

Endometrial Stromal Sarcoma of the Sigmoid Colon Arising in Endometriosis

: A Case Report with a Review of Literatures

Most of malignant tumors arising in ovarian and extraovarian endometriosis are carcinomas. Mixed müllerian tumor and endometrial stromal sarcoma arising in intestinal endometriosis are rarely described, but its clinicopathologic features have not been well characterized. Here we report a case of endometrial stromal sarcoma of the sigmoid colon arising in endometriosis with a review of six additional cases of endometrial stromal sarcoma arising in intestinal endometriosis found in English literatures. The patients ranged in age from 36 to 64 yr. Presenting symptoms were pain, bloody diarrhea, and tenesmus. Some patients had a previous history of endometriosis. Most of the tumors arose in the rectosigmoid colon. The histologic features were the same as their uterine counterpart. No death of disease had been reported. This rare tumor should not be confused with gastrointestinal stromal tumor clinically and histologically.

Key Words : Endometriosis; Sarcomas, Endometrial Stromal; Intestines

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Received : 11 December 2000
Accepted : 1 June 2001

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INTRODUCTION

Since Sampson first described the development of ovarian carcinoma in endometriosis in 1925, numerous case reports have documented the development of malignancies from endometriosis (1-3). Most of the malignant neoplasms are adenocarcinomas of endometrioid and clear cell types, and extrauterine endometrial stromal sarcoma is an extremely rare tumor (1-7). Recently, Yantiss et al. (7) reported only one endometrial stromal sarcoma among seventeen malignant neoplasms arising in gastrointestinal endometriosis. We found only six cases of endometrial stromal sarcoma of the intestinal tract arising in endometriosis in English literatures (1, 6-8). A case of endometrial stromal sarcoma of the sigmoid colon arising in endometriosis is described with a review of literatures.

CASE REPORT

The patient was 48-yr-old female who complained of difficult defecation and tenesmus. She had a history of subtotal hysterectomy for myoma uteri three years before, followed by total hysterectomy and salpingo-oophorectomy due to endometriosis of the left ovary the following year. On endoscopic examination, a polypoid lesion was found at the rectosigmoid area. Under the diagnosis of gastrointestinal stro-

mal tumor (GIST), segmental resection of the sigmoid colon with regional lymph node dissection was performed. On operation, tumor invasions to the urinary bladder and ureter were noted. Multiple nodules on the mesentery and lymph nodes enlargement were also found. Grossly, resected sigmoid colon had multinodular masses of 1-3 cm in size, involving from the mucosa to serosa (Fig. 1). Histologically, the lesion was characterized by round or tongue-like multinodular growth of closely packed plump spindle cells (Fig. 2) with prominent spiral arterioles (Fig. 3). The cellular atypism was minimal, and mitoses were rare. Rod- or star-shaped hyaline plaques were found. At the periphery and surface of the lesions, normal-appearing proliferative phase endometrial glands and stroma, corresponding to benign endometriosis, were present (Fig. 4). There were no periglandular stromal condensations that is characteristic of müllerian adenosarcoma. On immunohistochemical stain, the tumor cells were positive for vimentin, but negative for smooth muscle actin, S-100 protein, CD34, or c-kit (CD117). Most of the cells were also strongly positive for progesterone receptor, and weakly positive for estrogen receptor. Ki-67 labelling was low. Mesenteric nodules represented metastatic stromal sarcoma. The patient was complicated by postoperative disseminated intravascular coagulation (DIC), which was successfully managed and recovered. The patient was alive four months after the resection with no evidence of recurrence.

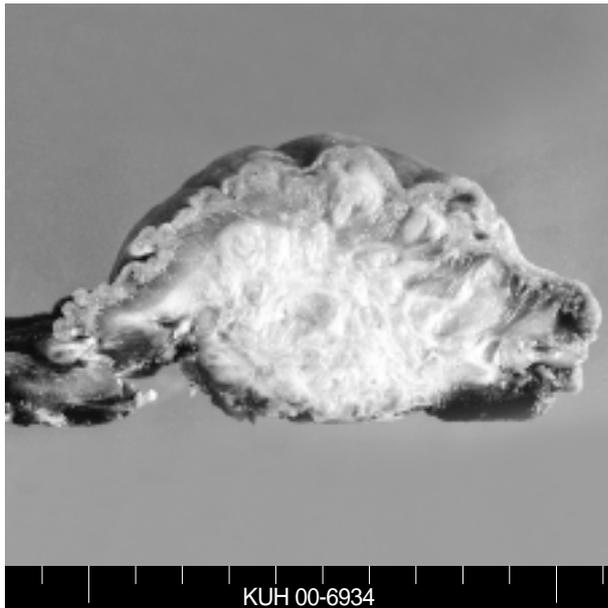


Fig. 1. Grossly, the resected sigmoid colon had multinodular masses involving the wall from the mucosa to the serosa.

