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# Retrorectal Chordoma: Case Report and Literature Review

**Martínez Hernández Magro Paulino[1]*, Martínez Salcedo Néstor[1], Koelliker Ayala Enrique[2] and Guerrero Medrano Jaime[3]**

[1]*Colorectal Surgery Department, Hospital Guadalupano de Celaya, Celaya Gto, México*
[2]*Pulmonology, Hospital Guadalupano de Celaya, Celaya Gto, México*
[3]*Pathology Department, Hospital Guadalupano de Celaya, Celaya Gto, México*

## Abstract

**Objective:** To present a case of retrorectal chordoma and a literature review of this condition.

**Introduction:** Tumors of the retrorectal area are rather rare. Retrorectal lesions can be cystic or solid, benign or malignant, and can be classified as congenital, neurogenic, osseous or miscellaneous. Diagnosis is frequently delayed until the tumors reach considerable size. Wide en bloc resection should be performed once the diagnosis is made.

**Case report:** A 63 year old male patient, who complaint of progressive and recent onset constipation, and light pain at the coccigeal area. At the digital examination of the rectum we appreciate a firm, smooth presacral mass swelling the posterior rectal wall, with intact rectal mucosa. The MRI showed a multilobulated retrorectal mass; and the patient underwent to a wide en bloc resection of the tumor. The histopathological study of the specimen corroborates the presence of chordoma.

**Conclusions:** Retrorectal tumors are rare; their diagnosis is difficult and late. Treatment is surgical with wide resection; chordomas have poor sensitivity to radiotherapy and chemotherapy.

**Keywords:** Retrorectal tumors; Sacrococcygeal chordoma; Presacral tumors

## Introduction

The retrorectal space can harbor heterogeneous and rare lesions that may be silent and are thus difficult to diagnose. Tumors of the retrorectal area are rather rare; their incidence has been estimated to be 1 in 40,000 hospital admissions [1,2]. Retrorectal lesions can be cystic or solid, benign or malignant, and can be classified as congenital, neurogenic, osseous or miscellaneous [2,3].

The sacrococcygeal chordoma although rare is the most common malignancy of this area. Approximately one half of these lesions is located in the sacrococcygeal area, predominates in men rather than woman and is rarely founded in young patients.

Diagnosis is not infrequently delayed until the tumors reach considerable size. The relative rarity and anatomical position of retrorectal tumors may lead to difficulty in diagnosis and surgical treatment 4. Most surgeons encounter few of these tumors during their professional life; we present a case of a patient with a retrorectal tumor and constipation as a main symptom.

## Case Report

A 63 year old male patient with history of chronic obstructive pulmonary disease secondary to chronic cigarette smoking that comes to evaluation for a recent apparition and progressive constipation and light pain at the coccigeal area. He notes that his constipation starts two months ago, with straining at stool and denies bleeding, weight loss, and family history of cancer. At the physical examination he looks thin, with barrel chest, cyanosis in the nail beds, and expiratory wheezes, but with good general appearance. Abdominal region soft and not tender with no palpable organs or masses, inguinal area with no palpable nodes, perineal region appears normal, but at the digital examination of the rectum we appreciate a firm, smooth presacral mass swelling the posterior rectal wall, but with intact rectal mucosa at the rectoscopy confirming the extrarectal origin of the tumor. Colonoscopy discards other intra-colonic lesions, and the carcinoembryonic antigen was normal. Due to the presence of a presacral mass we perform a MRI that showed a multilobulated retrorectal mass localized below S5, with this findings our preoperative impression was limited to a retrorectal tumor, (biopsy should be avoided unless the lesion seems unresectable) (Figure 1). At pulmonologist evaluation they founded in chest films hyperinflation with increased lucency of the lungs, and low flattened diaphragms, $SaO_2$ 84% and at spirometry forced expiratory volume in one second ($FEV_1$) of 38%, and recommend pulmonary physiotherapy prior to a surgical procedure. We decide both with the patient planning a surgical treatment and perform a posterior sagittal approach, with wide *en bloc* tumor and coccyx resection (Figures 2-4) under spinal anesthesia (due to his respiratory condition), thrombosis prophylaxis

