Leiomyosarcoma of the Anal Canal: About One Case Report and Review of the Literature


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Abstract

Introduction: The leiomyosarcoma of the anal canal is a very rare tumor with poor prognosis. Its diagnosis without immunohistochemistry is impossible and its management is not yet clear. We report one more case and a review of the literature. Case report: A 65-year-old woman consulted in October 2018 for anal pain and a rectal syndrome that had been evolving for the last year, without rectal bleeding, nor transit disorder; but with the impairment of general conditions. The digital rectal assessment, which was painful, perceived a non-stenosising, circumferential tumor process of the anal canal, of stony consistency with sphincteric hypotonia. At the rectoscopy, the anal tumor was circumferential polypoid, neoplastic appearance whose biopsy with immunohistochemical study concluded to an anal leiomyosarcoma. In the extension assessment, pelvic MRI; and pelvic abdominal and thoracic CT-scan did not reveal metastases. An abdominopelvic amputation was performed without incident and with good postoperative progress. Conclusion: The frequency of leiomyosarcomas of the anal canal is extremely low and only a few cases have been reported. The confirmation of the diagnosis is by biopsy and immunohistochemistry. The surgery suggested in the literature is an abdominoperineal amputation (PAA), made in our case with good results.

Keywords: Anal leiomyosarcoma, immunohistochemistry, abdominoperineal amputation.

INTRODUCTION

The Leiomyosarcomas are mesenchymal tumors that originate from smooth muscle fibers. The rectal and anal localizations are rare, representing less than 0.1% of all rectal malignancies [1]. In addition to its rarity, the optimal treatment of anal leiomyosarcoma is not yet clear and the prognosis remains poor. We report an exceptional case of anal leiomyosarcoma and discuss the management with literature data based.

CASE REPORT

It was a 65-year-old patient with no history who had been seen for anal pain associated with a rectal syndrome for 1 year without rectal bleeding and transit disorder associated in the context of impairment of general condition. The digital rectal examination, which was painful, perceived a left lateral tumor process of the anal canal, of stony consistency with a decreased sphincter tone.

At the rectoscopy, the anal tumor was polypoid and hemicircumferential, of neoplastic appearance whose biopsy with immunohistochemical study concluded to an anal leiomyosarcoma.
The other hand, only Yeh described, but their general nerve sheath can be excluded by the negativity of S100, which is the malignant counterpart of benign soft tissue tumors, such as neurofibromas and schwannomas. HMB45 is 100% specific and 93% sensitive for malignant melanoma [10]; thus, melanoma can be excluded by negative HMB-45 and S-100. Gastrointestinal stromal tumors are mesenchymal tumors originating from Cajal interstitial cells and characterized by a c-kit positivity closely resembling leiomyosarcomas. In fact, historically, many GISTs have been wrongly diagnosed as these. Here, GIST was excluded because of the negative expression of c-kit and CD 34. The hematogenous or contiguous dissemination (with metastases especially to the liver or lungs) and rarely involves lymph nodes [11]. In our case, the extension assessment was negative.

Due to its scarcity and limited published data, the treatment of leiomyosarcoma in the large intestine is controversial. Local excision or radical abdominal and perineal amputation (PAA) are the 2 surgical treatment options. Some authors recommend conservative treatment for localized tumors of low grade and less than 3 cm and reserve PAA for high-grade tumors and or more than 5 cm [12]. Chemotherapy and / or adjuvant radiotherapy for colorectal sarcomas have been described, but their place is not confirmed. The authors suggest that adjuvant therapy should be discussed in a clinical trial [4, 13, 14]. In our case, no postoperative treatment was indicated given the low grade and lack of data in the literature. In examining recurrence rates, Diamante and Bacon reported a rate of 86% after local excision [15]. Other authors reported that the local recurrence rate after WLE is 67.5%, while after the PAA it was 19.5% [16]. Although there is a difference in local recurrence in favor of PAA, there is no significant difference in survival [16, 17]. In fact, the most important prognostic factor is histological grade, while localisation, tumor size, age, and sex do not appear to affect local relapse or survival [14, 16, 17]. On the other hand, only Yeh et al., suggests that age <50 years is a factor of poor prognosis [18]. The prognosis of ano-rectal LMS is poor and the survival rate after surgical treatment is between 33 and 174 months [19]. Long-term follow up should be considered given the late nature of recurrence [13]. Overall survival at 1 year and 5 years is 83% and 46% respectively [20].

In summary, optimal treatment of leiomyosarcoma of the anus is not known. The surgical treatment for resectable tumors of the anal region is PAA. In some patients, conservative surgery followed...
by external beam radiotherapy and interstitial radiotherapy may be an alternative to radical surgery, with the aim of locally controlling the disease and preserving the anal sphincter.

**CONCLUSIONS**

The anal leiomyosarcoma is a very rare tumor, with a poor prognosis especially for high grade tumors. Surgery is the major therapeutic weapon and the indication of adjuvant treatment is not yet well established given the scarcity of this entity and the lack of prospective studies.

**REFERENCES**