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Case Report

Presacral tumor, experience of two cases in Mexico and review of the literature

Mauricio Gutiérrez A.[1], Daniel I. Camacho M.[2], Jorge Leal H.[1], Kevin J. Fuentes C.[1]*

[1]Department of General Surgery, Hospital, Médica Sur, Mexico City, Mexico
[2]Department of Colorectal Surgery, Hospital, Médica Sur, Mexico City, Mexico

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***Correspondence:**
Dr. Kevin J. Fuentes C.,
E-mail: kjfuentescalvo@gmail.com

ABSTRACT

Presacral or retrorectal tumors are rare, they can present different histologic strains and they can also be benign or malignant. These tumors represent a challenge for the surgeon due to their location in the presacral space and due to the low incidence and written information. Various surgical approaches have been described. We present the case of two patients who were diagnosed with presacral tumors (hamartoma and schwannoma) and who underwent a posterior approach for the removal of the tumors. There are several approaches for the treatment of presacral tumors, but in our experience, the posterior approach has shown adequate results without recurrence in the medium-term follow-up. The purpose of the manuscript is to document our experience in the management of two patients.

Keywords: Schwannoma, Hamartoma, Surgery, Pathology, Technique

INTRODUCTION

Presacral tumors also known as retrorectal tumors are a heterogeneous group of diseases that originate in the space between the sacrum and the rectum.[1-3] The limits of the presacral space are: posteriorly sacrum and coccyx, anteriorly rectum, superiorly S-2 and laterally ureters and iliac vessels.[3,4] The narrow space of the pelvis and the anatomical complexity of the presacral region present great difficulties in the surgical treatment of presacral tumors. Surgical errors in planning or during the operation can cause serious complications, such as bleeding from the presacral venous plexus and injury to the rectum or sacral nerve root. Because of the complex anatomy of this region, surgical treatment should usually be determined by cooperation between orthopedists, general surgeons, urologists and gynecologists.

Presacral tumors have been a challenge for surgeons for a long time due to the low incidence and written information. The incidence of presacral tumors ranges from one per 40,000 patients to 63,000 patients per year.[2-5] They are more frequent between the ages of 45 and 50 years with a predominance in women. A congenital and hormonal etiology has been proposed; hormonal changes in this age group appear to enhance the development of congenital presacral tumors.[1]

Currently, the Uhlig and Johnson classification is utilized, which categorizes tumors into congenital, neurogenic, osseous and miscellaneous.[4-6] Another way of dividing them into benign (the most frequent) or malignant, congenital or acquired; they are also classified according to their histologic type.[2,3]

Some authors have reported that the most common tumor is a congenital teratoma while others have reported a predominance of acquired teratomas.[2,7] Benign tumors are more common in females and in the earlier stages of life while malignant tumors are more common in males and in later stages of life.[2,5] We present the case of two patients

who were diagnosed with presacral tumor and underwent resection through a posterior approach.

CASE REPORT

Patient one

A 56-year-old male, originally from and resident of Mexico City. He presented with perianal pain associated with erectile dysfunction and constipation. He went to an outpatient clinic where electromyography was performed, and he was diagnosed with sensory neuropathy of the pudendal nerve (demyelinating type) and axonal degeneration. He began treatment with pregabalin with partial response. Two months after therapy, the patient experienced an exacerbation of symptoms. A physical examination with adequate coloration and hydration of the mucous membranes and integuments. Without cardiopulmonary compromise of the chest. The abdomen is soft and not painful, with no evidence of peritoneal irritation. A rectal palpation revealed a tumor on the posterior wall of the rectum at approximately 5 cm from the anal margin. The tumor was soft, with irregular borders.

Therefore, contrasted magnetic resonance imaging (MRI) of the pelvis was conducted. The results revealed a cystic tumor measuring 3.5×2.3×4.5 cm with a multiloculated aspect in the presacral space, with an apparent dependence on the sacral nerve tracts S4 an S5 and the inferior rectal nerve (Figure 1).

