Leiomyosarcoma of the rectum: Report of two cases

BERNA ÖKSÜZOĞLU¹, NİLÜFER GÜLER¹, AYŞEGUL ÜNER², ERHAN HAMALOĞLU³

Departments of ¹Medical Oncology and ²Pathology, Hacettepe University Institute of Oncology, ³Department of General Surgery, Hacettepe University Faculty of Medicine, Ankara-Turkey

Rectal leiomyosarcomas are rare neoplasms with poor prognosis. Currently surgery is considered the treatment of choice, however, the role of adjuvant chemotherapy and radiotherapy are still under investigation. In this report, two cases of progressive rectal leiomyosarcoma despite therapeutic efforts, are presented. [Turk J Cancer 2001;31(3):131-134]

Key words: Rectum, leiomyosarcoma, chemotherapy

Rectal leiomyosarcomas are uncommon neoplasms, accounting for fewer than 1% of all malignancies of colon and rectum (1). Besides its rarity, optimal treatment of rectal leiomyosarcoma is still not clear and the prognosis is poor. Herein we report two cases of rectal leiomyosarcomas with unusual presentation.

Case 1

A 43-year old woman was admitted to the obstetrics and gynecology department with a complaint of inguinal pain. Pelvic examination, transvaginal ultrasonography and abdominal computed tomography revealed a left 75x60 mm. adnexial cystic mass and another 5 cm solid mass in the Douglas pouch. Serum CA-125, CEA and CA 19-9 concentrations were within normal limits. In the rectosigmoidoscopy, mucosa appeared normal, however there was a narrowing of the lumen at 5 cm due to a possible extrinsic mass was noted. Total abdominal hysterectomy and bilateral salpingooopherectomy was performed and the pathological examination revealed bilateral ovarian follicular cysts, a hemorrhagic corpus luteum in the left ovary, chronic cervicitis and proliferative endometrium. One month after the operation, a 2-3 cm mass was palpable in the rectal examination but the patient refused another intervention. Six months later, she was admitted with the complaints of rectal pain and tenderness. Rectal examination revealed a 5 cm mass, 7 cm from the anal canal. It was excised and the macroscopic evaluation showed a well delineated mass in the rectal wall without mucosal involvement. The cut surface had a whorled appearance. The tumor was firm, with no apparent foci of necrosis or hemorrhage. The tumor was composed of long spindle cells with pale
eosinophilic cytoplasm. The tumor cells were arranged in whorls and fascicles with rare mitotic figures (2-3/50 HPF). One year later, a polyoid mass on the right wall of the rectum at 6 cm was identified and reexcised. The pathological examination revealed a 2.5 cm mass which showed histologically leiomyosarcoma. Six months later, she was readmitted with the complaints of rectal tenderness and bleeding and an abdominoperineal resection was performed. Pathological examination showed a 5 cm mass in the wall of rectum with morphological features of leiomyosarcoma. Thoracic computed tomography was normal and adjuvant radiotherapy and thereafter adjuvant chemotherapy including anthracycline and ifosfamide was started. After two courses of chemotherapy, she had a pelvic recurrence which was reexcised.