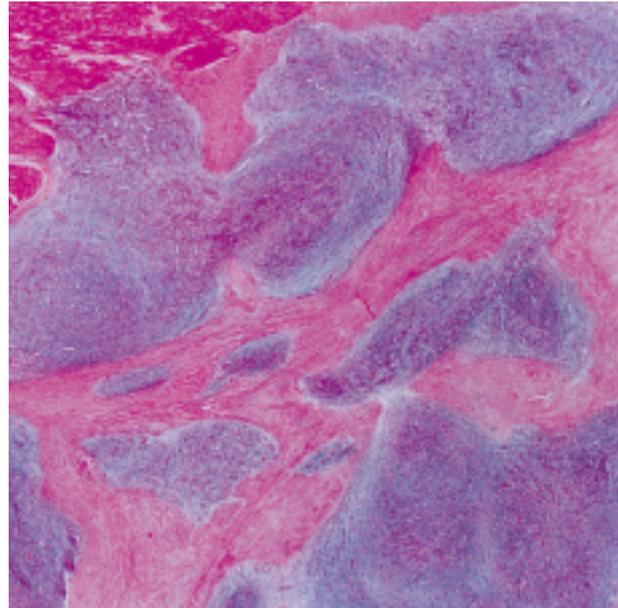


Fig. 2. The tumor is characterized by tongue-like multinodular proliferation of closely packed spindle cells (H&E, × 12.5).

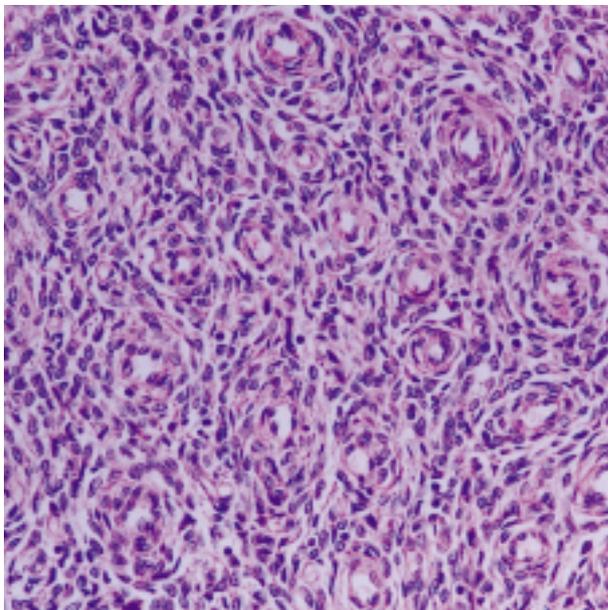


Fig. 3. The tumor was composed of short fascicles or sheets of monotonous plump spindle cells with abundant arterioles. A perivascular whorl arrangement is prominent (H&E, × 200).

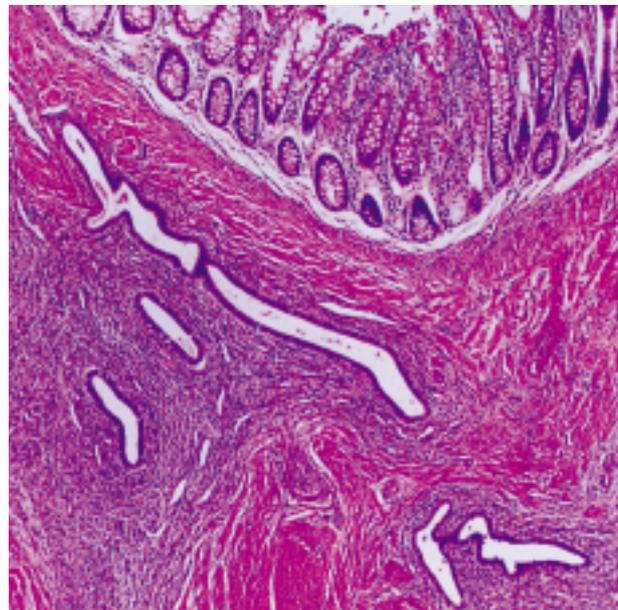


Fig. 4. At the periphery and surface of the tumor, glands and stroma, corresponding to benign endometriosis, are present. There are no periglandular stromal condensations that is characteristic of müllerian adenocarcinoma (H&E, × 40).