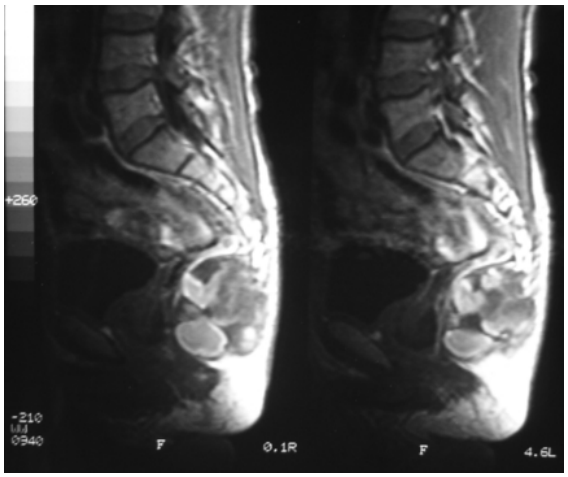


**Figure 1:** MRI with retrorectal mutilobulated tumor, below S5.

***Corresponding author:** Paulino Martínez Hernández Magro, Eje Vial Norponiente No 200 Consultorio 509, Col. Villas de la Hacienda, CP 38100, Celaya Gto, México, Tel: +52 (461) 6162123; E-mail: paulinomhm@hotmail.com

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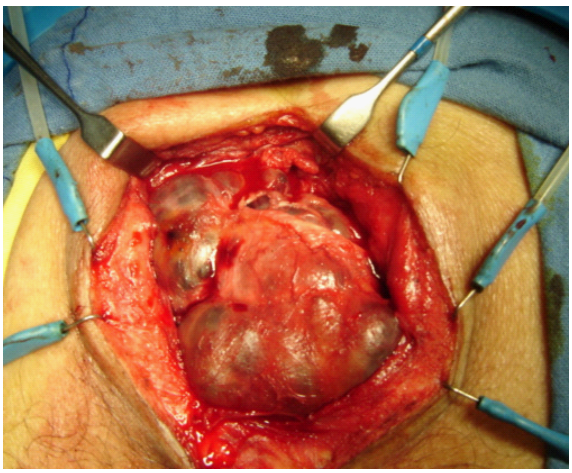

**Figure 2:** Posterior sagittal approach. A multilobulated tumor can be appreciated.

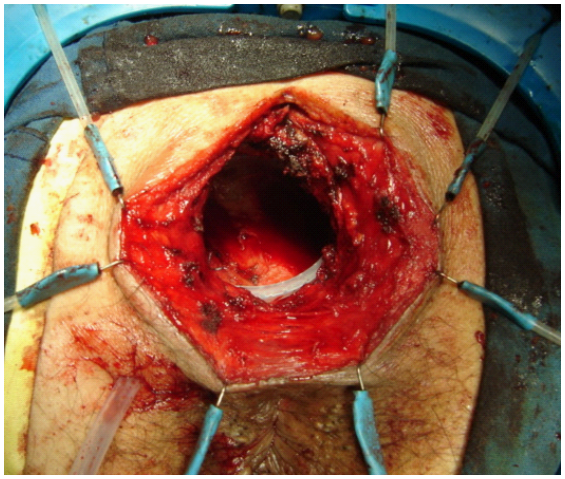

**Figure 3:** En bloc resection of the tumor and coccyx.

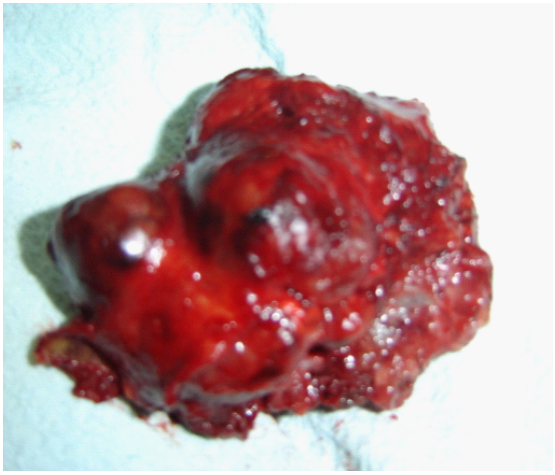

**Figure 4:** Surgical specimen.

and complete bowel preoperative preparation, finding a soft, multilobulated tumor approximately of 10 x 8 x 6 cm, with mucinous content which was resected in its totality, (resection was complete following the limits of the retrorectal space: the rectum anteriorly, the pre-sacral fascia posteriorly, and the endopelvic fascia laterally, the superior border of the space is the posterior peritoneal reflection of the rectum and the inferior border is Waldeyer fascia) and place a closed drainage at the resection site which was removed on the third day, with good outcome and resolution of his symptoms (constipation) and at a 15 month follow up without recurrence. The histopatological study of the piece confirms chordoma (conventional type) (Figure 5).

## Discussion

Retrorectal space is located anterior to the sacrum, and posterior to the rectum, its superior boundary is the peritoneal reflection, and inferior boundary the rectosacral fascia. Lesions in this area can arise from a variety of tissues, (this is an area of embryological fusion between hindgut and proctodeum, neural elements and bone) so a good knowledge of the embryology and anatomy of this space is essential to provide appropriate treatment [5].