Case 2

A 64 year-old man was admitted with the complaint of chest pain. A 7x7 cm mass in the left lung lower lobe, destructing the ribs was identified on thoracic computed tomography. A capsulated mass was excised with the involved parts of the 6, 7 and 8th ribs. Pathological examination revealed a malignant mesenchymal tumor, with vascular invasion and necrosis which was vimentin (+), cytokeratin (-), S-100 (-). Postoperatively, adjuvant radiotherapy and 6 courses of combination chemotherapy including anthracycline and ifosfamide, was administered. Two years after the operation, a solitary 20 mm nodule appeared in the left lower lobe of the lung. Left lower lobectomy was performed. The pathological examination showed a metastatic malignant mesenchymal tumor. After 4 months of metastasectomy, a new nodule appeared in the left upper lobe. A second metastasectomy procedure was not advised by the cardiovascular surgery department. He was treated with salvage chemotherapy including vincristine, actinomycin and cyclophosphamide. Since the metastatic nodules progressed, the treatment was stopped. After 1.5 years of metastasectomy, and 3.5 years after the initial presentation, he was admitted with the complaints of rectal bleeding and tenesmus. In digital rectal examination and rectosigmoidoscopy; a 7-8 cm polypoid mass at the rectosigmoid junction was found and a low anterior resection (LAR) was performed. Pathological examination revealed a malignant mesenchymal tumor composed of pleiomorphic cells with prominent mitotic activity and areas of necrosis. Focal area of epitheloid differentiation was also seen. Mucosa was also involved. A high grade sarcoma consistent with leiomyosarcoma was diagnosed. The histological properties of this tumor was different from the tumor excised from the thoracic wall, hence this was accepted as second primary sarcoma. Four months after LAR, widespread dissemination of the tumor with multiple metastasis to the liver and local recurrence in the pelvis was observed. The patient rapidly deteriorated and died within a month.

Discussion

Leiomyosarcomas are rare tumors of the rectal wall. Colorectal leiomyosarcomas have a predilection for rectum and sigmoid colon and are commonly associated with rectal bleeding and pain (2). Evans (3) reviewed 56
cases of smooth muscle neoplasm of gastrointestinal tract followed for a minimum of 10 years; only 4 of them was located in the rectum. Randleman et al (4) reported a series of 22 cases of rectoanal leiomyosarcoma seen over 35 years and Walsh et al (5) reported a series of 48 anorectal leiomyosarcoma in 31 years.

Symptoms seen frequently included changes in bowel habit, rectal pain, pressure, and bleeding. One of our patient was admitted with the complaints of rectal pain and the other with rectal bleeding. Rectum leiomyosarcoma can originate from muscularis mucosa, muscularis propria, or the wall of blood vessels. They may grow into the lumen or into the perirectal tissues (6,7). The mucosa is involved in about 30-50% of the cases (6). Since mucosa is uninvolved in our first patient, a gynecological pathology was initially considered until rectal symptoms became apparent. Lymphatic metastases are rare. Blood-borne metastases to the liver and lungs are the most common cause of death (6).

Optimal treatment of rectum leiomyosarcoma is unclear. Since high grade and/or large leiomyosarcoma (>5 cm) tend to have a worse prognosis, conservative surgical approaches such as wide local excision appear to be insufficient. In small (≤3 cm), and localised (≤T2) tumors with low grade histological appearance, conservative surgery may be sufficient (8). By the year 1994, Khalifa et al (9) reviewed 135 cases of rectum leiomyosarcoma from the literature and reported a recurrence rate of 67.5% after local excision and suggested that the abdominoperineal resection should be the procedure of choice for rectal leiomyosarcomas. Besides that, some authors claim wide local excision would be enough for lesions <2.5 cm which are localised to the intestinal wall (1). Transrectal ultrasonography would be helpful when conservative surgery is planned to define the extent of disease (1,8). In our first patient, conservative surgery in the form of wide local excision did not prevent local recurrence. Furthermore, adjuvant radiotherapy and chemotherapy following abdominoperineal resection, did not prevent local pelvic recurrence in this patient. Although some authors claim that postoperative radiotherapy is beneficial (10), others suggest adjuvant chemotherapy and radiotherapy should be given only in the context of a clinical trial (7,8,11).

There is no definitive treatment strategy for rectal leiomyosarcomas, but surgery is the main treatment modality. As other sarcomas, leiomyosarcomas are chemoresistant and radioresistant so the efficacy of adjuvant treatment is unclear. The prognosis of rectal leiomyosarcoma is poor and the survival rates after radical surgical treatment ranges between six months to six years (6). Since five year survival rate is only 20%, more effective adjuvant chemotherapeutic regimens should be evaluated postoperatively (7).

References