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Metastatic Rectal Leiomyosarcoma Involving the Liver: A Case Report and Literature Review

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Abstract **Case Report**

Rectal leiomyosarcoma is a very rare tumor with a poor prognosis. We report the clinical case of a 45-year-old patient who presented with an externally palpable mass during defecation, accompanied by mild rectal bleeding. Colonoscopy revealed an ulcerated, budding, hemorrhagic, semi-circumferential rectal tumor. An anatomopathological examination with immunohistochemical study confirmed the diagnosis of leiomyosarcoma. The staging showed multiple secondary liver lesions. The case was discussed in a multidisciplinary meeting, and the decision was made to initiate first-line chemotherapy. However, the patient passed away before the treatment could commence.

Keywords: leiomyosarcoma, rectum, metastasis.

INTRODUCTION

Leiomyosarcomas are mesenchymal tumors that arise from smooth muscle fibers. Leiomyosarcoma of the rectum is a rare entity, accounting for less than 0.1% of all malignant rectal tumors [1, 2]. Only a few studies with long-term follow-up data are available regarding the optimal treatment of rectal leiomyosarcoma. Prognostic factors related to tumor progression and patient survival are not well known. Due to its uncommon location, we add another case of metastatic rectal leiomyosarcoma involving the liver to the literature and review the available data in the literature.

CASE REPORT

We present the case of a 45-year-old patient with a history of passive smoking, who presented with an externally palpable mass during defecation that had been progressively increasing in size for five months. The patient also experienced mild rectal bleeding and exhibited rectal syndrome symptoms (imprint, tenesmus, and false urges). All these symptoms occurred in the context of general weakness and apyrexia. Clinical examination revealed a patient with poor general condition, with a Karnofsky Performance Status score of 60, pale mucocutaneous appearance, and discolored conjunctiva. The patient was malnourished.

Digital rectal examination revealed a tumor located 0.5 cm from the anal margin, extending over 6 cm (the upper pole was not perceived). The tumor was ulcerated, budding, non-stenosing, and highly sensitive to touch. The sphincter tone was preserved. Vaginal examination revealed a bulge in the posterior wall of the vagina. No other abnormalities were detected during the clinical examination. Colonoscopy revealed a tumorous process located 5 cm from the anal margin, with an ulcerated, budding, hemorrhagic, semi-circumferential appearance extending up to 10 cm from the anal margin, causing narrowing but maintaining luminal patency. A biopsy was performed. Histopathological examination revealed a mesenchymal tumor proliferation consisting of spindle-shaped cells with a mitotic index of 7 mitoses per 10 high-power fields (Figure 1).

