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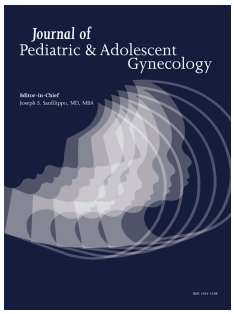

Foley Catheter As Vaginal Stent In A Toddler With Vaginal Rhabdomyosarcoma

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LH CASE REPORT

INTRODUCTION:
Sarcoma botryoides is a variant of embryonal rhabdomyosarcoma (RMS). This malignant tumor originates from muscle progenitor cells and although it is highly malignant, cure rates exceed 90%.[1] While RMS is the most common soft tissue malignancy in children, vaginal presentation is rare.[2] The average age of diagnosis of vaginal RMS is 21 months of age. Treatment consists of chemotherapy combined with local radiation and/or surgery to achieve local tumor control while preserving organ function.[1] Sequelae of vaginal radiation therapy in this age group have not been well defined. However, vaginal stenosis is a known result of local radiation therapy in adult women treated for a variety of pelvic cancers. In adults, vaginal dilation is the primary method of prevention and treatment.[3] In pre-pubertal children and toddlers, regular vaginal dilation could be a traumatic experience for both parent and child and is not a reasonable option.

A recent review article published in Obstetrics and Gynecology outlines the physiologic basis for vaginal dilation as well as recommended treatment guidelines in the prevention of vaginal stenosis in adult patients undergoing pelvic radiation. Vaginal dilation should be initiated 2-4 weeks following completion of radiation. Regular dilation for 10-20 minutes at least three times a week prevents adhesion formation and may release underlying myofascial tissue and prevent fibrosis in this layer.[4] Evidence suggests that radiation induces chronic oxidative stress, which can lead to late radiation-induced normal tissue injury. Markers for oxidative stress can be seen in affected tissues for up to 6 months following completion of radiation therapy.[5] Although difficult to quantify, in one study of adult patients undergoing radiation therapy for cervical cancer the incidence of vaginal stenosis is approximately 38 percent. [6] An older study evaluating 12 female patients who had undergone pelvic radiation therapy as children reported 5 of 12 had vaginal stenosis.[7]

CASE:
Our patient initially presented at age 18-months with bleeding and passage of small cysts from the vagina for one week. Vaginoscopy and biopsy confirmed diagnosis of vaginal RMS, subtype boytroid embryonal RMS. Patient underwent radiologic staging with whole body positron emission tomography (PET)/computed tomography (CT) scan and magnetic resonance imaging (MRI) of the primary site. She was classified with Intergroup Rhabdomyosarcoma Study Group (IRSG) Stage I/Group III embryonal RMS (PMID 18521303). Recommended systemic chemotherapy consisted of vincristine, actinomycin, and cyclophosphamide (VAC). She received a cumulative cyclophosphamide dose of 16.8 grams.

Prior to port placement and initiation of chemotherapy, our patient's family was counseled regarding ovarian preservation options. They elected to proceed with ovarian tissue freezing and unilateral oophorectomy was performed at the time of port placement. Ovarian cryopreservation is considered experimental and was performed under an IRB-approved research protocol at Children's National Medical Center.

The patient tolerated her chemotherapy well without any unexpected adverse events; and achieved complete radiographic response by PET and MRI at week 19. Vaginoscopy with biopsy was performed during week 19, and despite no residual disease seen on MRI, vaginoscopy demonstrated a pedunculated lesion in the vaginal canal. This lesion was excised and pathology confirmed residual disease. The case was reviewed through the Children's National Medical Center multidisciplinary tumor board. The decision was made for adjuvant