

## Case Report

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# Rectal leiomyosarcoma, three-year follow-up

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**ABSTRACT:** Rectal leiomyosarcomas are rare tumors originated from smooth muscle cells. Differential diagnosis includes gastrointestinal stromal tumors (GIST), leiomyomas or schwannomas, and the differentiation of these tumors is usually made through immunohistochemistry. Due to its rarity, the standard treatment has not been defined. The purpose of this study was to present the follow-up of a patient with leiomyosarcoma of medium rectum submitted to exclusive operative treatment. The tumor size was 6 cm and it had a high mitotic index. The patient remains with good urinary function and good sphincter function, and free of the disease after a three-year follow-up.

**Keywords:** leiomyosarcoma; rectal neoplasms; immunohistochemistry

**RESUMO:** Os leiomiossarcomas retais são tumores raros e originários de células de músculo liso. O diagnóstico diferencial inclui tumores estromais do trato gastrointestinal (GIST), leiomiomas ou schwannomas, e a diferenciação desses tumores normalmente é feita por imunohistoquímica. Devido a sua raridade, o tratamento-padrão ainda não está bem definido. O objetivo deste relato foi mostrar o seguimento de um paciente com leiomiossarcoma de reto médio submetido a tratamento operatório exclusivo. O paciente se manteve com bom controle urinário e boa função evacuatória. O tumor tinha alto índice mitótico e tamanho de 6 cm, mas não há sinais de recidiva após três anos da operação.

**Palavras-chave:** leiomiossarcoma; neoplasias retais; imunohistoquímica

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## INTRODUCTION

Rectal leiomyosarcoma is a rare disease and there are no prospective studies defining the best therapy to treat this type of tumor. For rectal tumors, abdomino-perineal amputation has been the treatment of choice when abdominal rectosigmoidectomy is not possible<sup>1</sup>. Recurrence rates after local excision seem to be very high, and the therapy with abdomino-perineal amputation has considerably reduced these rates. For being associated with high morbidity and definitive colostomy, less aggressive treatments are favored. More conservative surgeries are preferred for tumors of small size and low mitotic index, as they are less aggressive<sup>2,3,4</sup>. Some

authors suggest the use of preoperative radiotherapy to reduce the tumor size and, this way, allow the use of more conservative techniques that preserve the sphincter function, saving the patient from definitive colostomy<sup>1</sup>. This report presents the case of a patient with tumor of medium rectum, of 6 cm and high mitotic index, submitted to an exclusive operative treatment.

## CASE REPORT

A 70-year-old male patient, with history of tenesmus, recent intestinal constipation and hematochezia with 3-month progression. He presented prior history of diabetes *mellitus* and transurethral resection of the

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prostate (TURP) for seven years. Colonoscopy showed vegetative lesion of around 6 cm diameter, 4 cm from the anal margin; the remaining portion of the colon was normal until the cecum. The histological exam showed a neoplastic process with fusiform cells, suggesting a gastrointestinal stromal tumor (GIST, leiomyomas or neural tumor).

No neoadjuvant chemotherapy or radiotherapy was performed. The patient was submitted to abdominal rectosigmoidectomy combined with removal of perirectal fat, preserving the anal sphincter, and termino-terminal anastomosis of descending colon to lower rectum, 2 cm from the anal margin. No macroscopic metastases were observed. Protective ileostomy was performed. The patient did not present important complications in the immediate postoperative period. After the second week following the surgery, the patient presented urinary retention and infection, secondary to urethral stricture, associated with a prior urethra surgery. Urethral structure was treated with a transurethral surgery performed 18 months after the rectal tumor surgery.

The anatomopathological exam showed, through macroscopy, a voluminous lesion of 6 cm diameter, 1.5 cm from the distal margin of the surgical specimen. The microscopic analysis showed mesenchymal tumor of fusiform cells in the large bowel, polypoid growth type; moderate atypias and more than 10 mitotic figures per 50 high-power fields (HPF). This lesion affected the mucosa, the submucosa and part of the muscular tunica, without tumor emboli. The specimen margins were free of tumor. Fourteen lymph nodes were dissected from peri-intestinal fat and 1 lymph node of the mesocolon, all without metastases. The immunohistochemical analysis was positive for smooth muscle actin and negative for C-kit, CD 34 and S-100, confirming that it was a rectal leiomyosarcoma.