DISCUSSION

In 1925, Sampson defined an entity as originating from endometriosis when it satisfied three criteria: clear examples of endometriosis in close proximity to the tumor, no other primary site of malignancy, and histological appearance compatible with an origin from endometriosis (1). Many cases of

malignant neoplasms arising in ovarian and extraovarian endometriosis have been described thereafter (1-3). The frequency of malignant transformation of endometriosis is unknown, but it is estimated that up to 1% of women with endometriosis will develop endometriosis-associated neoplasm (2). Nearly 75% of the reported tumors arose in the ovary, but 21.3% appear in extragonadal sites (2, 4). The majority

of malignant tumors arising from endometriosis were adenocarcinomas, particularly endometrioid and clear cell carcinomas, but stromal sarcoma and mixed müllerian tumors can be developed (1-9). The same kinds of neoplasms have been reported in endometriosis of the intestinal tract (1, 6-9).

Recently, Yantiss *et al.* (7) described a large series of neoplastic and pre-neoplastic lesions of gastrointestinal endometriosis: among the seventeen cases, endometrial stromal sarcoma was only one and endometrioid adenocarcinomas were eight with four müllerian adenosarcomas, one endometrioid adenofibroma of borderline malignancy, one endometrial atypical hyperplasia, and one adenocarcinoma in situ. Using Sampson's criteria, we found only five additional cases of endometrial stromal sarcoma arising from intestinal endometriosis in English literatures (1, 6-9).

Seven cases including our case were analyzed to define the clinical and pathologic characteristics of this tumor. The seven patients ranged in age from 36 to 64 yr (average 51 yr). The two younger patients were nulliparous, and as for the other three, parity was not stated. Presenting symptoms were variable; pain in two, bloody diarrhea in one and tenesmus in one. Two patients had undergone previous surgical procedures aimed at treating the endometriosis (8). Among seven cases, five tumors were located in the rectosigmoid area. The rectosigmoid is an area of bowel having the highest incidence of endometriosis (4). Like their endometriotic precursors, the tumors occurred at various locations in the bowel wall. The pathologic features of the tumors were very similar and were virtually the same as their uterine counterpart. Although data concerning the survival were difficult to assess because the follow-up had generally been limited with only four cases with a documentation of follow-up for five or more years, no death of disease has been reported.

The endometrial stromal sarcoma of the intestinal tract arising in endometriosis should always be included in the differential diagnosis of small round cell or spindle cell tumors of the intestinal tract of female patients. The distinction from GIST should be considered, because the GIST is the most common mesenchymal tumor occurring in the intestinal tract and is managed differently from endometrial stromal sarcoma. Especially, if the underlying benign endometriosis is obscured, endometrial stromal sarcoma may be confused with GIST. In contrast to endometrial stromal sarcoma, most GISTs tend to be well-circumscribed with broad and pushing borders. The presence of characteristic morphologic features such as invasive "tongues" of tumor at the periphery of the neoplasm, short fascicles or sheets of monotonous plump spindle cells, and prominent arterioles should raise the suspicion of an endometrial stromal sarcoma. Immunopositivity for c-kit in GIST may be useful to distinguish these two entities (10). Müllerian adenosarcoma with sarcomatous overgrowth should also be included in differential diagnosis. Periglandular cellular stromal condensation, polypoid extensions of cellular

stroma into the glandular lumens, and various müllerian types of glandular epithelium favor the diagnosis of müllerian adenosarcoma with sarcomatous overgrowth. In our case, the presence of endometrial glands without periglandular stromal condensation and the immunohistochemical characteristics such as negative stainings for smooth muscle actin, S-100 protein, CD34, and c-kit were not consistent with GIST or müllerian adenosarcoma with sarcomatous overgrowth.

The present case was postoperatively complicated by DIC. A case of metastatic endometrial stromal sarcoma to the right ventricle associated with DIC has been described by Matsumoto *et al.* (11). Although the mechanism of DIC is not clear and its occurrence is rare, the possibility of DIC should be considered in the management of the patient with endometrial stromal sarcoma.

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