

Colonic Metastases of Endometrial Stromal Sarcoma: A Case Report

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ABSTRACT

Endometrial stromal sarcoma (ESS) is a rare tumor classified into two distinct subtypes: low-grade and high-grade. Low-grade ESS is characterized by slow growth and the potential for recurrence or metastasis.

In this case report, we present the case of a 39-year-old woman who was admitted with severe pelvic pain. Her diagnostic workup revealed a low-grade ESS with colonic metastases.

The diagnosis is primarily established through a precise anatomic-pathological examination, specifically utilizing immunohistochemical analysis.

This case is of significant importance due to the rarity and uniqueness of metastatic colon cancer originating from uterine sarcomas. It underscores the importance of considering the possibility of metastatic disease, particularly when solitary colonic lesions are identified.

Keywords

Endometrial stromal sarcoma, Colonic metastases, Morphological and immunohistochemical study.

Introduction

Colorectal cancer (CRC) is the most common tumor of the digestive tract. In contrast, secondary colon tumors are a rare entity, accounting for approximately 1% of all colorectal cancers [1]. The most common pathway for metastatic spread to the bowel is peritoneal seeding, although hematogenous and lymphatic spread to the colon have also been reported, as seen in breast carcinoma, lung carcinoma, and melanoma [1].

In this report, we describe the case of a patient with colonic polypoid lesions that were subsequently identified as metastatic lesions of an endometrial stromal sarcoma (ESS). Endometrial stromal sarcoma is a relatively rare malignancy characterized by its indolent nature, with local recurrences and distant metastases potentially occurring even 20 years after the initial diagnosis [1]. However, the presence of colonic metastases is an unusual

clinical scenario, and preoperative radiologic findings of ESS are nonspecific, making anatomic-pathological diagnosis essential.

Case Report

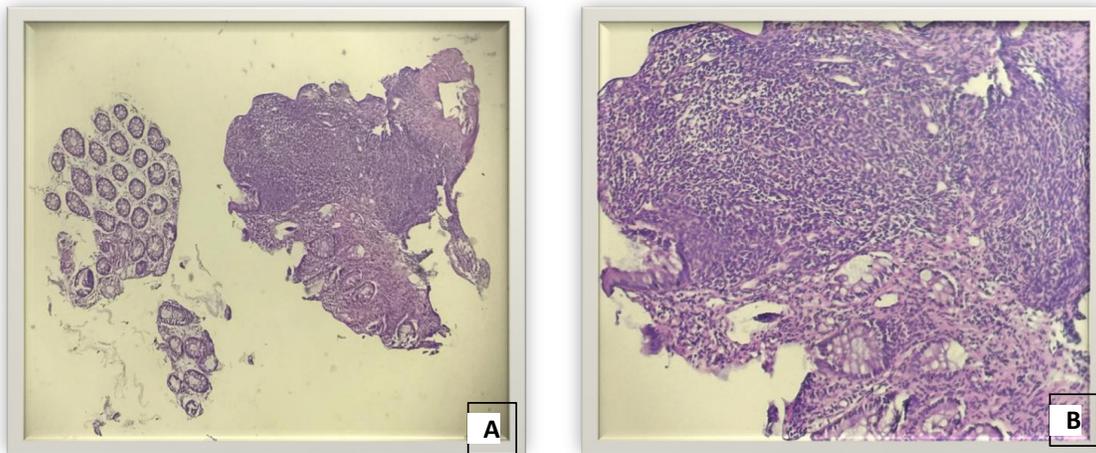
A 39-year-old female patient was admitted to the emergency department for pelvic pain of high intensity that had been evolving for 5 months with a recent deterioration in general condition and a weight loss of 5 kg over the past 2 months, without metrorrhagia or leucorrhoea. She was a Para 5, Live 4 and had a history of contraceptive use. Her general and systemic examinations were normal. An abdomino-pelvic CT scan was performed, which revealed a large pelvic mass exhibiting hemorrhagic changes. Additionally, a complementary MRI was conducted, confirming the presence of a retro-uterine pelvic mass measuring 16x11 cm, with signs of hemorrhaging.

