Retrorectal Hamartoma, a Rare Congenital Tumor. Case Report

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INTRODUCTION

Retrorectal hamartoma is a rare congenital tumor that is benign in more than 85% of cases.[1] The condition is often asymptomatic or may present with nonspecific symptoms resulting from compression of adjacent organs (e. g. constipation caused by rectal compression), infection, or pain. It is most frequently observed in middle-aged women. A definitive diagnosis requires an anatomic pathologic examination, and the recommended treatment is a complete surgical resection of the lesion. The prognosis is generally favorable.
The aim of this paper is to present a clinical case of a rare, and therefore, difficult-to-diagnose tumor. We will highlight the relevant clinical, diagnostic, and therapeutic aspects

CASE

The patient is a 65-year-old woman with no significant medical history who presented to the emergency department with severe pain in the sacrococcygeal region that has persisted for three months. She is experiencing difficulty sitting and has noticed a tumor at the level of the coccyx. The patient denies any changes in her bowel habits. During the physical examination, a painful, firm mass was felt in the right para-coccygeal area. A digital rectal examination revealed an extra-rectal, posterolateral elastic mass, with normal rectal mucosa. Laboratory tests were all within normal limits.
Abdominal and pelvic magnetic resonance imaging showed a 6 cm multilobulated cystic mass with homogeneous contents located in the precoccygeal retrorectal space, suggestive of cystic hamartoma (Fig. 1).
It was decided not to perform an endoscopic examination as the lesion was extra-rectal and had a clear cleavage plane with the rectum.
A transcoccygeal approach was performed using the Kraske technique in the jackknife position. A transverse incision was made at the level of the sacrococcygeal junction with resection of the coccyx and sacrococcygeal ligament. The lower retro-rectal space was then approached, the tumor was separated from the posterior aspect of the rectum, avoiding its opening, and was completely excised. (Fig. 2).
The pathology report revealed a cystic hamartoma or tailgut cyst with no evidence of malignancy. (Fig. 3).

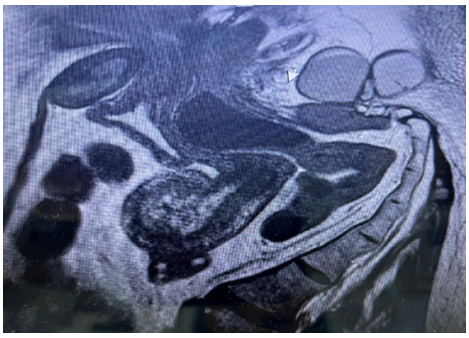

Figure 1. *Sagittal T2-weighted magnetic resonance imaging shows a well-defined multilobulated cystic mass of approximately 6 cm in diameter with homogeneous contents in the precoccygeal retrorectal space. There is no continuity with the rectum and no agenesis or discontinuity of the anterior surface of the coccygeal vertebrae. Based on these findings, a presumptive diagnosis of cystic hamartoma is made.*

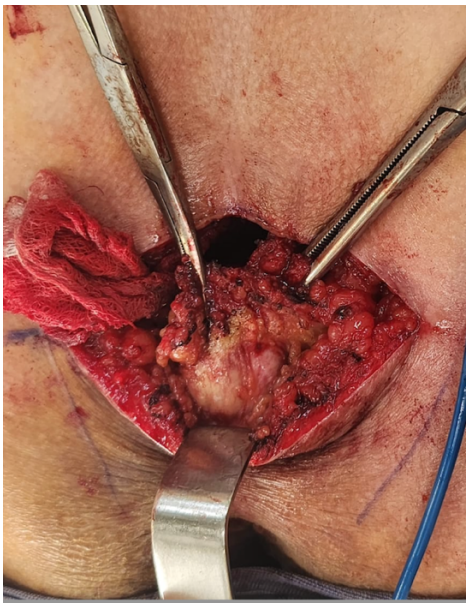

Figure 2. *Excision of the cystic lesion using the Kraske approach, through a transverse incision at the level of the sacrococcygeal junction.*