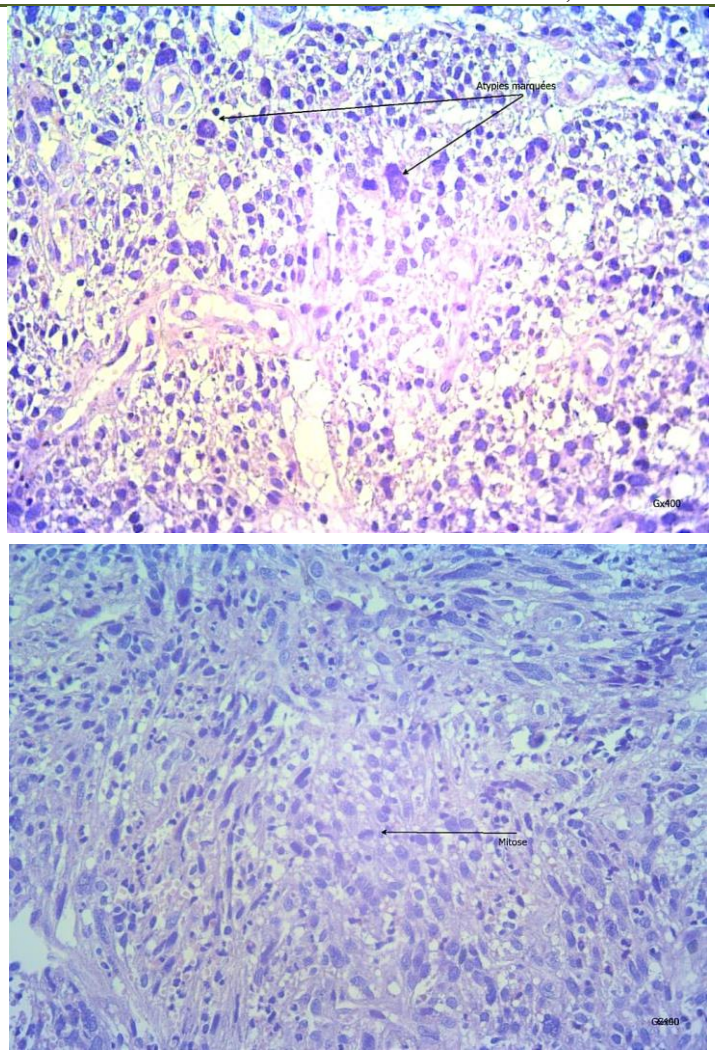


Figure 1: mesenchymal spindle cell proliferation with marked atypia and high mitotic index (HE X40)

The cytoplasm appeared pale eosinophilic. This proliferation was accompanied by abundant vascularization without notable tumor necrosis. The tumor infiltrated and ulcerated the mucosa. Immunohistochemical analysis was conducted using antibodies against pancytokeratin, desmin, CD34, CD117, and S100 protein. Strong cytoplasmic positivity for desmin was observed in the tumor cells (figure 2). The antibodies against CD117, CD34, cytokeratin, and S100 protein were negative (figure 3). The diagnosis of leiomyosarcoma was established.

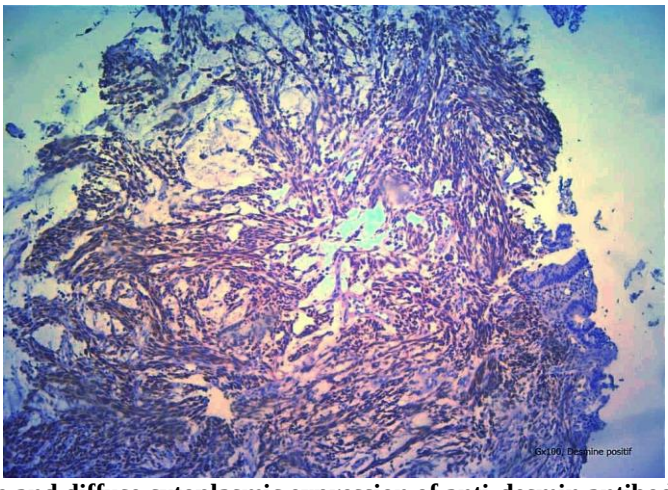
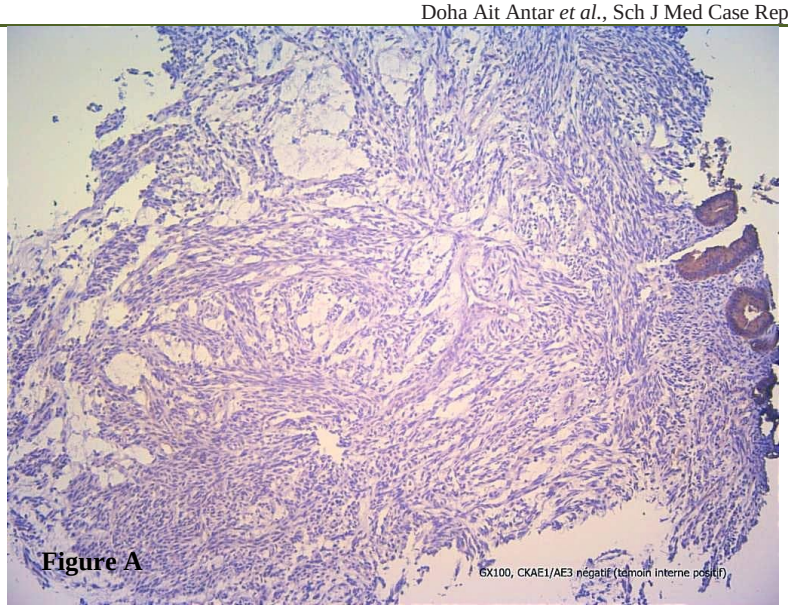
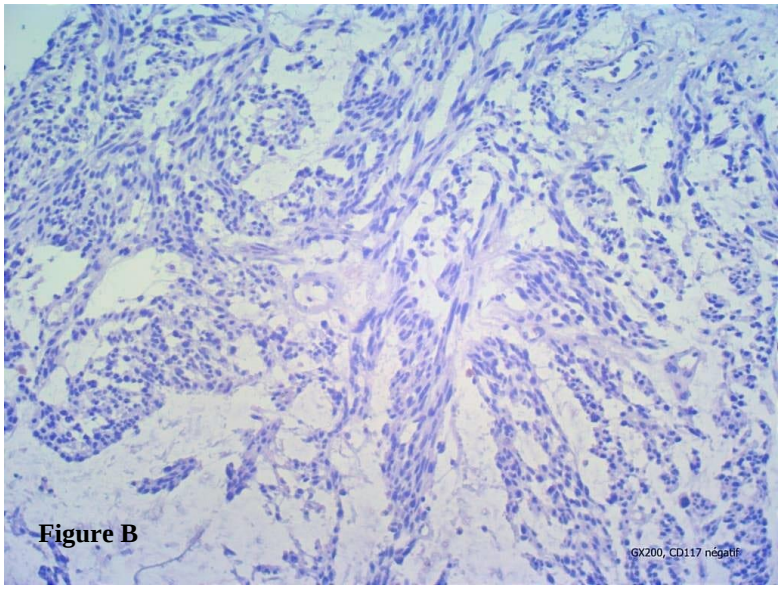


