The First Documented Case of High-Grade Synovial Cell Sarcoma of the Rectum

Richard B. Hostetter, MD; Min Yan, MD; Houman Vaghefi, MD, PhD; Kenneth Pennington, MD; and Gary Cornette, DO

Abstract

A patient presented with signs and symptoms of tenesmus, urgency, and rectal bleeding that she had been experiencing over the course of several months. Full endoscopic evaluation showed a 6-cm submucosal mass approximately 10 cm from the dentate line projecting as an endoluminal mass with a large broad base. An initial endoscopic resection was attempted but aborted because of significant hemorrhage, and surgical oncology was consulted. After stabilization, the patient underwent a transanal resection of the mass the following day. An endoscopic gastrointestinal anastomosis stapler resulted in a margin-negative complete resection of what was later determined to be a high-grade synovial cell sarcoma. This case report presents the first known documented case of synovial cell sarcoma of the rectum. (JNCCN 2012;10:947–950)

Case Report

A patient presented with signs and symptoms of tenesmus, urgency, and rectal bleeding that she had been experiencing over the course of several months. Full endoscopic evaluation showed a 6-cm submucosal mass approximately 10 cm from the dentate line projecting as an endoluminal mass with a large broad base (Figures 1 and 2). An initial endoscopic resection was attempted, but was aborted because of significant hemorrhage, and surgical oncology was consulted. After stabilization, the patient underwent a transanal resection of the mass the following day. An endoscopic gastrointestinal anastomosis stapler resulted in a margin-negative complete resection of what was later determined to be a high-grade synovial cell sarcoma (Figures 3–7).

The rectal mass was removed using 2 surgical procedures. The first was an endoscopic biopsy and consisted of 3 irregular segments of tan-yellow soft tissue that were slightly lobulated and focally lined with granular pale-tan eroded mucosa. These 3 fragments measured 6 × 4.5 × 1.5 cm in aggregates. The next day an excisional biopsy was taken of a polypoid soft tissue mass measuring 5.2 × 4.8 × 3 cm. One side was designated as the previous resection site. The opposite side showed a nodular contour, which was smooth.

Microscopically, a spindle cell tumor was noted in the submucosa. A portion protruded into but not through the muscularis mucosa. However, the overlying mucosa was free of tumor. Focally, the mucosa was eroded with hemorrhage. Tumor cells were spindle-shaped, with a high nuclear-cytoplasmic ratio growing in solid sheets or fascicles. The nuclei had finely stippled chromatin. Cell margins were indistinct. In focal areas, increased collagen bundles were present. No calcification was noted. No epithelial or glandular component was seen. Mitotic activity was increased (4/10 high-power field). A hemangiopericytoma-like vascular pattern was noted in focal areas. Occasional large atypical cells with degenerative nuclear atypia were noted. No necrosis was seen. Based on morphology, differential diagnosis included gastrointestinal stromal tumor, leiomyosarcoma, and other high-grade sarcoma.

Special studies revealed that tumor cells were diffusely positive for TLE-1 and focally positive for AE1/3, OSCAR, and CK5/6. The tumor cells were negative for CAM5.2, CD117, desmin, SMA, S100, CD34, and myogenin. Reverse transcriptase polymerase chain reaction showed that tumor cells were positive for SYT-SSX2 synovial sarcoma fusion transcript and negative...
for SYT-SSX1 fusion transcript. This stain pattern
and the SYT-SSX2–positive oncogene supported the
diagnosis of monomorphic synovial cell sarcoma.

The patient’s medical history consisted of meta-
static hormone-positive breast cancer to the bone.
That cancer had been stable and treated for approxi-
mately 10 years. She had periodic bone scans, PET
scans, blood tests, and other routine surveillance
for her breast cancer, but never had an endoscopic
evaluation of her bowel until her symptoms prompt-
ed this workup. Subsequent postresection staging
studies continued to show persistent active, but not
progressive, breast cancer bony disease and no addi-
tional evidence of metastatic synovial cell sarcoma.

The patient’s care and management were pre-

tented before the multidisciplinary sarcoma confer-
ence, and the decision was made to proceed with a
more thorough surgical staging and more radical on-
cologic surgical therapy for this disease. The patient
underwent a laparoscopic coloanal resection with
mesorectal excision with primary anastomosis. To
help manage her breast cancer, a bilateral oophorec-
tomy was performed concurrently. A rectal resection
was performed, which was 19 cm in length by 4 cm
in diameter. No residual tumor was seen, grossly or
microscopically. The muscularis propria was intact,
confirming tumor aroused above muscularis pro-
pria. No metastatic tumor was identified in 12 small
lymph nodes. The fallopian tubes and ovaries were
unremarkable.
After radical surgery was performed and on determining the presence of no residual disease, the sarcoma board recommended no further chemotherapy or radiation therapy (RT). The patient already had documented widely metastatic, although stable, breast cancer.

The propensity for distant and regional nodal disease and local recurrence has been historically evident with this high-risk sarcoma type.

**Discussion**

Surgical resection with adequate margins is the primary treatment for retroperitoneal and pelvic soft tissue sarcomas. Multiple retrospective studies have shown that these sarcomas may be treated with preoperative or postoperative RT. The rationale is that negative surgical margins are difficult to achieve, and therefore RT would improve resectability when administered preoperatively. However, the use of postoperative RT to address microscopic or positive margins is justified. Generally, preoperative RT is given at a lower total dose of 50 Gy in 25 fractions, and postoperative RT is given in higher total doses (60–66 Gy in 30–33 fractions). The advantage of adjuvant therapy is that the final pathology is clearly established.

However, the role of adjuvant therapy in submucosal rectal sarcoma is unclear. The use of postoperative RT at higher doses could potentially produce a greater incidence of complications and morbidity in this anatomic area. The possibility also exists that preoperative RT could downstage this surgically unstaged tumor.

The potential benefit of chemotherapy for high-grade synovial cell sarcoma is starting to become more evident with new therapies. Response rates suggest the potential benefit of chemotherapy with this disease, and therefore would prompt consideration of more complete surgical staging to clearly identify the risk/benefit ratio.

Understanding the biology and predicted behavior of this unusual soft tissue sarcoma might suggest possible favorable factors in this location. The existence of multiple natural barriers to tumor progression continued to be intact in this patient (eg, muscular mucosa, serosa, mesorectum). Therefore, a surgical approach could provide a local control ben-
benefit that would not be present in most deep retroperitoneal pelvic sarcomas. This might, in fact, represent a biologic characteristic of a similar sarcoma in the extremity that was treated with radical amputation, in the sense that appropriate natural anatomic barriers could safely be resected with minimal morbidity and mortality. Moreover, for patients who are not candidates for curative treatments, surgical resection is an important option for palliation of symptoms.  

Although synovial cell sarcoma has been reported in almost every organ and location of the body, no case of submucosal rectal presentation has been documented to date. The biologic behavior of this sarcoma in this location prompts a thorough and careful understanding of high-grade synovial cell sarcoma behavior and the integration of a multidisciplinary plan and approach.

References  