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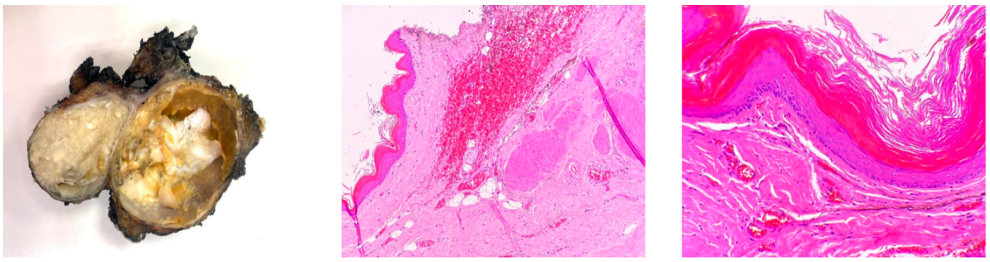

Figure 3. *Excised specimen. Multicystic lesion with a grayish content. Histopathology is consistent with a benign tailgut cyst.*

DISCUSSION

Retrorectal tumors are a rare condition. As its name suggests, the retrorectal space is bordered anteriorly by the rectum and posteriorly by the sacrum. The majority of these lesions are benign and 15% are malignant.[1] These tumors may originate in nerves, bones, or be metastases of another primary tumor. They may also be congenital, such as dermoid cysts, epidermoid cysts, rectal duplication cysts, or cystic hamartomas, each with its particular histological characteristics. Dermoid and epidermoid cysts are lined with stratified squamous epithelium. Dermoids also have hair follicles and sweat glands. Rectal duplication cysts are lined with typical gastrointestinal epithelium and two layers of smooth muscle and nerve plexuses.[4] Cystic hamartoma, arising from the aberrant remnant of the incompletely involuted hindgut, contains several types of epithelium, such as mucus-secreting glandular, squamous, and transitional epithelium, and, unlike duplication cysts, the smooth muscle fibers are not organized in layers and do not contain nerves.[2,4]
Cystic hamartomas are more common in middle-aged women and rarely become malignant. They are difficult to diagnose because most are asymptomatic. When infected, they cause pain, local inflammation, and fistulas; constipation may be the only symptom when the rectum is compressed.[2] Digital rectal examination reveals an extramucosal mass or, as in our patient, may present as a tumor in the coccygeal region causing severe pain when sitting.
Imaging studies can also be a diagnostic tool in the absence of clinical findings. On the other hand, once the diagnosis is made or suspected, CT and especially MRI of the pelvis are the studies of choice for locoregional assessment of size, location, and proximal limits to plan the surgical strategy and to evaluate possible malignant features, such as whether the cyst has a solid component. Cystic hamartoma is characterized on MRI by hypointensity on T1 images and hyperintensity on T2 images. A wall thickening and intracystic vegetations suggest malignancy.[3]
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Treatment consists of complete resection of the lesion to eliminate symptoms and prevent recurrence and possible malignant transformation.2 The approach may vary depending on the extent and location of the lesion. A preoperative pathological diagnosis is not required, except in inoperable or unresectable patients in whom biopsy becomes relevant.
In our patient, a Kraske approach was performed, which consists of placing the patient in a jackknife position, allowing direct access to the retrorectal space. This is the best approach for lesions below S3 that do not extend into the pelvic viscera. For those above S3, the abdominal approach is recommended.[3] A resection of the coccyx may or may not be necessary.
Benign lesions have a good prognosis, but malignant lesions have a high recurrence rate of 10-15%. To reduce this rate, R0 resection, as in our case, is essential.

CONCLUSION

Los tumores retrorrectales son una entidad muy poco frecuente, de caracteristicas benignas en su amplia mayoría. El diagnóstico es incidental o por manifestaciones de las complicaciones de la lesión. Los estudios de imágenes, sobre todo la resonancia magnética son de fundamental importancia para la valoración preoperatoria, siendo la cirugía con escisión total, el tratamiento curativo. La histologia de la pieza confirmará el diagnóstico.

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