Undifferentiated spindle cell sarcoma at the anastomosis after ileocecal resection for colon cancer: A case report

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Case Report

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ABSTRACT

Key Clinical Message (44/50 words)

This report describes the first case of spindle cell sarcoma (SCS) occurring at the colonic anastomosis after ileocecal resection for colorectal cancer. SCS is rare, and the treatment strategy remains controversial. We report the diagnosis, management, and outcome of an 86-year-old woman with SCS.

KEYWORDS: Primary spindle cell sarcoma, colon cancer, surgery, ileocecal resection, case report

INTRODUCTION (1,638/3,000 words)

Primary spindle cell sarcoma (SCS) is a subtype of mesenchymal sarcoma, a broad term for cancers that arise in connective tissues, such as muscles, bones, cartilage, fat, blood vessels, and tendons. Primary mesenchymal sarcomas of the gastrointestinal system are rare, comprising merely 0.1–3% of all gastrointestinal tumors.[1]

SCS can occur in almost any part of the body,[2] but it is more common in superficial sites, such as the head, neck, and upper and lower limbs.[3] Individuals of all ages can be affected, but middle-aged and older adults (50–70 years) are the most common age groups, and men tend to be more affected.[4]One study reported that abdominal occurrence (133 in 3,299 cases) was a poor prognostic factor compared with occurrence in other parts of the body.[2]

Various treatment options have been described, including chemotherapy, radiotherapy, and surgery. Surgical resection achieves a better prognosis than non-resection,[3] and surgical therapy remains the gold standard for soft tissue sarcomas. However, the importance of lymph node dissection and extensive resection remains controversial.

In this report, we describe a case of SCS that arises at the anastomosis site after ileocecal resection for ascending colon cancer. SCS occurring in the intestinal tract is rare, and this is the first report of SCS occurring at an anastomosis site after colon resection for colorectal cancer.

CASE HISTORY

An 86-year-old woman presented to our hospital with a chief complaint of epigastric pain. She had undergone ileocecal resection for ascending colon cancer 10 years previously (pT2N0M0StageI; TNM classification 8th ed.), and no recurrence was reported. In June 2022, she visited her local doctor, who referred her to our hospital after finding an encapsulated fluid retention and scattered small nodules in the abdominal cavity on computed tomography (Figure 1).

Differential diagnosis, investigations, and treatment

The primary lesion was not identified on various examinations, but the lesion resolved spontaneously after approximately 6 months of follow-up. In February 2023, the patient was referred to our hospital for progressive anemia. A circumferential type 2 tumor was found at the anastomosis after resection of the ascending colon by colonoscopy, which was not found in July 2022 (Figure 2). The circumferential type 2 tumor was located at the ascending colon, and a biopsy revealed a malignant spindle tumor and the patient was referred to our department. Blood tests showed no elevation of tumor markers, and other tests were normal. Computed tomography revealed irregular wall thickening of 25 mm in the ascending colon and a 37-mm tumor in the mesentery of the small intestine, but no other obvious lesions (Figure 3). Magnetic resonance imaging showed a pale high signal on T2-weighted and diffusion-weighted images and a high signal on fat-suppressed T1-weighted images, suggesting mucus accumulation (Figure 4).

The patient was diagnosed with a malignant spindle cell tumor localized at the anastomosis. In March 2023, the patient underwent an open right hemicolectomy of the colon and limited lymph node dissection (because the lymph nodes had already been dissected in the previous surgery). Only one small nodule was found intraoperatively in the abdominal cavity. The nodule was submitted for pathological examination and diagnosed as scar tissue. The operative time was 252 min, with a blood loss of 120 mL. The patient was discharged on day 8 without any postoperative complications. Macroscopically, the anastomosis around the ascending colon, which was the primary site, revealed a circumferential 84×52 mm type 3 tumor with superficial necrotic tissue. The necrotic tissue adhered to the superficial layer (Figure 5A). Histologically, the tumor was greyish-white, dense, and situated in the intrinsic muscularis propria, with atypical spindle-shaped cells in the tumor area, infiltrating the subplasma membrane and showing atypical nuclear fission (Figure 5B). The colon serosa was filled with fibrous hyperplasia, vessel hyperplasia, inflammatory cell infiltration, and necrotic tissues. Further immunohistochemical analysis showed that the tumor cells positively stained for α1-antichymotrypsin (Figure 5C) and vimentin (Figure 5D), were S-100- and HMB-45-negative, and were MDM2-/CDK4-positive. Thus, the possibility of dedifferentiated liposarcoma could not be denied, and the pathological examination revealed an undifferentiated SCS centered on the anastomosis.

