

Case report

Successful robotic surgery for primary resection of a vaginal leiomyosarcoma: A case report

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A B S T R A C T

Primary vaginal leiomyosarcoma (LMS) is a rare entity with limited data on optimal treatment approach. Most previously reported cases utilize an open or transvaginal surgical approach for primary tumor resection. Minimally invasive surgery is an important tool in complex pelvic surgery and the limits of its utility continue to expand. Here, we report a rare case of an 11.7 cm primary vaginal LMS in a 45-year-old female that was successfully resected with a robotic approach. Our case demonstrates an innovative use of the robot and the feasibility and efficacy of this approach for primary resection of large vaginal tumors.

1. Introduction

Primary vaginal malignancies are rare and estimated to comprise just 1.3% of all gynecologic malignancies worldwide (Bray et al., 2018). The vast majority of these are squamous in histology, with primary vaginal leiomyosarcoma (LMS) representing a mere 2% of all vaginal neoplasms (Khosla et al., 2014). Surgical resection of LMS is the preferred mode of treatment, though the optimal route of surgery for vaginal tumors is not known (Ciaravino et al., 2000). Minimally invasive surgery has become an important tool in gynecologic oncology and its indications continue to be explored. Here, we report a case of a primary vaginal LMS that was successfully resected with a minimally invasive robotic approach. Written informed consent was obtained from the patient for publication of this case report and the accompanying images.

2. Case summary

A 45 year old gravida 0 presented to a primary care clinic with three months of clear, malodorous vaginal discharge and a two week history of voiding dysfunction. Her medical and surgical history was significant for uterine leiomyoma, morbid obesity (BMI 41), and a prior loop electrosurgical excision procedure. She had not been sexually active for more than five years.

On bimanual exam, a 10 cm fungating, necrotic mass was noted in the vagina. She was referred to a gynecologist where biopsies were obtained. Pathology was consistent with high grade malignant spindle cell neoplasm, favoring LMS. She was then urgently referred to a gynecologic oncologist for further evaluation and treatment planning.

MRI of the pelvis revealed a $7.6 \times 8.6 \times 10.2$ cm heterogenous, moderately T2 hypointense mass arising from the posterior cervix or posterior vagina. The mass did not appear to extend through the posterior wall of the vagina (Fig. 1). Along the left pelvic sidewall there were mildly prominent lymph nodes (largest 1.2×0.9 cm) concerning for nodal metastases. Computed tomography scan of the chest, abdomen and pelvis was consistent with the MRI of the pelvis and also showed scattered nonspecific sub centimeter pulmonary nodules and numerous benign-appearing hypodensities of the liver. After examining the patient and reviewing the imaging, primary resection utilizing the *DaVinci Xi* system was recommended in the interest of improving perioperative morbidity compared with an open approach.

The patient was taken to the operating room by a board-certified gynecologic oncologist for a robotic-assisted radical hysterectomy with upper vaginal tumor resection, bilateral salpingo-oophorectomy, left pelvic lymph node dissection, and cystoscopy. Bimanual exam revealed a massive vaginal tumor that was approximately 10 cm in size and seemed to be arising from the left posterior cervix versus upper vagina. The rectum was free of palpable tumor. On laparoscopy, the upper abdominal survey was normal. One pedunculated leiomyoma arose from the uterine fundus. The ovaries were densely adherent to the pelvic sidewalls and posterior cul-du-sac, with areas of old endometriosis noted. The mass itself was arising from the left vaginal fornix with no apparent involvement of any underlying submucosal tissue, rectum or bladder. The *DaVinci Xi* system was used to perform a radical hysterectomy. The uterine arteries were sacrificed at the level of the ureters and ureterolysis was performed to the level of the bladder. The vesicovaginal and rectovaginal planes were well developed, with the bladder and the rectum both dissected free from the vagina

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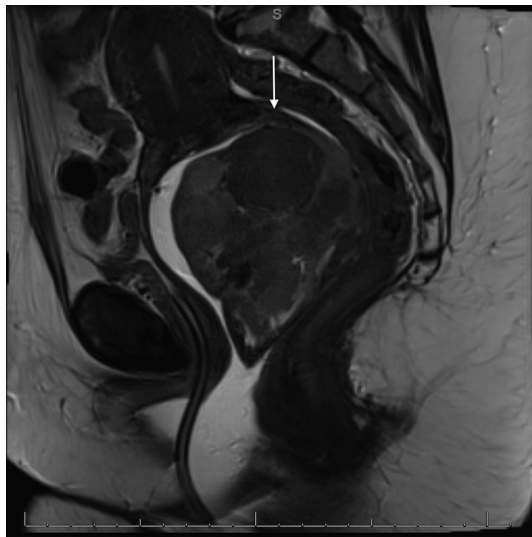