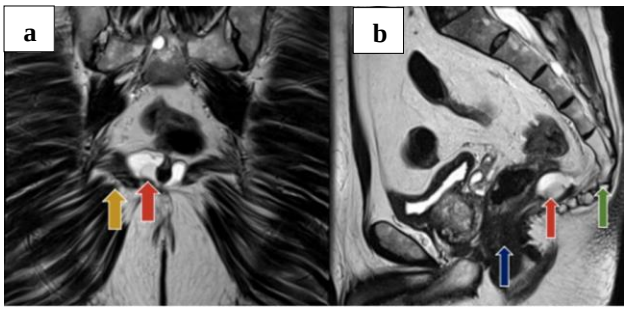

Figure 1: Contrasted magnetic resonance imaging of the pelvis (a) yellow arrow indicates nerve roots of S4 and S5 (pudendal nerve tract) displaced by tumor of cystic aspect, indicated with red arrow; and (b) blue arrow indicates tumor rectum, red arrow indicates tumor in pre sacral space, green arrow indicates sacral bone and coccyx.

In his medical history he denies smoking, and any allergies, transfusion or traumatic event. His surgical antecedents include an appendectomy in childhood, and the release of the left ulnar nerve. He reports being hypertensive of long evolution in treatment with amlodipine and candesartan.

The procedure involved the resection of a presacral tumor with a posterior approach and the resection of the coccyx, S-4 and S-5 (Figure 2). The findings of the procedure were a presacral tumor of approximately 4 cm.

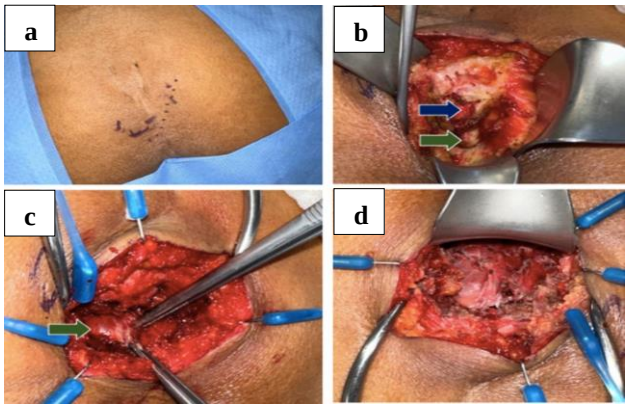

Figure 2: Resection of a presacral tumor with a posterior approach and the resection of the coccyx, S-4 and S-5 (a) marking of the incision to perform the posterior approach, in the lower part anal orifice; (b) the blue arrow indicates coccyx and with green arrow pre sacral tumor; (c) with green arrow pre sacral tumor being removed; and (d) resacral space with absence of coccyx, S4 and S5 to achieve R0 reseccion.

An examination of the pathology report revealing a diagnosis of retrorectal cystic hemartoma (Figure 3). After six days, he was discharged without any complications. His follow-up period was 4 months with no recurrence data.

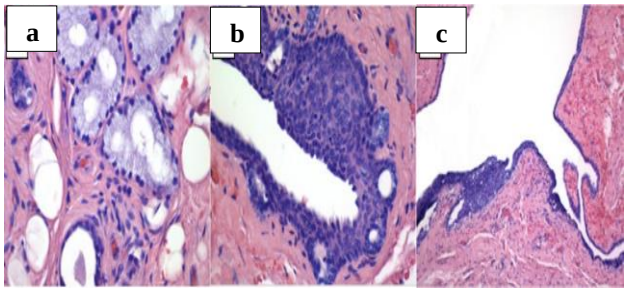

Figure 3: Histopathologic sections of pre sacral region (a) cystic space lined by cuboidal epithelium and urothelium immersed in perifectal smooth muscle, (b) benign urothelial lining of the cyst wall; and (c) adipose tissue, mucinous glands trapped at the cyst wall level.

Patient two

A 66-year-old female patient with an intermittent pulsating pain for 5 months in the left iliac fossa, intensity 5/10 and with occasional irradiation to the left lower extremity. Later she presented an exacerbation of the symptoms and irradiation to the dorsal region, so she went to orthopedics for evaluation. A magnetic resonance imaging (MRI) of the pelvis was requested, which revealed a presacral tumor of approximately 4 cm.

According to her medical history her mother passed away due to liver cancer, and her brother due to gastric cancer. Native and resident of Mexico City. She affirms to be allergic to sulfas, ampicillin and naproxen. A transfusion of a red cell concentrate was performed at 41 years of age due to spontaneous abortion. As for the surgical history, a laparoscopic cholecystectomy and arthroscopy of the left shoulder with repair of the rotator cuff ligament. Diagnosis of systemic arterial hypertension following treatment with lercanidipine and nevibolol/hydrochlorothiazide. Type 2 diabetes in treatment with metformin.

