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RECTAL LEIOMYOSARCOMA MIMICKING AS A PARARECTAL GIST (GASTROINTESTINAL STROMAL TUMOUR) AND MANAGEMENT BY ABDOMINOPERINEAL RESECTION & LITERATURE REVIEW.

Oncology

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ABSTRACT

Rectal leiomyosarcomas are rare tumors originating 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 a rare case report of rectal leiomyosarcoma in a 55 year old male mimicking as a pararectal GIST and successfully managed with abdominoperineal resection.Patient was discharged without any complications and is disease free 4 years after surgery.

KEYWORDS

leiomyosarcoma; rectal neoplasms; immunohistochemistry; abdominoperineal resection.

INTRODUCTION: Rectal leiomyosarcoma is a rare disease and there are no prospective studies defining the best therapy to treat this type of tumor. They are often confused for a pararectal GIST and managed accordingly but can be differentiated making use of Immunohistochemistry examination.For rectal tumors, abdominoperineal resection has been the treatment of choice when abdominal rectosigmoidectomy is not possible[1] .Recurrence rates after local excision seem to be very high, and the therapy with abdominoperineal resection 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[2,3,4]. 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[1]. This report presents the case of a patient with tumor of lower rectum 12 X 10 cm submitted to an exclusive operative treatment.

DISCUSSION

Rectal leiomyosarcoma corresponds to 0.1 to 0.5% of all malignant tumors of the rectum[5]. The incidence is higher in people of 50 to 70 years of age, regardless of the gender[6]. The dissemination tends to be hematogenic or by contiguity (with metastases especially to liver or lungs) and it rarely involves lymph nodes[3]. 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[5,6]. 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[5,7]

The spread of rectal leiomyosarcoma is mainly local or hematogenous, although lymphatic metastasis has been reported in some poorly differentiated tumors [15]. In our case, no metastasis was found in resected perirectal lymph nodes. The optimal treatment modality in patients with rectal leiomyosarcomas is controversial. Wide local excision and radical surgery, such as anterior resection or abdominoperineal resection, are commonly used. As shown in many studies, radical surgery is associated with a lower recurrence rate than wide local excision [12]. However, differences in survival rates were not statistically significant, regardless of the treatment modality [12,13]. Pelvic radiation therapy is generally considered unsuccessful. Minsky et al. reported moderate success using radiation therapy following surgery [13]. In contrast, Consentino et al. demonstrated that neither adjuvant radiation therapy nor chemotherapy is effective [14]. Chemotherapy has been generally unsuccessful in treating this tumor. The 2 most commonly used agents, doxorubicin and dacarbazine, are associated with low response rates ranging from 15% to 30% [14]. Unfortunately, there are few data regarding the efficacy of adjuvant treatments in the literature to draw definitive conclusions. The overall prognosis is poor, with reported survival of 20–40% at 5 years and significant additional mortality reported during later years. Tumor size and the degree of differentiation are known to be the most significant prognostic factors.

If they are not detected by endoscopic examination, an accurate diagnosis may be difficult because their clinical presentation can mimic that of primary ovarian carcinoma. In a case report by Yung-Taek Ouh et al a woman who had a large pelvic mass that was suggestive of a primary ovarian malignancy but was finally proven to be a leiomyosarcoma of the rectum.[10]

The tumor is an intraluminal or polypoid mass and, sometimes, with ulcerated inflammed or necrotic mucosa. Some reports also describe intramural tumor, lobulated mass or transmural plaque-like lesion with extensive mucosa ulceration[3,8]. 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[8]. High-degree tumors are those of 10 or more mitotic fgures per 50 HPF and low-degree tumors have less than 5 mitoses per 50 hpf[8]. The immunohistochemical pattern is positive for α-smooth muscle actin and desmin (positive in 66% of the cases)[8] and CD 99 and negative for c-kit (or CD 117) and S-100.

A distinguishing feature of our case is that the rectal tumor mass was not identified on total colonoscopy. Generally, colonoscopic examination including sigmoidoscopy shows a polypoid, submucosal mass occupying the lumen, leading to the suspicion of a tumor originating in the rectum. In such cases, rectal bleeding or obstruction can occur, prompting an evaluation of the possibility of colorectal problems. However, some rectal leiomyosarcomas growing away from the lumen, so-called exocolic growth, may not be detected on colonoscopic examination. The growth pattern of the tumor and the lack of specific symptoms might be responsible for the failure of preoperative diagnosis.

Pathologically, leiomyosarcomas can be distinguished from leiomyomas on the basis of the following features: larger tumor cells, fewer stromal fibers, increased mitotic activity, and nuclear pleomorphism .Among these findings, the presence of mitoses are the hallmark of malignancy (5 or more mitoses per 10 high-power fields) [11].

In our case the tumor was present in lower third rectum and was confused as a pararectal GIST. We made use of immunohistochemistry to make a correct diagnosis consistent with a rectal leiomyosarcoma which is extremely rare in this location.

HPE exam was suggestive of a grade 3 leiomyosarcoma with a mitotic count of more than 20 per 10high power fields.

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[2,5].

Leiomyosarcoma seems to have a better prognosis than the gastrointestinal stromal tumour with the same index of mitoses[8]. 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[5].

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 resection was the treatment of choice, due to its lower recurrence rate[5,6] 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[2,3,4]. 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[4]. 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 resection [2].In a study of twelve cases by Wang et.al from china ,role of Abdominoperineal resectionas an appropriate treatment for large rectal leiomyosarcomas has been established .[9]

Kiffer et al. defend the idea of postoperative pelvic radiotherapy to all patients, as it has no signifcant side effects, and that patients with smaller tumors could have more benefits with radiotherapy than those with larger tumors, of worse prognosis[3]. In our case , we didn't give any adjuvant radiotherapy. 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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