Fig. 1. Magnetic resonance imaging of the pelvis, sagittal view. White arrow points to vaginal tumor.

approximately five centimeters inferior to the cervico-vaginal junction. Placement of a uterine manipulator was not feasible due to the size and location of the mass, so ring forceps were used to delineate the vaginal anatomy. Cautery was used to create the anterior colpotomy over the ring forceps. Using cautery this incision was carried circumferentially, first on the right-hand side, then posteriorly and finally on the left hand side. The tumor appeared to arise from the left upper vagina and therefore this area had to be further dissected free from the rectum an additional two centimeters. It required complex visualization and manipulation, including use of a 30° camera and placement of an end-to-end anastomosis sizer in the rectum in order to finally release the tumor from the remaining portion of the vagina. The tumor was removed through the patient's vagina prior to closing the vaginal cuff. Given enlarged lymph nodes on imaging, a left pelvic lymphadenectomy was performed.

The total operative time was 353 min, with an estimated blood of 200 ml. Following the procedure, the patient was admitted to the hospital due to an episode of symptomatic hypotension that responded to fluid resuscitation. She was discharged on post-operative day 1. There were no postoperative complications.

Final pathology showed an 11.7 cm tumor arising rising from the posterior aspect of the upper vagina with no gross evidence of invasion into the cervix or surrounding tissues (Fig. 2). Histologically, the tumor had a fascicular growth pattern with pushing borders and was composed of spindle cells with eosinophilic fibrillary cytoplasm. The nuclei were cigar-shaped with moderate to severe atypia. Several areas of tumor coagulative necrosis were present. The tumor was highly mitotically active with up to 23 mitosis/10HPF, many of which were atypical (Fig. 3). All the findings support the diagnosis of high grade primary vaginal leiomyosarcoma. The surgical margins were negative for malignancy, and there was no evidence of spread to the lymph nodes.

Following surgery, the patient received 4 cycles of adjuvant gemcitabine and docetaxel. Unfortunately, approximately five months after primary tumor resection, computed tomography scan showed multiple new lung lesions concerning for metastatic disease. Biopsy of the pulmonary lesion was consistent with recurrent leiomyosarcoma. The patient has since undergone multiple regimens of systemic therapy with pulmonary disease progression. She has not had local recurrence. At 28 months following presentation, the patient transitioned to hospice care.

3. Discussion

Vaginal LMS is a rare entity and there are still no explicit guidelines



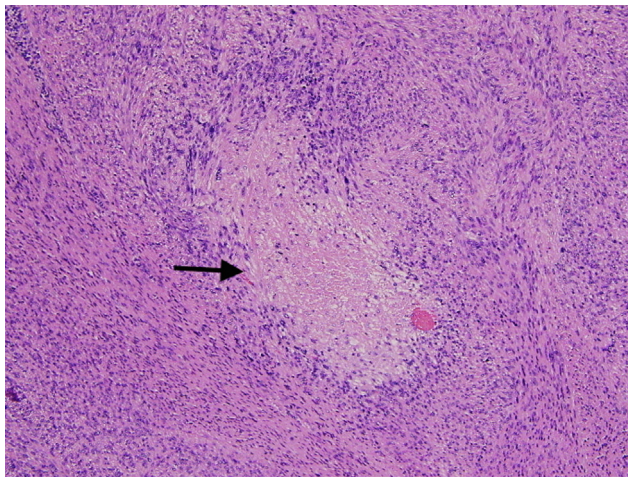
Fig. 2. Radical hysterectomy and upper vaginal tumor resection specimen. Blue arrow points to endometrium. Red arrow points to uterine cervix. Yellow arrow shows large mass arising from the posterior vagina. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

on optimal treatment. We have presented a case demonstrating an innovative use of the robot in the surgical resection of a large vaginal LMS.

