challenge. Regardless of their definitive nature, they follow similar study and management algorithms; only in the case of imaging, suspicion of malignancy is a prior anatomopathological study required. The surgical approach depends above all on the tumor size and the relationship with the S3 sacral vertebra.

In the case presented of a retrorectal mature teratoma, the perineal approach is a valid therapeutic option since it is a lesion below the third sacral vertebra. Complete resection of the lesion is essential to avoid loco regional recurrences.

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