Figure 2: intense and diffuse cytoplasmic expression of anti-desmin antibody in tumor cells





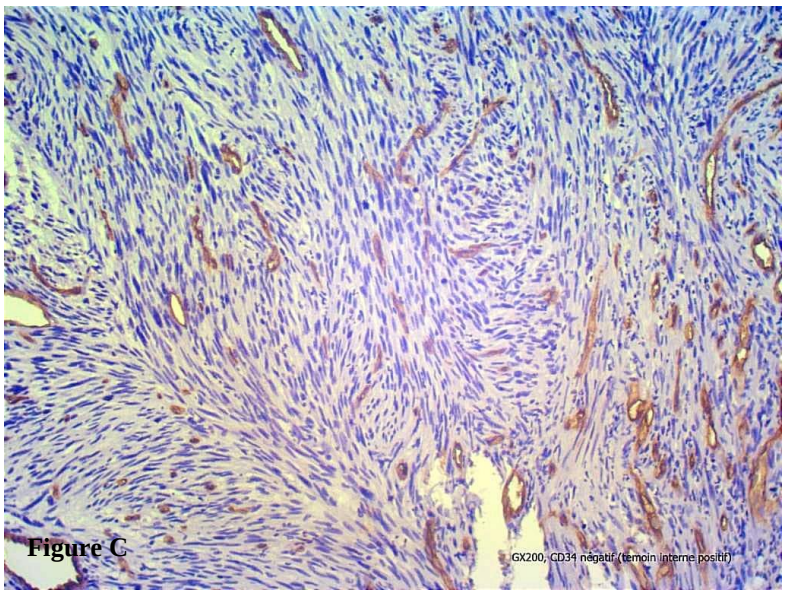


Figure 3: tumor cells not expressing anti-CK (figure A), anti-CD117 (figure B), anti-CD34 (figure C) and anti-pS100 (figure D) antibodies

A thoraco-abdominopelvic CT scan revealed a lesion in the middle and upper rectum, located 5 cm from the anal margin, budding and measuring 60.5 X 6.3 X 45 mm, spontaneously isodense, heterogeneously enhanced after contrast agent injection. It is associated with nodular infiltration of perirectal fat, with a nodule on the left side located less than 1 mm from the rectal fascia. Widely necrotic left internal iliac lymph nodes, with the largest measuring 32 mm in its smallest dimension. Enlarged liver (hepatic arrow measures 18.7 cm) with fairly well-defined masses, hypodense on spontaneous contrast and heterogeneously enhanced after contrast agent injection.