The patient underwent a laparotomy, which revealed the presence of a friable mass with a soft consistency that bled upon contact. The mass measured approximately 10 cm and was adherent to the posterior surface of the uterus, positioned to the left. The

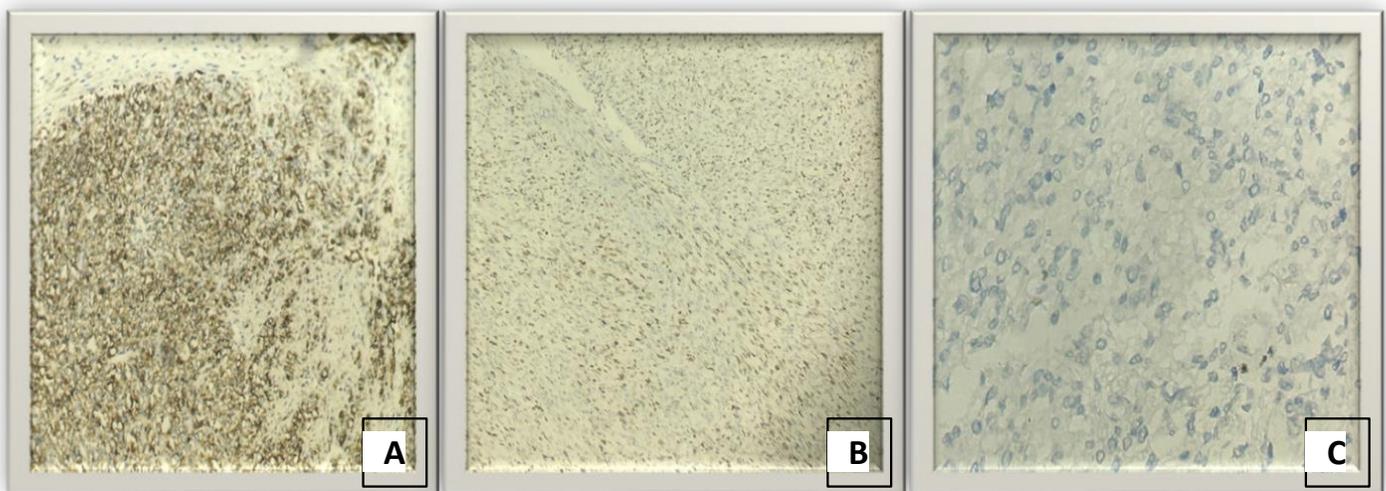
exploration also detected nodules in the sigmoid upon palpation. A right and left adnexectomy was performed along with the resection of the mass. Additionally, biopsies of the right and left parietocolic gutters and an omentectomy were conducted. After the surgery, a colonoscopy was performed, revealing subcentimetric polypoid lesions located at the rectosigmoid junction, with the largest measuring 7 mm. Biopsies were conducted. Histological examination showed a tumoral proliferation densely cellular, multinodular, monomorphic, made up of round cells, sometimes elongated, with fine chromatin nuclei, producing in places tubes or cords. Within this tumor proliferation, abundant vascularization was observed, made up of small blood vessels, surrounded by tumor cells.

An immunohistochemical study, as well as a FISH (fluorescent in situ hybridization) analysis, was conducted in our laboratory to confirm the diagnosis. The tumor showed strong positivity for CD10, estrogen, and progesterone receptors. It tested negative for AML, desmin, H- caldesmon, CK, and EMA.

On FISH analysis, the JAZF1 gene exhibited a rearrangement in more than 20% of the tumor cells. The morphological features, confirmed by immunohistochemical and molecular studies, allowed us to establish the diagnosis of an SSE. Based on these results, a conclusive diagnosis of low-grade endometrial stromal sarcoma (ESS) metastasized to the sigmoid colon was established, and the patient is a candidate for treatment with aromatase inhibitors.



A- Low power view of the colon biopsy (H&E stain 100× magnification): proliferation of small blue cells in the colonic mucosa.
B- Small spindled to oval uniform tumor cells (H&E stain 200× magnification).