Retrorectal lesions can be classified as either congenital or neoplastic. Congenital lesions represent 40% and neoplastic lesions 60%. Developmental cysts are the most common retrorectal cystic lesions in adults, occurring mostly in middle-aged women [6]. The Mayo clinic reported that 43% of all retrorectal tumors were malignant [7].

The first description of a sacrococcygeal chordoma was in 1900 by Henning. The sacrococcygeal chordoma is a tumor arising from remnants of the notochord [8-10], characterized by a slow and progressive growth that usually spans a period of years. Its invasion is by direct extension. Usually thought like a local disease but has been demonstrated distant metastases in > 40% of cases. The usual sites of distribution of chordoma are 50% sacrococcygeal, 35% sphenooccipital and vertebral in 15% [8]. The male: female ratio is 2:1 [8,11], and over one half of the lesions occur in persons aged 50-70 years [12]. The principal symptom is pain in 82% of the cases [10,13], symptoms are produced as the tumor proliferates, often reaching considerable size before diagnosis is made [10,14], the second most common symptom is constipation. If a malignant tumor invades sacral roots (S2-S4), fecal or urinary incontinence may ensue [13].

Chordomas are lobulated, pseudoencapsulated, gelatinous masses, with variable consistency, the lesions are malignant and tend to be locally aggressive, they often spread along the nerve roots in the sacral

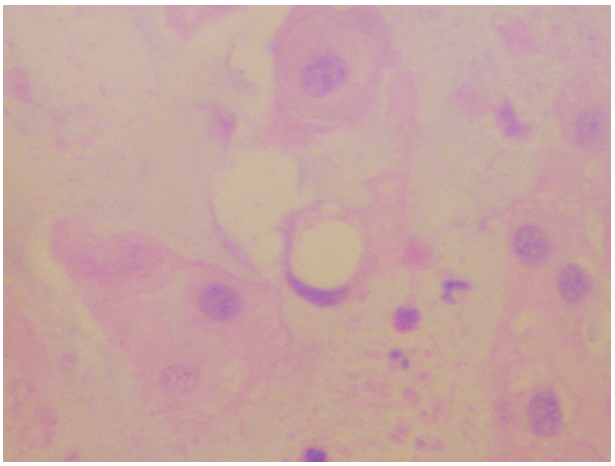

**Figure 5:** Histopatologyc study demonstrates the presence of physaliferous cells that confirms chordoma.

plexus, and usually do not infiltrate adjacent organs but recurrent tumors have this tendency [5]. Microscopically the large vacuolated cells (physaliferous cells) are characteristic of this tumor [8].

Nearly all of the patients have a palpable retrorectal mass on digital examination, so rectal examination is therefore the most important, most effective, and least expensive means to identify the tumor. Endoscopic and barium enemas can show extrinsic compression. Plain pelvic radiographs could show bone destruction in sacrum or coccyx. Endoluminal ultrasound can help to distinguish solid from cystic lesions. CT or IRM should be performed to confirm the presence of a retrorectal mass and to evaluate not only the tumor size but also its spatial relationships with pelvic organs and sacrum to decide the best approach for surgery.

It is recommended that all the retrorrectal lesions be extirpated, although they be benign and without symptoms [2]. Chordomas have poor sensitivity to radiotherapy and chemotherapy; they mainly are treated by surgery [8,15,16]. Complete removal of the tumor at the time of initial surgery is important for good prognosis. Based on CT or MRI findings, a surgical plan can be made for tumor excision, small and low lying (below level of S3) can be removed by a posterior approach [17]. If the upper pole of the tumor extends above the S3 level, and anterior-posterior approach is preferred [9]. In some cases of benign retrorectal tumors, laparoscopic approach has been reported [18].

The value of biopsy before curative resection is controversial. There are some reports which indicate that biopsy may cause seeding of tumor through otherwise unaffected tissue planes [19].

Metastasis is rare, and when it occurs is usually to lymph nodes, lungs and liver. The incidences of pulmonary and bone metastases could be high in patients with long term follow-up [8]. Recurrence after curative resection is frequent and may lead to a slow but relentless progression until death due to invasion of local pelvic structures [13]. In cases of recurrence, reexcision is a reasonable therapeutic option.

## Conclusions

Retrorectal tumors are rare; their diagnosis is difficult and late. Once the diagnosis is made, surgical therapy is mandatory even if the patient is asymptomatic, wide en bloc resection should improve survival, and decrease recurrence rates. Chordomas have poor sensitivity to radiotherapy and chemotherapy.

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