Hepatic MRI was performed to better characterize the liver lesions. It revealed an enlarged liver with multiple large right-sided liver lesions, with the largest measuring 14.74 x 11.14 cm. They are oval-shaped with hypointensity on T1, hyperintensity on T2, and heterogeneously enhanced after contrast agent injection. In our department, multidisciplinary tumor board meetings for colorectal malignancies have been established, involving visceral surgeons, medical oncologists, radiation oncologists, pathologists, and radiologists. Specific discussions on relevant clinical, pathological, and imaging data should take place regularly. The case was discussed in a multidisciplinary tumor board meeting, and the decision was to initiate palliative chemotherapy with doxorubicin and ifosfamide. However, the patient passed away before starting the treatment.

DISCUSSION

Digestive leiomyosarcomas are very rare tumors. The most common site is the uterus, followed by the gastrointestinal tract and then the retroperitoneal region [3]. Anorectal localization remains exceptional, representing 7% of gastrointestinal leiomyosarcomas [4]. It was in 1908 that Exner reported the first case of anorectal leiomyosarcoma. Evans reported 56 cases of gastrointestinal LMS in 10 years, of which 4 were located in the rectum [4]. Randleman *et al.* [4] reported a series of 22 cases of anorectal leiomyosarcoma over a period of 35 years, and Walsh *et al.* [2] reported a series of 48 cases of ano-rectal leiomyosarcoma over a period of 31 years. The median age was 65 years, ranging from 24 to 88 years [5]. Common symptoms of rectal leiomyosarcoma include transit disorders, pelvic pain, and rectal bleeding [6]. In our case, the patient was admitted with an unusual symptomatology characterized by reducible rectal prolapse.

The histology of leiomyosarcoma is characterized by a dense network of spindle cells with elongated and blunt nuclei. Hyperchromasia and nuclear pleomorphism are generally characteristic. The cytoplasm varies from typically highly eosinophilic to pale. Necrosis, hemorrhage, and cystic degeneration can frequently be observed [7]. In immunohistochemical examination, stains for SMA, desmin, and h-caldesmon are positive in the vast majority of specimens (>70%), while stains for CD117, CD34, S-100, and DOG-1 are negative in leiomyosarcomas [8]. According to the American Joint Committee on Cancer (AJCC) staging system for soft tissue sarcomas, histological grade, tumor size, and tumor depth are three clinicopathological risk factors for leiomyosarcomas [9]. Regarding histological grade, the Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) distinguishes the grade of malignancy based on the three factors of differentiation, necrosis, and mitotic rate [10]. However, the staging of rectal leiomyosarcoma has not been determined due to its rarity. Yamamoto *et al*. [11] reported that tumor size (≥5 cm) was significantly associated with poor prognosis. They also suggested that tumor depth and necrotic area could affect the prognosis. Yeh *et al.* [12] stated that young age (<50 years) and high histological grade of the tumor were independent prognostic factors for rectal leiomyosarcoma.

Hematogenous dissemination is the most frequent route of spread, and liver metastases represent the most common cause of death [13]. Lymphatic metastases are rare and may be found in poorly differentiated tumors [14]. Due to the rarity of rectal leiomyosarcoma, only a few case reports and minimal relevant data could be found in the literature. The standard treatment for rectal leiomyosarcoma remains to be determined. Surgery is the fundamental treatment for rectal leiomyosarcomas, but surgery alone is insufficient for locally advanced tumors. Radiotherapy and chemotherapy have been shown to be effective as perioperative treatment, although their efficacy is not remarkable [15].

