Vaginectomy and Buccal Mucosa Vaginoplasty as Local Therapy for Pediatric Vaginal Rhabdomyosarcoma

Rodrigo L. P. Romao and Armando J. Lorenzo

We report a case of vaginal rhabdomyosarcoma where vaginectomy with buccal mucosa vaginoplasty was performed to avoid radiation therapy to the young pelvis.

The patient presented at 30 months with an exophytic vaginal mass, found to be botryoid rhabdomyosarcoma. After receiving neoadjuvant vincristine, actynomycin D, and cyclophosphamide chemotherapy with good response, she underwent surgery. It was performed using an anterior sagittal approach on the prone position, which allowed for a safe circumferential dissection of the vagina all the way to the cervix and en bloc resection. Two buccal mucosa grafts were used for vaginoplasty. Pathology revealed negative margins. The patient completed therapy in October 2014 and remains disease-free. UROLOGY 102: 222–224, 2017. © 2017 Elsevier Inc.

Despite portraying excellent overall survival rates, local control in vaginal rhabdomyosarcoma (RMS) represents a conundrum. Radical mutilating surgeries performed in the 1970s, such as pan (radical) hysterectomy, led to prohibitive permanent sexual and reproductive impairment and have thus been abandoned[1]; the preferred method of local control in recent years has been radiation therapy. Notwithstanding the benefit of organ preservation, radiation also carries varying degrees of morbidity to all exposed organs contained in the female pelvis (pelvic bone, bladder, uterus, vagina, and rectum), with clinical significance being inversely proportional to the patient's age.

Recent multicentric cooperative trials under the auspices of the Children's Oncology Group have attempted to omit radiation in low-risk patients with great chemotherapy response, only to reveal high rates of local recurrence (~50%).[2,3] It becomes clear that the development of a better local control strategy for this disease is desirable, particularly for younger girls (<3 years of age).

Herein we report a case of vaginal RMS where local control was achieved through a subtotal vaginectomy through an anterior sagittal approach. Vaginal reconstruction was performed using a buccal mucosa graft. The surgical strategy was chosen with the deliberate goal of avoiding radiation to the young pelvis.

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CASE REPORT

A 30-month old girl presented to the emergency department with an exophitic vaginal mass noted by her mother. She was otherwise well. A vaginoscopy with biopsy confirmed the diagnosis of embryonal (fusion negative) RMS of the botryoid variant. Staging did not reveal any other suspicious areas of disease. The patient was classified as having a favorable site, stage I, group III, low-risk RMS and was treated with vincristine, actynomycin D, and cyclophosphamide chemotherapy as per D9803 protocol.

Repeat vaginoscopy at 12 weeks showed marked improvement; however, abnormal areas could still be noted on the distal aspect of both the anterior and posterior vaginal walls. Biopsy of those areas confirmed the presence of rhabdomyoblasts only. After extensive multidisciplinary discussion at our tumor board meeting, we decided to proceed with a surgical strategy for local control. The team agreed that if a negative margin surgical resection was achieved, radiation would be omitted from this girl's initial treatment plan.

At 24 weeks, she underwent a subtotal vaginectomy using an anterior sagittal approach. This approach is similar to the anterior sagittal trans-rectal approach procedure described for vaginoplasty in congenital adrenal hyperplasia with a high vagina[4,5] and to the one used for the surgical management of cloacal anomalies.[6] The patient was positioned prone in a modified jackknife position. A midline sagittal incision was performed from the rectum to the vagina, extending circumferentially around the latter. The rectal wall was not incised, and dissection was carried down in the plane between the rectum and the vagina initially and then circumferentially to the level of the cervix (Fig. 1). The plane between urethra and vagina was the most tenuous to identify, and having a Foley catheter inside each structure aided in separating them adequately.

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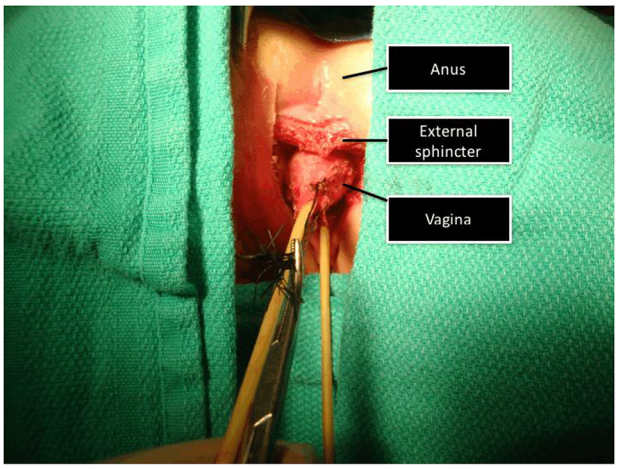

Figure 1. Patient is in the prone position and circumferential dissection around the vagina has been completed. Key structures are identified and 2 Foley catheters can be seen, 1 in the vagina and the other in the urethra. (Color version available online.)

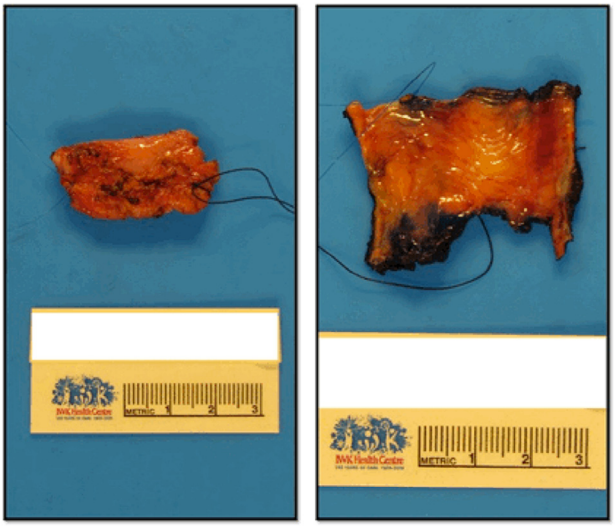

Figure 2. En bloc vaginectomy specimen sent to pathology. (Color version available online.)