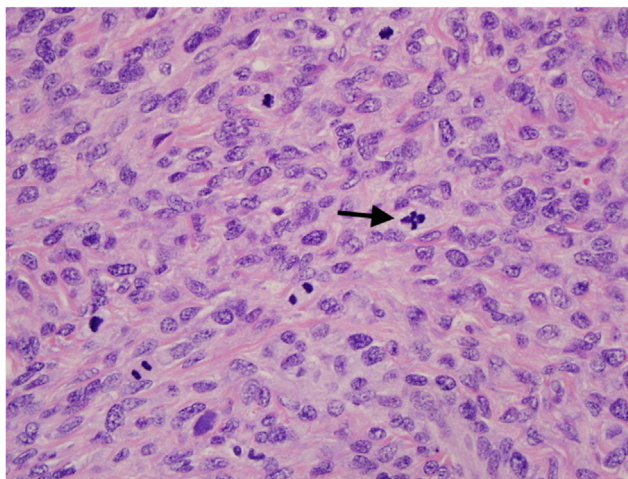
The average age at diagnosis for vaginal LMS is roughly 50 years, though cases have been reported in patients from age 21 to 86 (Khosla et al., 2014; Ciaravino et al., 2000; Peters et al., 1985). These malignancies most commonly present as asymptomatic masses, though vaginal discharge or dyspareunia may be present (Ciaravino et al., 2000; Peters et al., 1985). Vaginal LMS occur most frequently in the posterior wall and can be exophytic or intramural. These tumors can be large with no or minimal invasion into the surrounding tissue, as in the case of our patient (Ciaravino et al., 2000). The FIGO staging for vaginal carcinoma focuses on the size and depth of invasion, however, there is no formal staging established for vaginal LMS. The histologic appearances are similar to those encountered in the uterus. The criteria used to diagnose smooth muscle tumors include the presence of moderate to marked cytologic atypia, increased mitotic activity (at least 5 mitosis/10 HPF), coagulative tumor necrosis and infiltrative margins.

The data to guide treatment is limited. Primary surgical management remains the mainstay of treatment (Khosla et al., 2014). In a review article of 48 patients with primary vaginal leiomyosarcoma, the 5 year overall survival of all patients was 43%. Of those undergoing primary surgical management, the 5 year overall survival was 57%, which was significantly greater than those undergoing primary chemotherapy or radiation treatment ($p = .0006$). There was no survival benefit when chemotherapy or radiation was added to surgery (Ciaravino et al., 2000). Interestingly, in a case series of 17 patients with this disease, 3 patients who underwent pelvic exenteration were the only patients who had long term survival (Peters et al., 1985). Ciaravino supported that this is a reasonable recommendation for those able to tolerate such a morbid surgery. However, as in the case of our patient, vaginal leiomyosarcoma can recur with distant metastases in the absence of local recurrence (Anderson and Bodurka, 2008). One case report of an isolated recurrence of vaginal LMS described the management of pulmonary metastases with chemotherapy followed by thoracotomy. The patient was alive with no evidence of disease 2 years later (Anderson and Bodurka, 2008).

Minimally invasive surgery has an important role in gynecologic oncology. In 2012, the LAP2 study demonstrated an improved safety profile in patients who underwent a minimally invasive approach for comprehensive surgical staging of uterine cancer, without compromising progression free or overall 5-year survival (Walker et al., 2012).



(a)



(b)

Fig. 3. (A) Tumor shows fascicular growth pattern and is composed of spindle cells with eosinophilic fibrillary cytoplasm and cigar shaped nuclei with moderate to severe atypia. Black arrow pointing to area of coagulative necrosis. (H&E, 10 $\times$). (B) The tumor contains frequent mitoses with black arrow showing atypical mitotic figure. (H&E, 40 $\times$).

As a result of this study, a minimally invasive approach has been the preferred method for endometrial cancer. A similar shift has been noted for the management of early stage cervical cancer, although this approach has recently been scrutinized as shorter disease free and overall survival rates were noted in patients undergoing minimally invasive surgery compared to laparotomy (Ramirez et al., 2018). Data is emerging on the feasibility of minimally invasive ovarian cancer surgery, although long term outcomes are not well understood (Cardenas-Goicoechea et al., 2019).

Surgical approach has not been investigated in vaginal LMS. The limited number of cases prevent meaningful survival analysis based on surgical approach. Given the need for enhanced deep pelvic visualization, as well as a desire to minimize postoperative complications, robotic surgery was utilized in our case.

Unfortunately, our patient's clinical course has been consistent with

other cases of vaginal LMS. Because of the rare nature of this aggressive disease, optimal management guidelines may never be established. As of now, surgery remains the recommended primary treatment. Our case demonstrates that a robotic surgical approach for the primary resection of vaginal LMS is feasible, effective, and safe.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Disclosures

None.

Author contribution statement

We acknowledge all authors have contributed to this paper. Dr. Erickson was involved in the care and treatment of this patient. Drs. Hagen, Wilhite, Erickson, and Tarbunova were involved in the literature review and writing of the case report.

Declaration of Competing Interest

The authors have no conflicts of interest to disclose.

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