Surgical approaches, including radical surgery such as anterior resection, abdominoperineal resection, and local excision, should aim to achieve macroscopically complete resection and minimal microscopic positive margins. Radical surgery is associated with a lower recurrence rate than wide local excision, must aim to achieve macroscopically complete resection and minimal microscopic positive margins. Radical surgery is associated with a lower recurrence rate than wide local excision, but there was no difference in overall survival between the two modalities [15]. In order to maintain local control and preserve the rectal sphincter, experimental perioperative radiotherapy for rectal leiomyosarcoma can be considered an extension of the treatment principles for rectal cancer and limb sarcomas [16].

Adjuvant chemotherapy may be an option for high-risk patients. Only two agents, doxorubicin and ifosfamide, have been confirmed to be effective as first-line general chemotherapy. In retrospective trials, the median progression-free survival was approximately 6 months, and overall survival was approximately 12 to

15 months for patients treated with doxorubicin and ifosfamide [17]. According to the SARC002 study [18] the combination of fixed-dose rate infusion of gemcitabine and docetaxel has been shown to be effective against pre-treated leiomyosarcomas with first-line chemotherapy and resulted in median progression-free survival and overall survival of 6.2 and 17.9 months, respectively.

García-Del-Muro *et al.* [19] demonstrated the feasibility and efficacy of a fixed dose of gemcitabine plus dacarbazine as second-line treatment for leiomyosarcomas and achieved median progression-free survival and overall survival of 4.9 and 13.8 months, respectively, in a randomized phase II trial. Trabectedin has been explored and shown a high rate of disease control (with progression-free survival rates of 26 to 30% at 6 months), particularly in patients pre-treated with doxorubicin and ifosfamide [20]. The prognosis of anorectal leiomyosarcomas is poor. One-year and five-year overall survival rates are 83% and 46%, respectively [21].

CONCLUSION

Anorectal LMS is a very rare tumor with a poor prognosis, especially for metastatic tumors. Currently, there is limited data available to define an optimal treatment.