On physical examination, the chest showed no signals of cardiopulmonary compromise. The abdomen exhibits a globular shape, is depressible, does not cause pain upon palpation, and does not exhibit any evidence of peritoneal irritation. A rectal examination revealed a palpation of a tumor at approximately 6 cm from the anal margin, with regular edges, soft consistency.

It was decided to perform resection of the presacral tumor with posterior approach. A resection of the coccyx and S-5 was performed, and then a skin flap was advanced to achieve primary closure. The findings of the procedure were a presacral tumor of approximately 1.5 cm, with regular borders (Figure 4).

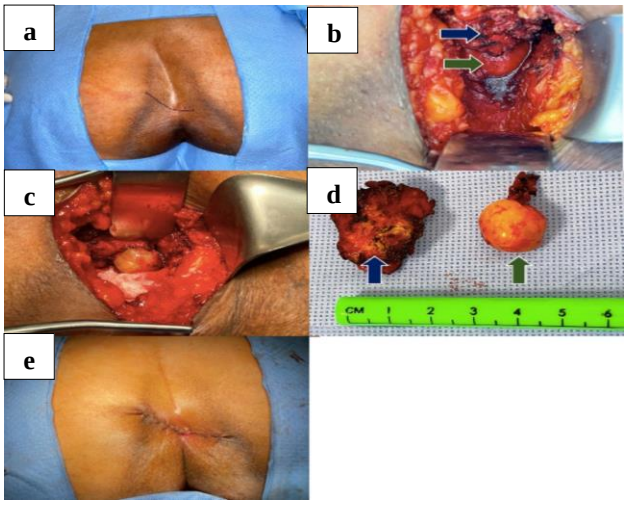

Figure 4: Approach to resection of the pre sacral tumor, (a) marking of the incision to perform the posterior approach, in the lower part, the anal orifice; (b) presacral space, with blue arrow coccyx and S5 with green arrow presacral tumor; (c) presacral space with absence of coccyx and s% to achieve R0 resection; (d) with blue arrow coccyx bone and S5, with green arrow pre sacral tumor of approximately 1.5 cm; and (e) closure of the incision with advancement of skin flaps.

A pathology report was received with microscopic description of PS-100 positive for neoplastic cells, actin negative, focal CD-34 positive, Ki-67 <3%, and a diagnosis of Schwannoma of 1.8 cm major axis (Figure 5). She was discharged without complications after three days. After two years of follow-up, she remained without recurrence.

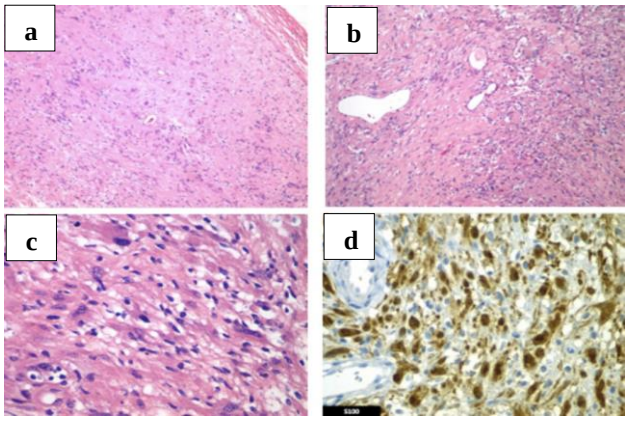

Figure 5: Pathology sections that suggest Schwannoma (a) benign encapsulated mesenchymal neoplasm with a growth pattern of long fascicles crisscrossing each other; (b) hyalinized vessels, intermingled mast cells; (c) spindle -shaped nuclei, ending in spikes and mild nuclear atypic; and (d) immunohistochemical reaction for protein S-100 positive in nucleus and cytoplasm of the neoplastic cells.