The vagina was resected en bloc (Fig. 2), and only a minimal amount of mucosa was spared at the dome (fornix) to allow reconstruction. The cervix was also preserved. Anterior, posterior, proximal, and distal margins were carefully identified with different suture materials. Intraoperative frozen section confirmed that margins were negative. Two buccal mucosa grafts were harvested at the beginning of the case from each cheek (approximately 4 × 1 cm) as described previously for urethral reconstruction.[7,8] Donor sites were left open. Each graft was laid longitudinally (on a posterior and anterior orientation) and sewn initially to the vaginal dome (forniceal) mucosa and then to the introitus using 5-0 polyglactin sutures. Finally, the grafts were sewn to each other to reconfigure the lateral vaginal walls, including adjacent tissues for better fixation (Fig. 3). A 24 Fr pediatric chest tube was sutured in place as a vaginal stent and removed on postoperative day (POD) 9.

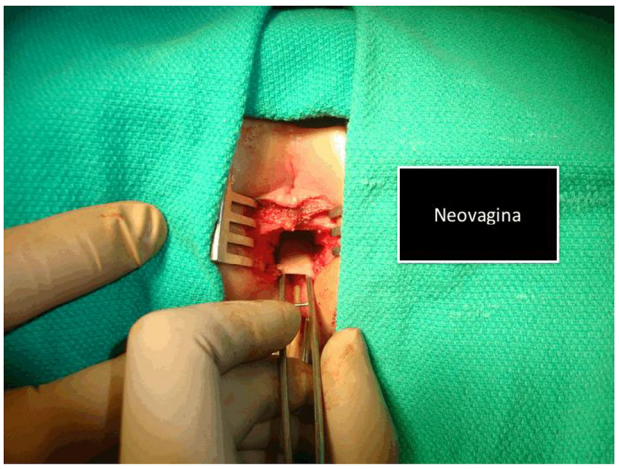

Figure 3. Final aspect of the buccal mucosa vaginoplasty. (Color version available online.)

The patient tolerated the procedure well and was discharged on POD 3. She had prolonged constipation postoperatively and developed a superficial dehiscence of the perineal body on POD 15, which related to passage of a large amount of hard stool. She was re-admitted and an examination under anesthesia was performed at the time; there was no evidence of deep-space infection or fistulas. She was kept on oral antibiotics and the area healed well without complications.

Chemotherapy was concluded, and the patient underwent an end-of-treatment vaginoscopy, which revealed a patent vagina and no evidence of recurrent disease. At 34 months of follow-up, the patient has been disease free based on magnetic resonance imaging of the pelvis and examination under anesthesia performed concomitantly.

DISCUSSION

Female genital tract RMS is a rare disease, accounting for less than 5% of all RMS.[2] About half of those cases affect the vagina. Adequate local control is paramount in the treatment of these patients, as illustrated by the high recurrence rates observed in patients treated with chemotherapy alone as detailed in the beginning of the paper. Nonetheless, overall survival was excellent in those patients where radiation was spared initially, implying that salvage strategies exist and are reassuringly effective in rescuing patients who developed a local recurrence.[2] Hence the rationale for the surgical strategy described here.

The current trend in local control for genitourinary RMS revolves around surgery with organ preservation and radiation when the latter is not achievable. Nonetheless, the negative effects of radiation on the young female pelvis are irrefutable. Historically, surgical options described for these patients included pelvic exenteration and panhysterectomy,[1] which are clearly too extreme and unacceptable. Vaginal

reconstruction has been plagued by technical challenges, such as severe strictures with grafts and flap techniques and the need for laparotomy and other morbidities associated with bowel vaginoplasty.

In recent decades, a variety of procedures have been added to the armamentarium of pediatric genitourinary and anorectal reconstruction. The posterior and anterior sagittal approaches offer a reliable and reproducible means of accessing the perineal structures without entering the abdominal cavity.[4,5,9] Specifically, surgical management of cloacal anomalies and congenital adrenal hyperplasia has been greatly impacted by modifications introduced by such approaches, as well as total and partial urogenital sinus mobilization.[10-12] Buccal mucosa grafts are now used routinely in both adult and pediatric urology for urethral reconstruction with acceptable results.[13,14] Recent reports have established that buccal mucosa vaginoplasty leads to good cosmetic and functional outcomes in patients with Meyer-Rokitansky-Kuster-Hauser syndrome (agenesis of müllerian structures and vagina), complete androgen insensitivity syndrome, and repair of urogenital sinus.[15-18] Grimsby and Baker provide a comprehensive overview of complications reported with different vaginoplasty techniques and report on a personal series of 7 cases of total neovagina creation using buccal mucosa grafts in a similar fashion to the one described herein. Five out of the 7 are sexually active and have no dyspareunia.[19]

In conclusion, the technique proposed herein builds on recently developed surgical strategies to offer a reasonable alternative to radiation therapy in patients with vaginal RMS. Obviously long-term follow-up is required to ensure adequate oncological and functional outcomes are attained.

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