REFERENCES

1. Minsky, B. D., Cohen, A. M., & Hajdu, S. I. (1991). Conservative management of anal leiomyosarcoma. *Cancer*, *68*(7), 1640-1643.
2. Walsh, T. H., & Mann, C. V. (1984). Smooth muscle neoplasms of the rectum and anal canal. *British Journal of Surgery*, *71*(8), 597-599.
3. Talikoti, M. A., Deo, S. V., Shukla, N. K., Kallianpur, A. A., & Gupta, M. (2011). A rare case of giant leiomyosarcoma in a filarial scrotum: a case report. *World Journal of Surgical Oncology*, *9*(1), 1-5.
4. Randleman, C. D., Wolff, B. G., Dozois, R. R., Spencer, R. J., Weiland, L. H., & Ilstrup, D. M. (1989). Leiomyosarcoma of the rectum and anus: a series of 22 cases. *International journal of colorectal disease*, *4*, 91-96.
5. Hoshino, N., Hida, K., Kawada, K., Sakurai, T., & Sakai, Y. (2017). Transanal total mesorectal excision for a large leiomyosarcoma at the lower rectum: a case report and literature review. *Surgical Case Reports*, *3*, 1-5.
6. Zbar, A. P., Sokolowsky, N., Sandiford, N., & Prussia, P. R. (2004). Leiomyosarcoma of the rectum. A report of two cases and review of the literature. *The West Indian Medical Journal*, *53*(2), 122-125.
7. Zambo, I., & Veselý, K. (2014). WHO classification of tumours of soft tissue and bone 2013: the main changes compared to the 3rd edition. *Ceskoslovenska patologie*, *50*(2), 64-70.
8. Robin, Y. M., Penel, N., Pérot, G., Neuville, A., Vélasco, V., Ranchère-Vince, D., ... & Coindre, J. M. (2013). Transgelin is a novel marker of smooth muscle differentiation that improves diagnostic accuracy of leiomyosarcomas: a comparative immunohistochemical reappraisal of myogenic markers in 900 soft tissue tumors. *Modern Pathology*, *26*(4), 502-510.
9. Edge, S. B., & Compton, C. C. (2010). The American Joint Committee on Cancer: the 7th edition of the AJCC cancer staging manual and the future of TNM. *Annals of surgical oncology*, *17*(6), 1471-1474.
10. Trojani, M., Contesso, G., Coindre, J. M., Rouesse, J., Bui, N. B., De Mascarel, A., ... & Lagarde, C. (1984). Soft-tissue sarcomas of adults; study of pathological prognostic variables and definition of a histopathological grading system. *International journal of cancer*, *33*(1), 37-42.
11. Yamamoto, H., Handa, M., Tobo, T., Setsu, N., Fujita, K., Oshiro, Y., ... & Oda, Y. (2013). Clinicopathological features of primary leiomyosarcoma of the gastrointestinal tract following recognition of gastrointestinal stromal tumours. *Histopathology*, *63*(2), 194-207.
12. Yeh, C. Y., Chen, H. H., Tang, R., Tasi, W. S., Lin, P. Y., & Wang, J. Y. (2000). Surgical outcome after curative resection of rectal leiomyosarcoma. *Diseases of the colon & rectum*, *43*, 1517-1521.
13. Deyden, W. E., & Wilkinson, C. A. (1978). Leiomyosarcoma of the rectum. *Southern Medical Journal*, *71*(6), 743-745.
14. Cuffy, M., Abir, F., & Longo, W. E. (2006). Management of less common tumors of the colon, rectum, and anus. *Clinical colorectal cancer*, *5*(5), 327-337.
15. Xu, D., Lv, Y., Chen, L., Chen, M., Wang, F., Song, Z., ... & Wang, D. (2019). Multidisciplinary evaluation of locally advanced leiomyosarcoma in the lower rectum: a case report and literature review. *Translational Cancer Research*, *8*(4), 1618.
16. Yang, J. C., Chang, A. E., Baker, A. R., Sindelar, W. F., Danforth, D. N., Topalian, S. L., ... & Rosenberg, S. A. (1998). Randomized prospective study of the benefit of adjuvant radiation therapy in the treatment of soft tissue sarcomas of the extremity. *Journal of clinical oncology*, *16*(1), 197-203.
17. Penel, N., Italiano, A., Isambert, N., Bompas, E., Bousquet, G., Duffaud, F., & French Sarcoma Group (Groupe Sarcome Français/Groupe d'Etude des Tumeurs Osseuses. (2010). Factors affecting the outcome of patients with metastatic leiomyosarcoma treated with doxorubicin-containing chemotherapy. *Annals of Oncology*, *21*(6), 1361-1365.
18. Maki, R. G., Wathen, J. K., Patel, S. R., Priebat, D. A., Okuno, S. H., Samuels, B., ... & Hensley, M. L.

(2007). Randomized phase II study of gemcitabine and docetaxel compared with gemcitabine alone in patients with metastatic soft tissue sarcomas: results of sarcoma alliance for research through collaboration study 002 [corrected]. *J clin oncol*, *25*(19), 2755-2763.

19. García-del-Muro, X., López-Pousa, A., Maurel, J., Martín, J., Martínez-Trufero, J., Casado, A., ... & Buesa, J. M. (2011). Randomized phase II study comparing gemcitabine plus dacarbazine versus dacarbazine alone in patients with previously treated soft tissue sarcoma: a Spanish Group for Research on Sarcomas study. *Journal of Clinical Oncology*, *29*(18), 2528-2533.
20. Samuels, B. L., Chawla, S., Patel, S., Von Mehren, M., Hamm, J., Kaiser, P. E., ... & Demetri, G. D. (2013). Clinical outcomes and safety with trabectedin therapy in patients with advanced soft tissue sarcomas following failure of prior chemotherapy: results of a worldwide expanded access program study. *Annals of oncology*, *24*(6), 1703-1709.
21. Wang, H. S., Chen, W. S., Lin, T. C., Lin, J. K., & Hsu, H. (1996). Leiomyosarcoma of the rectum: a series of twelve cases. *Zhonghua yi xue za zhi= Chinese Medical Journal; Free China ed*, *57*(4), 280-283.