Immunohistochemistry of Endometrial Stromal Sarcoma

A/ Diffuse staining with anti-CD10 antibody (IHCX200); **B/** Immunoreactivity for estrogen receptor (IHCX200); **C/** and lack of desmin immunoreactivity (IHCX400).

Discussion

Endometrial stromal sarcomas (ESS) are exceedingly rare malignancies, accounting for approximately 0.2% of all uterine tumors. The annual incidence of ESS is 1-2 cases per million women [1]. ESS predominantly affects younger women, with an average age range of 42-58 years, and 10-25% of the affected individuals are premenopausal [1].

There are two distinct subtypes, low-grade and high-grade, distinguished by differences in morphological atypia and proliferative activity [2]. High-grade ESS is aggressive, while low-grade ESS is a slow-growing tumor with a significantly more favorable prognosis. Nonetheless, approximately 50% of low-grade ESS cases may lead to recurrence and metastasis, which are often identified many years after the initial treatment [2,3]. Low-grade ESS can metastasize to the vagina, pelvis, and peritoneal cavity, which are typically the most common sites. Less common locations include the lungs, liver, bladder, breast, heart, brain, and bone [4]. However, in our case, metastasis of ESS to the sigmoid colon was demonstrated. Yuki Asada et al. reported the case of a 49-year-old woman who had undergone a hysterectomy for low-grade endometrial stromal sarcoma three years earlier and presented with lower abdominal pain. A sigmoidoscopy revealed a polypoid submucosal tumor, and histopathological analysis of biopsies from the lesion exhibited features similar to those of the resected uterine ESS [5].

Diagnosing LG-ESS (Low-grade endometrial stromal sarcoma) can be challenging when dealing with limited tissue samples, primarily for two reasons. First, the histological features of malignancy in LG-ESS may not be fully assessed in a biopsy sample. Secondly, LG-ESS originating from extrauterine locations often presents with characteristics that mimic those of more prevalent primary mesenchymal tumors, necessitating further studies [6].

The histological aspects of extrauterine ESS are identical to those of uterine site. These are multinodular tumoral proliferations, monomorphic in appearance, infiltrating in glove fingers or in tongues. The tumoral cells are fusiform or oval, not very atypical with mitoses not exceeding 10. The vasculature is abundant, made up of small blood vessels, surrounded by cell sleeves tumors giving the appearance of spiral arterioles. Lymph emboli are common. LG-ESS is diffusely and strongly positive for ER, PR, and CD 10. A panel of immunohistochemical stains is recommended when differentiating stromal neoplasms from cellular leiomyomas or leiomyosarcoma (NCCN) [7]. Gastrointestinal stromal tumor (GIST) and ESS will stain for CD 10 and desmin, but h-caldesmon and SMMS-1 will be positive in GIST and negative in ESS [8]. Gastrointestinal stromal tumor (GIST) and ESS have overlapping staining profile, as they both stain for CD10, desmin and smooth muscle actin (SMA), GIST is positive for c-kit (CD117) and DOG-1, while LG-ESS is negative [6].

Endometrial stromal tumors are a genetically heterogenous group of tumors that harbor recurrent chromosomal translocations, producing specific gene arrangements. The JAZF1- SUZ12 translocation is specific for LG-SSE and is not present in other uterine mesenchymal neoplasms [6]. The proposed treatment should be validated in an expert multidisciplinary consultation meeting. Surgery remains the primary therapeutic approach for primary low-grade SSE. Additionally, hormone therapy has been demonstrated to be effective in treating ESS, owing to the presence of estrogen and progesterone receptors [1].

Mansi et al. indicated that progesterone therapy should be the first-choice treatment for recurrent low-grade ESS, with resolution or stabilization of recurrent or metastatic disease observed in more than 50% of patients treated with progestin agents [9]. Our patient is a candidate for adjuvant treatment with aromatase inhibitors.

Conclusion

ESS is a rare uterine tumor, and metastatic lesions in the colon are equally infrequent clinical occurrences. Our case presents an ESS that has metastasized to the sigmoid colon, rendering it a unique and unusual scenario. Our case underscores the significance of utilizing morphological, immunohistochemical, and molecular data to confirm the diagnosis of low-grade endometrial stromal sarcoma.

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