The ileostomy was closed six months after the abdominal resection. Anastomosis persisted with mild stenosis, which was digitally dilated for eight weeks. No adjuvant chemotherapy or radiotherapy was performed. The patient remains under monitoring and, three years after the diagnosis, remains free of the disease or metastases and presents excellent sphincter function according to the score of the Memorial Sloan-Kettering Cancer Center<sup>2</sup>.

## DISCUSSION

Rectal leiomyosarcoma corresponds to 0.1 to 0.5% of all malignant tumors of the rectum<sup>5</sup>. The incidence is higher in people of 50 to 70 years of age, regardless of the gender<sup>6</sup>. The dissemination tends to be hematogenic or by contiguity (with metastases especially to liver or lungs) and it rarely involves lymph nodes<sup>3</sup>. The most frequent symptoms are bleeding and perianal pain, as well as difficult evacuation, sensation of rectal pressure and diarrhea. However, up to 40% of the cases can be asymptomatic<sup>5,6</sup>.

Three fourths of all cases are located in the lower third portion of the rectum, 10% in the medium rectum and 15% in the upper rectum. More voluminous tumors are especially located in the upper rectum<sup>5,7</sup>. The tumor is an intraluminal or polypoid mass and, sometimes, with ulcerated, inflamed or necrotic mucosa. Some reports also describe intramural tumor, lobulated mass or transmural plaque-like lesion with extensive mucosa ulceration<sup>3,8</sup>. Histologically, it presents fusiform cells, which are similar to well differentiated smooth muscle fibers. Their nuclei are oval to moderately elongated and usually dead-end. They may have areas of coagulative necrosis or pleomorphism<sup>8</sup>. High-degree tumors are those of 10 or more mitotic figures per 50 HPF and low-degree tumors have less than 5 mitoses per 50 HPF<sup>8</sup>. The immunohistochemical pattern is positive for  $\alpha$ -smooth muscle actin and desmin (positive in 66% of the cases)<sup>8</sup> and negative for C-kit (or CD 117).

The prognostic factors vary in the literature and are not well defined, but there generally agreed that the size (over 5 cm) and the number of enlarged mitoses are the main factors of bad prognosis<sup>2,5</sup>. Leiomyosarcoma seems to have a better prognosis than the GIST with the same index of mitoses<sup>8</sup>. Yeh et al. suggest that being under 50 years old would also be a factor of bad prognosis and that extended follow-up is recommendable, as subsequent recurrences have been observed. In addition, they suggest that surgeries may not be adequate to this group, and that the use of adjuvant therapies should be considered<sup>5</sup>.

Due to its rarity, the best therapeutic approach has not been defined, as no prospective studies analyzing the treatment of this tumor have been conducted. Historically, abdomino-perineal amputation was the treatment of choice, due to its lower recurrence rate<sup>5,6</sup>,

but rectosigmoidectomy with coloanal anastomosis and extended local surgery have been used in cases of small and low-degree tumors, sometimes combined with adjuvant radiotherapy<sup>2,3,4</sup>.

Extended local resection can be used, without adjuvant therapies, for lesions smaller than 2.5 cm and confined to the wall, according to a series of 22 cases from Mayo Clinic<sup>4</sup>. Grann et al. suggest that, for tumors smaller than 5 cm involving anal sphincter preservation, adjuvant therapies with brachytherapy after transanal excisions can be an option to abdominoperineal amputation<sup>2</sup>.

Kiffer et al. defend the idea of postoperative pelvic radiotherapy to all patients, as it has no significant side effects, and that patients with smaller tumors could have more benefits with radiotherapy than those with larger tumors, of worse prognosis<sup>3</sup>.

In the case reported here, abdominal rectosigmoidectomy was performed with total excision of perirectal fat and colorectal anastomosis using the double-staple technique. The purpose was to have oncological resection and preserve anal sphincter and pelvic autonomic innervation, saving the patient from definitive colostomy. No adjuvant radiotherapy was performed, as the literature has no good evidences about the effectiveness of this treatment. Despite the tumor size of 6 cm and its high mitotic index, the patient progressed well and with good quality of life. The patient remains free of the disease after three-year follow-up.

It is not possible to recommend a standard therapy to treat this disease. Additional studies are required to analyze the progress of this type of tumor and the best treatment for it.

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