Case report

Complete remission after abdominoperineal resection with posterior vaginectomy for repeated recurrent low-grade leiomyosarcoma of the vagina

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ABSTRACT

Vaginal leiomyosarcoma is a very rare malignant lesion of the female genital tract accounting for less than 1% of all vaginal malignancies. Surgery remains the primary treatment in prolonging the overall survival. We report on an unusual recurrent low-grade leiomyosarcoma of the vagina in a 51-year-old woman who underwent four surgical resections over a 7-year period. Optimal debulking was performed after her third recurrence and the patient remained free of recurrence 6 years after the fourth surgery. Extensive and aggressive resection can improve the prognosis of selected patients with local recurrence of vaginal low-grade leiomyosarcoma and can be considered a treatment option.

1. Introduction

Vaginal malignancy is a rare disease, accounting for about 3% of all malignant neoplasms of the female genital tract.1 Leiomyosarcoma accounts for only a small proportion of vaginal cancer and only about 140 cases have been reported in the literature.2-4 There is no consensus on the treatment of vaginal leiomyosarcoma; wide surgical excision with or without radiotherapy is the most common treatment modality.2,5

We report on a case of repeated recurrent low-grade leiomyosarcoma of the posterior vaginal wall treated by wide local excision. The lesion was successfully controlled after the fourth resection.

2. Case report

A 51-year-old Taiwanese woman, gravida 2 para 2, presented in 2000 with a lump in the vagina for 2 months. Gynecological pelvic examination revealed a 5 × 4 cm firm tumor arising from the posterior wall of the lower third of the vagina which completely obscured the vagina. A Pap smear showed normal results. There was no parametrial or rectal involvement on bimanual and rectovaginal examination. The patient had no medical complications and no significant family history of malignancy. She underwent a wide local excision and cellular leiomyoma was diagnosed.

In 2004, a recurrent 3 × 2 cm vaginal tumor was found adjacent to the original resection scar. Wide local excision was repeated and the pathologic findings showed low-grade leiomyosarcoma. Six months later, she developed a 2 × 2 cm recurrent vaginal mass in the same area. She underwent a third excision procedure with histologic findings demonstrating a low-grade leiomyosarcoma.

Three years after the third surgery, a 7 × 4 cm recurrent tumor was found in the same area with extension to the perineum and rectum. Magnetic resonance imaging (MRI) of the abdomen and pelvis revealed a 7 × 4 cm heterogeneously enhancing mass arising from the posterior wall of the lower vagina and extending to the...
submucosa of the rectum and perineum (Fig. 1). Her chest radiography was normal. The patient was counseled to have a repeat wide local excision, with or without radiation, or abdominoperineal resection. The patient requested the most definitive operation with the lowest rate of recurrence. In 2007, she underwent a major operation including abdominoperineal resection with end colostomy, total hysterectomy, posterior vaginectomy, and regional lymph node dissection including those in the perirectal, inferior mesenteric and inguinal areas. The surgical specimen consisted of a

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Fig. 1. Magnetic resonance imaging (MRI) findings at the time of the fourth operation. A 7 × 4 cm heterogeneously enhancing mass lies between the vagina and rectum interspace with perineal extension. (A) T1-weighted sagittal MRI, (B) T1-weighted axial MRI.

Fig. 2. Gross appearance of the vaginal tumor. The neoplasm involves both the vaginal cuff and rectal wall but not the rectal mucosa.

Fig. 3. Photomicrographs of the vaginal tumor. (A) A cellular spindle cell tumor is noted in the submucosa of the rectum. (H&E, ×40). (B) The tumor consists of proliferation of spindle cells containing hyperchromatic elongated nuclei. Mitotic figures are seen (Arrow). (H&E, ×400).
7 × 4 cm protruding firm tumor involving both the vaginal cuff and rectal wall. The resection margin in the perineum was 1.0 cm from the vaginal tumor (Fig. 2). Histologic examination of the protruding vaginal wall tumor showed that it was composed of closely packed interlacing fascicles of spindle cells with slightly enlarged hyperchromatic and pleomorphic nuclei, frequent abnormal mitotic figures (>10 per 10 HPF on average), and absence of definite tumor necrosis (Fig. 3). The tumor invaded the submucosa of the rectum. The section margins of the perineum and perianal skin were all free of disease. A recurrent low-grade leiomyosarcoma of the vagina with extension to the submucosa of the rectum was considered. The body of the uterus and both ovaries were not involved. All lymph nodes were negative. The final pathologic diagnosis of the tumor was International Federation of Gynecology and Obstetrics Stage II vaginal low-grade leiomyosarcoma. No adjuvant treatment was given and she was disease-free six years following the fourth definitive surgery.

3. Discussion

Treatment of patients with vaginal leiomyosarcoma is challenging because of the rarity of this disease and a lack of controlled clinical trials comparing management strategies. At this time primary surgery remains the mainstay of management of vaginal leiomyosarcoma. Treatment plans should be individualized depending on the location, size, and clinical stage of the tumor. Different surgical methods such as wide local excision, radical surgery (total vaginectomy with or without vulvectomy), and pelvic exenteration have been described.

In our case, a recurrent vaginal tumor occurred 3 years after the third surgery. High local recurrence rates are in part due to incompletely removed tumors or positive microscopic margins. In a study of 2084 patients with primary soft tissue sarcoma, Stojadinovic et al reported that the presence of a positive microscopic margin nearly doubled the risk of subsequent local failure at the primary tumor site. Tailored aggressive surgery is therefore needed. A preoperative workup with MRI or computed tomography of the pelvic and abdominal extent of lesions is very important. The surgical choice depends not only on the size of the vaginal tumor but also on the location. Our patient had a 7-cm soft mass in the lower third of the posterior vagina adjacent to the perineum with extension to the lower rectum. An abdominoperineal resection with disease-free margins is the optimal treatment to prevent local recurrence.

Adjuvant radiotherapy has been shown to improve local control and decrease the local recurrence rate, but not to improve overall survival in patients with soft tissue sarcomas. In a randomized trial of early stage (stage I or II) uterine sarcomas, the use of adjuvant radiotherapy had no impact on survival outcomes for women with early stage uterine leiomyosarcoma. However, the role of radiotherapy in the management of vaginal leiomyosarcoma is not clear. Therefore, adjuvant radiotherapy is not recommended routinely.

In summary, we report on an uncommon case of recurrent low-grade leiomyosarcoma of the vagina which was treated successfully by radical surgery. Patients with vaginal low-grade leiomyosarcomas close to the dentate line of the lower rectum can be treated by abdominoperineal resection. Extensive resection with disease-free margins is the optimal treatment to prevent local recurrence.

Conflict of interest

The authors declare no conflicts of interest.

References