DISCUSSION

In presacral tumors, between 26% and 50% of patients are asymptomatic, this, associated with the nonspecific clinical presentation and the difficult anatomical localization can delay the diagnosis and treatment of presacral tumors.[1-3] Li et al have reported that the most common symptom, present in 56.8% of cases, is sacrococcygeal or perianal pain, followed by sacrococcygeal and perianal mass in 36.4% of patients.[1] A fundamental step is the rectal examination where it is possible to palpate the tumor in some cases providing a sensitivity of up to 62%, according to some series reported in the literature, tumors can be palpated between 82% and 90% of cases.[2,3,8] As with the first of our patients, other associated clinical data may appear, such as erectile dysfunction due to alteration of sacral roots S2-S4 or changes in bowel habit with constipation.[5]

Different imaging studies can be used to determine the location of the lesion and plan the therapeutic approach.[2] Computed tomography (CT) helps to differentiate between solid and cystic tumors and better visualizes the bony parts; on the other hand, magnetic resonance imaging (MRI) better visualizes the soft structures to plan the surgical approach.[2,5] MRI has a high accuracy (93%) to differentiate benign tumors from malignant tumors; however, the histopathological report continues to be the gold standard for definitive diagnosis.[1,4] The histopathologic report will establish the histologic lineage of the tumor, which will guide further therapeutic decisions such as neoadjuvant chemotherapy or transcatheter arterial embolization.[2] Recently it has been

observed that transcutaneous biopsy correlates with the final pathology report after surgical resection, thus helping to plan the safest surgical procedure by widening the surgical margins. However, this is still under discussion as the seeding of neoplastic cells with this technique has been proposed which could increase the recurrence rate.[4] The most common histologic strains are neurogenic tumors and epidermoid cysts, in our case the histologic strains reported are hemartoma and Schwannoma, those are enlisted in Table 1.[3]

Table 1: Histological strains.[10]

Congenital tumors	Dermoid or epidermoid cysts
	Teratoma
	Chondroma
Neurogenic tumors	Neurofibroma
	Neurofibrosarcoma
	Neurinoma
	Schwannoma
Osteogenic tumors	Osteoma
	Osteochondroma
	Sarcoma
Other	Lipoma
	Liposarcoma
	Fibroma
	Fibrosarcoma
	Hemartoma
Neuroendocrine neoplasms	

The gold standard for treatment is surgical resection which involves the removal of benign tumors that can lead to complications such as sexual, urinary or bowel dysfunction; or malignant tumors that can account for up to 40.9% of cases. Considering the possible etiology of these tumors, the administration of estrogen or progesterone receptor inhibitors has been proposed as a more conservative treatment.[1] Nonetheless, its utilization is subject to controversy owing to the resistance of these tumors.[2]

There exist various surgical techniques for resection: trans-sacrococcygeal, abdominal, abdominal-sacral, transrectal, transvaginal and even laparoscopic techniques.[2,3] In regard to the surgical technique, the posterior approach with trans-sacrococcygeal technique has been used more frequently due to his quicker approach and easy access. It also reduces the duration of hospitalization.[1,2] In our case, the two patients were treated with a posterior trans-sacrococcygeal approach. The literature describes that lesions smaller than 1 cm can be managed trans-sacrococcygeally and lesions larger than 1 cm or above S4 with an abdominal or mixed approach.[2-4] In our case, in both patients an R0 resection was achieved, however it was necessary to resect S-4, S-5 and coccyx in the first patient; while in the second patient S-5 and coccyx, data that correlates with what is suggested by some authors such as Dozois et al.[9]

In up to 26% of cases, complications such as bleeding, perforation of the rectum, nerve injury and urethral injury occur.[2] In our two cases there were no complications during the procedure.

The survival rate in certain series is variable, as evidenced by Li et al, wherein it is reported to be 100% in benign tumors.[1] It has been reported that malignant tumors have a 5-year survival rate of 75%.[2] In our case, in the four-month follow-up of one of the patients and two-year follow-up of the other, the survival rate was 100% without recurrences or complications.

Some studies such as Dziki et al report 11% recurrence in benign lesions and up to 40% in malignant lesions. It has been proposed that recurrences are due to lack of complete resection of the tumors or R1 resections.[2] Another important aspect proposed to avoid recurrences is to avoid tumor rupture during the surgical procedure, and also, it has been proposed to wash the presacral space with 2% iodine solution thus avoiding possible tumor seeding.[1,4] As part of the follow-up, PET-CT can be used.[2] In our case, since the lesions were benign, it was not necessary to perform further extension studies.

CONCLUSION

Although it is a rare pathology, the physician should be alert to patients who present with sacrococcygeal or perianal pain because it may be a presacral tumor which should be investigated further. As for the surgical approach, in the experience of MD. Daniel I. Camacho-Mauries, the posterior approach has shown good results in achieving an R0 resection.

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