Outcome and follow-up

The patient recovered well. At the 1-year follow-up, computed tomography and colonoscopy did not reveal any signs of local recurrence or distant metastasis.

DISCUSSION

Sarcomas are malignant mesenchymal tumors and are divided into osteosarcomas and soft-tissue sarcomas. Gastrointestinal sarcomas are more common in the small intestine, stomach, and esophagus, and they occur mainly in older adults and less commonly in younger patients.[5] The most common clinical manifestations, in order, have been reported to be abdominal pain, abdominal mass, and fever.[6,7] Undifferentiated sarcoma, a soft-tissue sarcoma first described and named by Ozzello et al.,[8]is a high-grade, poor prognosis disease with a reported median survival of < 6 months.[9] Conversely, there have been reports of SCS occurring intraperitoneally. However, there have been no previous reports of SCS occurring intracolonally or at the anastomotic site (as observed in this case), making the current report the first of its kind.

As part of the body's natural response to soft tissue injury, spindle cells in injured tissue divide to promote healing. Normally, the spindle cells stop replicating once the affected area is healed. However, for reasons that are not fully understood, cells may continue to divide uncontrollably. Excess cells may then accumulate and combine to form an SCS. Risk factors include Paget's disease of the bone, previous radiation therapy for bone infarction, and osteomyelitis. Encapsulated fluid retention and scattered small nodules, which had spontaneously disappeared preoperatively, could not be seen intraoperatively in the abdominal cavity, and the nodules that were submitted to pathology were only inflammatory changes. When the abdominal cavity and the nodules were present, they were accompanied by a mildly elevated inflammatory response. However, the inflammatory response improved when the abdominal cavity and the nodules disappeared after follow-up. Thus, we judged this to be most likely an inflammatory reaction. The cause of the inflammation is unknown, but this inflammation may have also contributed to this condition. We hypothesized that chronic inflammation and regeneration of the previous intestinal anastomosis contributed to this risk.

SCS is a group of tumors morphologically composed of spindle-shaped cells, such as fibrosarcoma, synovial sarcoma, and malignant nerve sheath tumors. A diagnosis is difficult to make based on morphology alone. The final diagnosis is based on immunostaining and genetic testing. In the present case, the diagnosis was possible only with immunostaining. The tumor developed at the anastomotic site, and carcinoma sarcomatoid changes were first considered. However, this was ruled out because there was no clear evidence of carcinoma, and AE1/AE3 testing was negative. Other results from immunostaining indicated high malignancy and suggested a high-grade SCS. Although the tumor originated within the intestinal wall, GIST and leiomyosarcoma were also ruled out because of differences in histology. Synovial and clear cell sarcomas were also excluded, leading to a sarcoma diagnosis with an unknown differentiation direction.

In contrast, cells were positive for MDM2 and CDK4. This suggested the possibility of a dedifferentiated component in dedifferentiated liposarcoma (DLPS). In such cases, a well-differentiated liposarcoma (WLPS) component is often present in the surrounding areas. In the present case, no WLPS component was found in the excised specimen. There was a preoperative history of fluid retention and spontaneous disappearance of the nodule. Although spontaneous resolution of WLPS has not been previously reported, spontaneous resolution of tissue is difficult to assess. Only one small nodule was found intraoperatively in the abdominal cavity and was submitted to pathology, but the diagnosis was scar tissue, and no WLPS component was detected in the resection specimen. Therefore, although the possibility of DLPS is low in this case, careful follow-up will continue because this condition is rare, and no firm treatment guidelines have been established.

