



LEIOMYOSARCOMA OF THE ANAL CANAL: ABOUT ONE CASE REPORT AND REVIEW OF THE LITERATURE.

K.Gharbi , M.A.Lkousse , Y.Ismail , J.Atmani , A.Elfarouki ,
A.Ait Errami , S.Oubaha , Z.Samlani , K.Krati

Gastroenterology service, Hospital Mohammed VI Marrakech Morocco

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-stenotising, circumferential tumor process of the anal canal, of stony consistency with sphincteric hypotonia. At the rectoscopy, the anal tumor was circumferential polyploid, 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.*

Key words: 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.

MATERIALS AND METHOD

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 polyploid and hemicircumferential, of neoplastic appearance whose biopsy with immunohistochemical study concluded to an anal leiomyosarcoma.

The study of the biopsy concluded to characteristics compatible with anal leiomyosarcoma with fused cells arranged in interconnected bundles, with irregular nuclei, confirmed by an immunohistochemical study: Anti-AML +, anti-Desmin +, anti-Ki67 +, and anti-CD34 -, anti-CD117 -, anti-S100 -, anti DOG1.

Pelvic MRI found a parietal circumferential tumor process of the anal canal extended to the anal margin; budding endoluminal, measuring 11 mm thick and 4.6 cm high, with infiltration of the levator muscles of the anus, especially the right, without invasion of neighboring structures and without lymphadenopathy. (Fig.1)

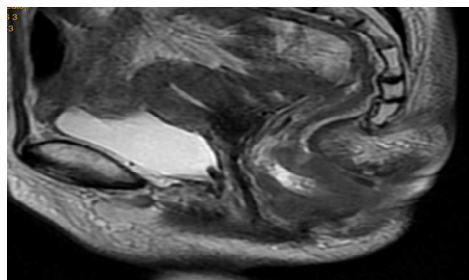


Figure 1. Pelvic MRI showing the tumor process

The thoracic and abdominal CT-scan as part of the extension assessment revealed no metastases, particularly hepatic or pulmonary, nor lymphadenopathies.

Thus, abdominal and pelvic amputation (PAA) was performed without incident during the surgery and with a good postoperative evolution.

RESULTS AND DISCUSSION

Discussion:

Digestive leiomyosarcoma are very rare tumors. Their incidence is higher among people aged 50 to 70, regardless of sex [2].

The most common site is the uterus followed by the gastrointestinal tract and the retroperitoneal region [3]. The anal and rectal localizations remain exceptional representing 7% of the leiomyosarcomas of the digestive tract [4]. The case of anal and rectal leiomyosarcoma was for the first time reported in 1908 by Exner . Evans [5] reported 56 cases of leiomyosarcoma of the gastrointestinal tract in 10 years, of which 4 were located in the rectum. Randleman et al [4] reported a series of 22 cases of anal and rectal leiomyosarcoma over a 35-year period and Walsh et al reported a series of 48 cases of anal and rectal leiomyosarcoma over a 31-year period.

There is no specific symptoms . The anal pain was the symptom for which our patient was admitted, associated with rectal syndrome as the master symptom.

Histologically, the leiomyosarcom of the rectum can come from the muscular mucosa, muscularis, or the wall of the blood vessels. They can develop in the light or in the perirectal tissue [6, 7]. The mucosa is affected in about 30-50% of cases [6].

In immunohistochemistry, leiomyosarcomas classically stain with smooth muscle actin, vimentin and desmin [8], and actin positivity of smooth muscle is present in 100% of cases. Basic calponin is a recently discovered marker that is an actin-binding protein, tropomyosin and calmodulin, isolated from smooth muscle and found in 75% of leiomyosarcoma cases [9]. Vimentin is likely to be positive in all these tumors and it would not help them to differentiate them.

The desmin is more of a skeletal muscle marker and may therefore be negative in leiomyosarcomas that have a smooth muscle origin. Malignant tumor from the peripheral 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.

CONCLUSION:

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.

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