

AMPUTATION OF CERVIX AND NEOCERVIX CREATION USING STURMDORF TECHNIQUE IN RECURRENT CERVICAL SARCOMA BOTRYOIDES: A CASE REPORT

Pribakti Budinurdjaja¹, Ihya Ridlo Nizomy¹, Inas Tsurayya Fauziah Lahdimawan²

¹ Division of Urogynecology and Reconstruction, Department of Obstetrics and Gynecology, Faculty of Medicine Lambung Mangkurat University/ Ulin Hospital, Banjarmasin

² Residency Program, Department of Obstetrics and Gynecology, Faculty of Medicine of Lambung Mangkurat University/ Ulin Hospital, Banjarmasin

Correspondence Author: inaslahdimawan@gmail.com

Abstract:

Sarcoma botryoides, or embryonal rhabdomyosarcoma, is soft tissue sarcoma arises from embryonic muscle cells and normally present with discharge, bleeding, or a visible mass at the vaginal introitus in infants and children. Aim of this study is to report a case of cervical sarcoma botryoides. Method used is a Case report. A 16-year-old girl, nulliparous, complained abnormal vaginal bleeding and a protruding vaginal mass. Patient underwent first surgery, mass recurrent five months later. The second surgery was performed by cervical amputation and neocervix creation using Sturmdorf sutures technique. Postoperative week 12 demonstrated a healthy neocervix in appearance, her menses have resumed, indicated patency. She has no evidence of disease 3 months after diagnosis. Cervical sarcoma botryoides is a rare malignancy. Clinicians must thoroughly examine any polyp or mass protrudes from vagina. Early diagnosis and treatment are essential for better prognosis. Sarcoma botryoides has a much improved prognosis with multimodality treatment. To determine the long-term survival and future potential fertility outcome, larger study and longer follow-up are needed.

Keywords: sarcoma, botryoides, embryonal, rhabdomyosarcoma, neocervix

Introduction

Sarcoma botryoides, known as embryonal rhabdomyosarcoma, is soft tissue sarcoma arises from embryonic muscle cells.^{1,2} Rhabdomyosarcoma is the most common soft tissue tumor in children, accounting for about half of all soft tissue sarcomas and 4,5% of all childhood cancers.^{3,4} Rhabdomyosarcoma can affect any part of the body, but the most common locations are the head and neck area and the genitourinary tract. The Intergroup Rhabdomyosarcoma Study Group (IRSG) described three histologic subtypes: embryonal, alveolar, and undifferentiated.⁴ The embryonal subtype are divided into classic, botryoid, and spindle cell.⁵ Botryoid variant is responsible for the majority of cases of embryonal rhabdomyosarcoma of the cervix. Approximately 20% of rhabdomyosarcoma in children located in the genitourinary tract and rarely in cervix (0.5%).⁴ Sarcoma botryoides is highly malignant, normally present with discharge, bleeding, or a visible mass at the introitus in infants and children.¹

We report a case of sarcoma botryoides that presented as a cervical mass protruding from the vaginal introitus in an adolescent, treated by amputation of uterine cervix followed by neocervix formation. We emphasize the diagnosis and management in limited resources setting.

Case Report

A 16-year-old girl, nulliparous, referred to our centre presented with abnormal vaginal bleeding since a year ago and a mass protruding through the vaginal introitus, initially small about 3 cm diameters then enlarged over time. She had surgery in previous hospital, CT scan revealed a solid mass in pelvic cavity, uterine prolapse was initially suspected. The histopathologic evaluation initially suggested an abscess with focal atypical cells and sarcoma cannot be excluded. Five months after the first surgery,

she complained about heavy vaginal bleeding and recurrent of vaginal mass. The past medical history and family history was unremarkable.

Physical examination was obtained, the patient vitally stable. Gynecologic examination has revealed a grossly cervical mass, irregular shape with grape-like feature protruding from introitus vagina measuring 13 x 10 cm diameters (Fig. 1). Other examination within normal limit.

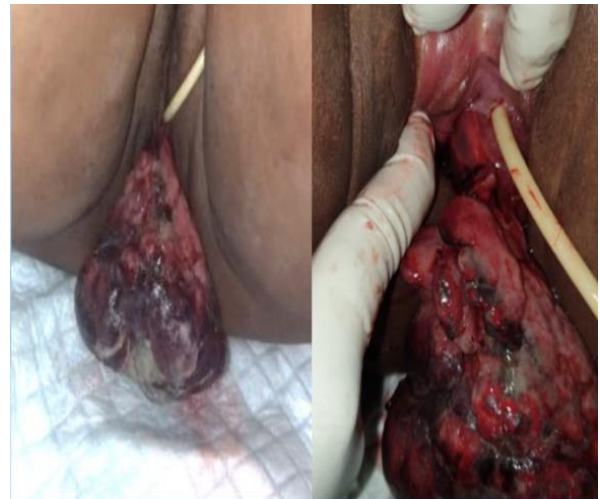


Figure 1. Preoperative, mass protruding from vaginal introitus, macroscopic appearance of the tumor "a grape like" feature

Ultrasonography imaging was obtained, uterus ante flexion with 3,23 x 1,9 x 3,5 cm in size, fundus and endometrial line was seen (Fig. 2). Due to the limited resource no MRI was available to evaluate the metastasis. The surgery was performed by cervical amputation and neocervix was formed by Sturmdorf sutures technique.