This is the first case of SCS occurring in the anastomosis reported in the literature; however, in general, the frequent recurrence of undifferentiated soft tissue sarcoma is problematic. In our patient, the tumor did not show distant metastasis and could be completely resected surgically despite undifferentiated sarcomas metastasizing early. Once distant metastases develop, they cannot be radically removed by surgery, and the prognosis is very poor. The overall 2-year survival rate for anaplastic sarcoma has been reported to be approximately 60%. The generally short follow-up period for patients with anaplastic sarcoma, with some patients missing follow-up data and no reports of survival rates beyond 5 years in the literature, indicates that the overall prognosis for patients with anaplastic pleomorphic sarcoma of the anastomosis may be even worse.[6] Local recurrence is also common in soft-tissue sarcomas, and Sawamura and Daigeler reported postoperative local recurrence within a median period of 19 and 15.7 months, respectively.[10,11] Thus, frequent follow-up is recommended after sarcoma resection, especially during the first 2 years.[12] Although complete surgical resection is the gold standard treatment, there are no fixed standards for lymph node dissection, partly because of the lack of reports. In the present case, although the lymph node dissection was limited because it had already been dissected in a previous surgery, complete resection was achieved grossly and on imaging, and a good prognosis was expected. Adjuvant radiation therapy and chemotherapy may be attempted postoperatively, depending on the patient's condition and intraoperative status; however, further research is needed to select a specific regimen because of individual differences in efficacy. Adriamycin and ifosfamide should be considered for the treatment of soft tissue sarcomas if recurrence is suspected; however, SCS of gastrointestinal origin is extremely rare, and there is no clear evidence. Further molecular biological studies of SCS may provide the basis for molecularly targeted therapies and immunotherapy and offer hope for improving the prognosis of patients with SCS.

CONCLUSION:

SCS is a rare disease, and this is the first reported case of SCS occurring in the colon or during colonic anastomosis. The treatment strategy remains controversial, and it is important to include more cases in future studies.

Author Contributions:

NK, TH, and MK conceived the presented idea, developed the theory, and performed the computations. MT, MN, TA, MI, RH, KT, and TT encouraged the investigation of specific aspects and supervised the findings of this study. All authors discussed the results and contributed to the final manuscript.

Acknowledgments

Not applicable

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Figure legends

Figure 1:

Abdominal CT and PET-CT findings A and B: CT showed fluid accumulation with contrast effect in the pelvis and abdominal cavity. PET-CT showed significant FDG accumulation.

Figure 2:

Colonoscopy findings: a circumferential type 3 tumor was found at the anastomosis after resection of the ascending colon, which was not found 1 year prior.

Figure 3:

Abdominal CT A and B: CT demonstrated an irregular wall thickening of 25 mm in the ascending colon and a 37 mm tumor in the mesentery of the small intestine, but no other obvious lesions.

Figure 4:

Magnetic resonance imaging (MRI) findings:

MRI showed a pale high signal on T2-weighted and diffusion-weighted images and a high signal on fat-suppressed T1-weighted images, suggesting mucus accumulation.

Figure 5:

A: Macroscopically, the anastomosis around the ascending colon, which was the primary site, revealed a circumferential 84×52 mm type 3 tumor with superficial necrotic tissue. The necrotic tissue adhered to the superficial layer.

B: The tumor was a grayish-white, dense tumor seated in the intrinsic muscularis propria, with atypical spindle-shaped cells in the tumor area, infiltrating into the subplasma membrane and showing atypical nuclear fission. The serosa layer of the colon was filled with fibrous hyperplasia, vessel hyperplasia, inflammatory cell infiltration, and necrotic tissues. The serosa layer of the colon was filled with fibrous hyperplasia, vessel hyperplasia, inflammatory cell infiltration, and necrotic tissues.

The dotted line is the anastomosis.

C and D: Further immunohistochemical analysis showed that the tumor cells were positively stained for α1-antichymotrypsin (Fig. 5C) and vimentin (Fig. 5D).

Figures

Figure 1

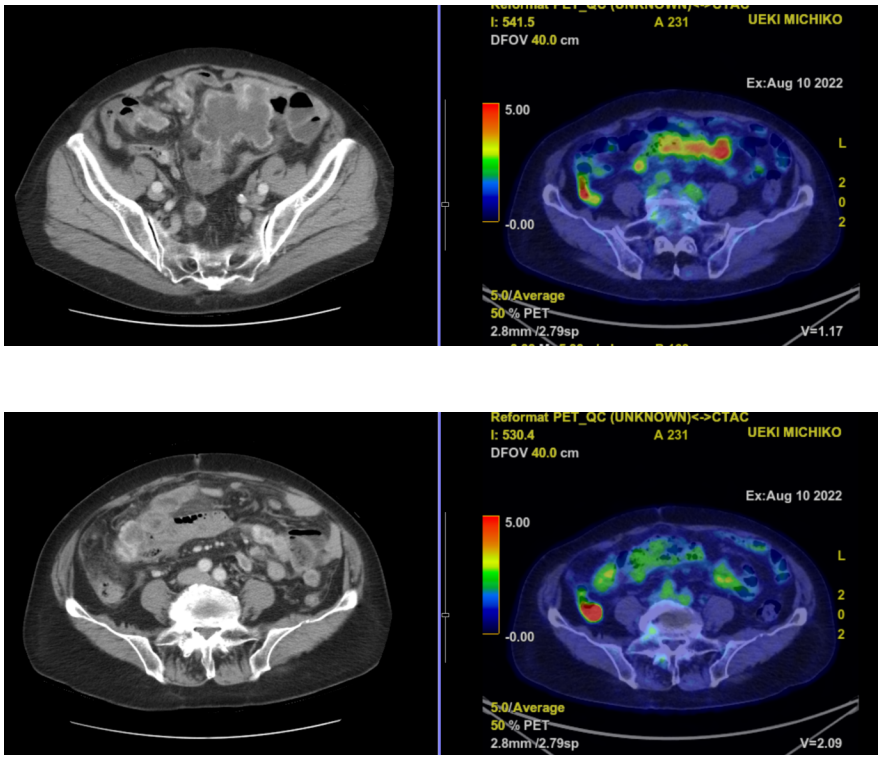


Figure 2

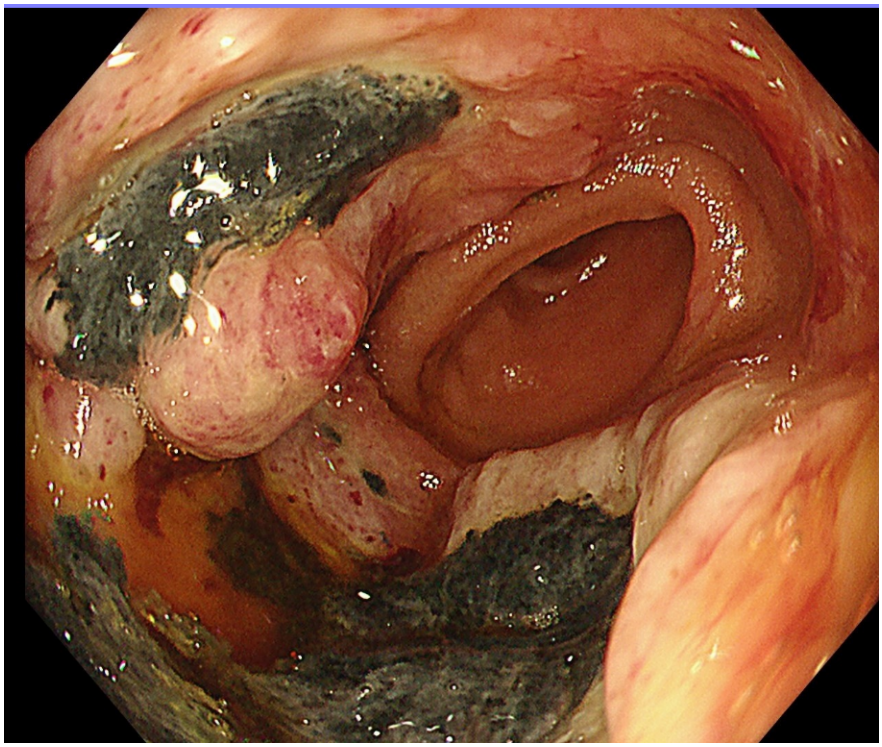
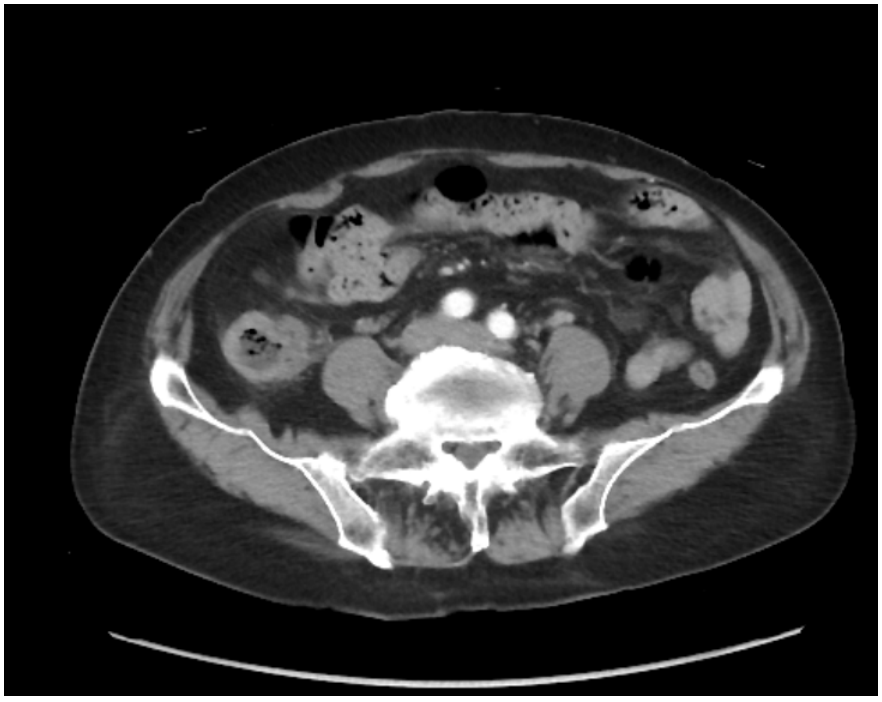


Figure 3



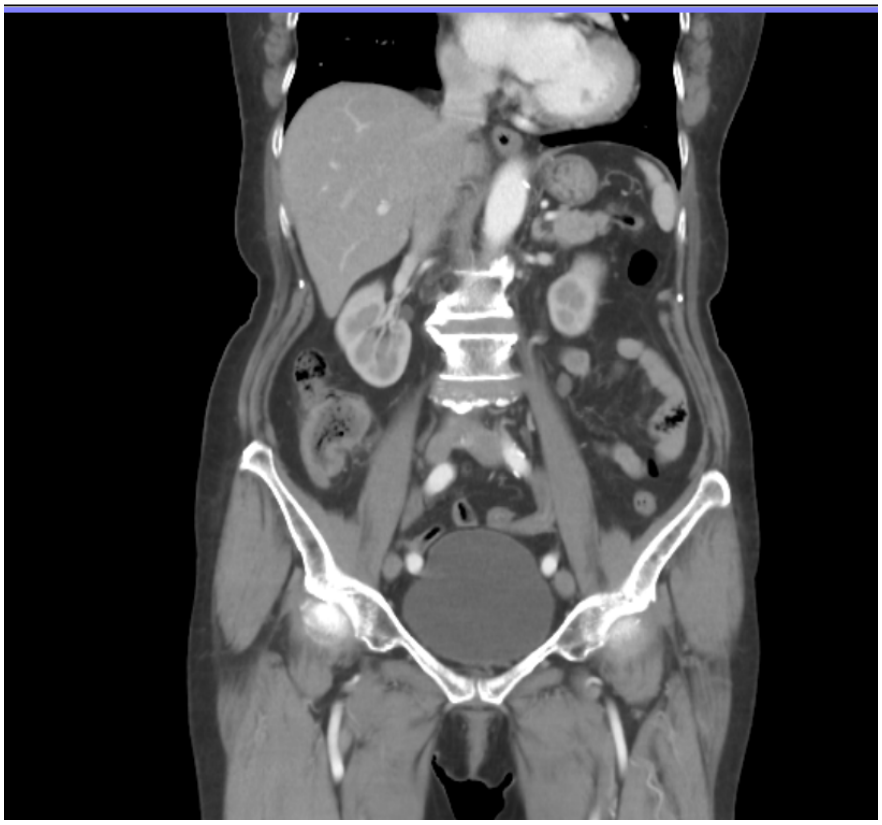
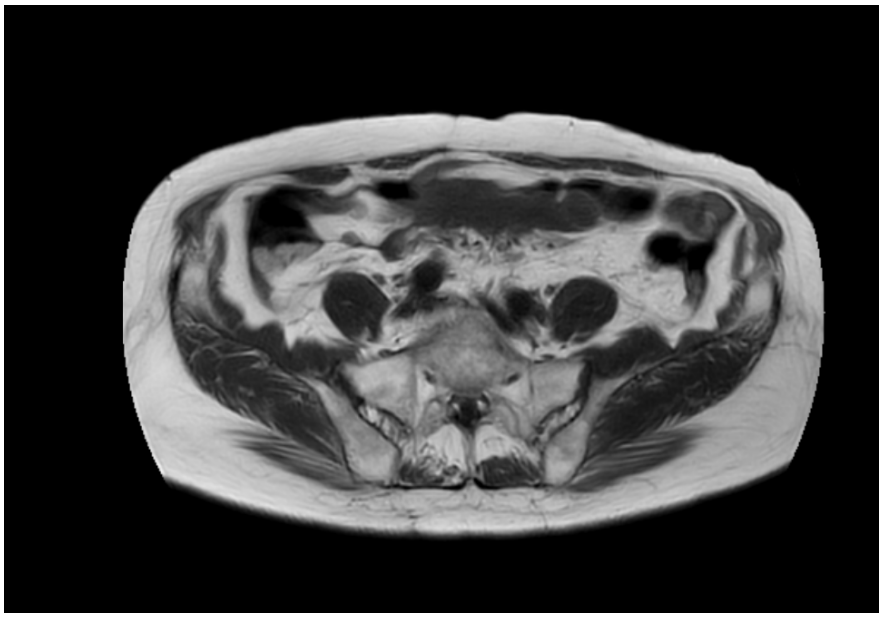


Figure 4



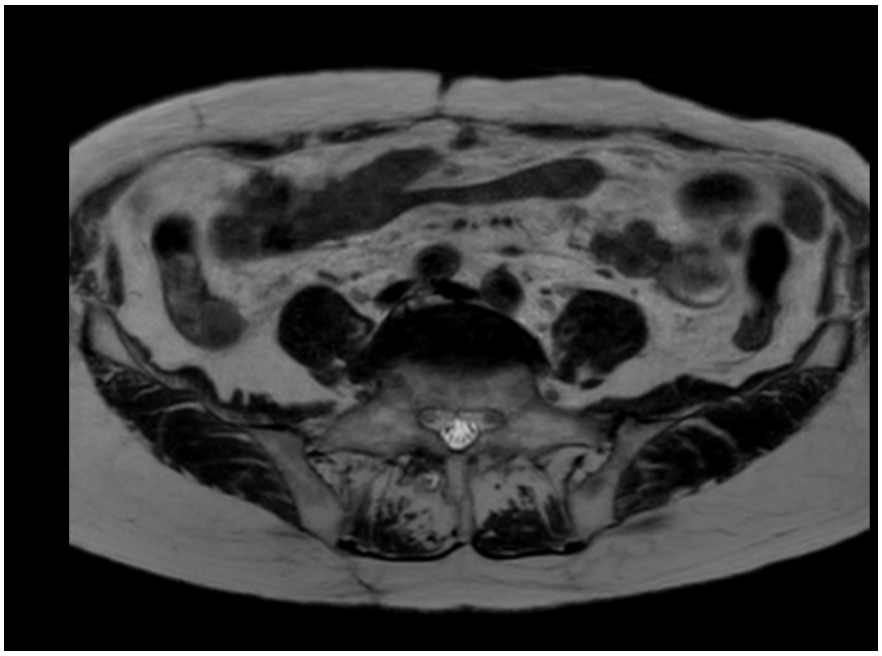


Figure 5