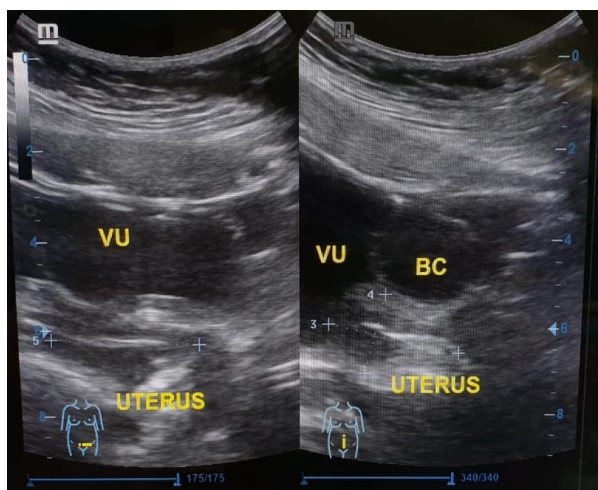


Figure 2. Ultrasonography imaging shown endometrial line of uterus

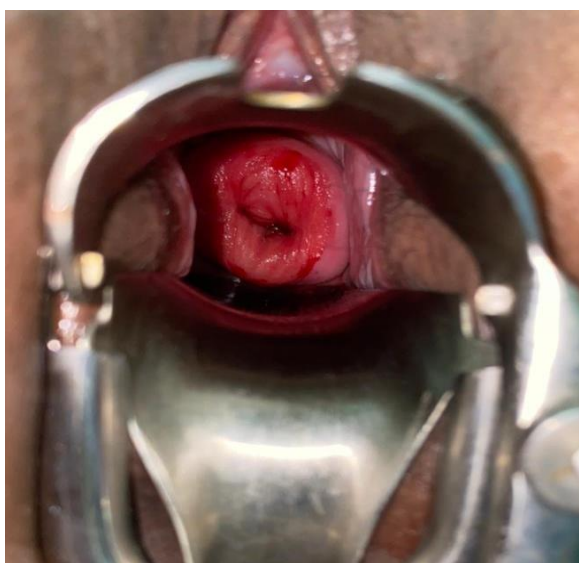


Figure 3. Postoperative, week 12, speculum examination shown healthy neocervix

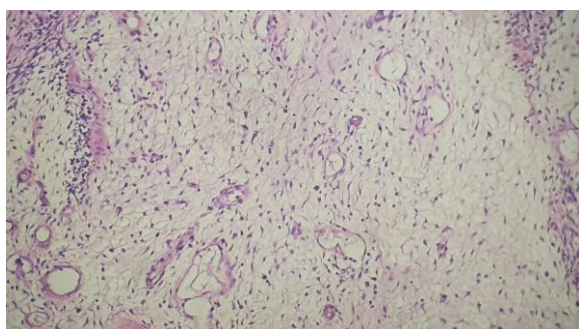


Figure 4. Histopathological finding consisted with a sarcoma botryoides (embryonal rhabdomyosarcoma)

The histopathological findings on microscopic examination, the tumor consists of cells proliferation arranged in a hypercellular area, the cell nucleus is spindle shaped cell, stellate and round, these findings were consisted with a sarcoma botryoides (embryonal rhabdomyosarcoma).

Discussion

Rhabdomyosarcoma (Greek for rhabdos, “rod”, mys “muscle”, sarkos “flesh”) is the most common soft tissue tumor among children and adolescents accounting for 40% of tumors in persons under 20 years old.² Although they may occur in any site, embryonal rhabdomyosarcoma typically arise in genitourinary tract. The botryoid subtype represents 10% of all rhabdomyosarcomas and is associated with an excellent 5-year survival rate (95 %).² Botryoid subtype occurs in hollow organs arising under the mucosal surface of body orifices such as the vagina, bladder, nasopharynx and biliar tract.² Vulvo-vaginal and uterine rhabdomyosarcoma is the most common malignancy of the pediatric female genital system.⁶

Sarcoma botryoides generally presents in the first few years of life. It may appear as abnormal vaginal bleeding, blood-tinged discharge, protruding vaginal mass, or abdominal-pelvic mass. Urinary symptoms may present especially when the tumor is anteriorly situated or tenesmus where there is posterior extension. The name botryoides originates from the Greek word “botrys,” a bunch of grapes, due to the characteristic macroscopic appearance of the tumor, may appear in gray-red colored, with hemorrhage and edema, associated with vaginal bleeding.^{2,3,6-8}

Diagnosis of sarcoma botryoides is based on histopathology and post-surgery immunohistochemistry, it may also be performed by preoperative biopsy or intraoperative frozen section. Vaginal lesions usually have embryonal or botryoid

embryonal histology and are associated with an excellent prognosis.^{2,9}

The multimodal treatment for rhabdomyosarcoma of the uterine cervix should always be given.¹⁰ In the past, exenterative surgery was used for these lesions, but survival was poor. More recently, conservative surgery has been used in conjunction with preoperative or postoperative chemotherapy and radiotherapy with significantly improved survival.¹ Standard optimal treatment is not yet established and also difficult to devise, given the rarity of disease and histologic variations.

In this case, patient underwent surgery. Considering the tumor in uterine cervix, cervical amputation was performed by circular incision of the cervix and the cervical walls are push upward, continued by incision of the tumor and cervix with 2 cm of margin. Evaluated of internal uterine ostium, uterus sounds at a depth of 5 cm. After cervix was sutured, neocervix was made by Sturmdorf technique from the middle. In 1915, Arnold Sturmdorf introduces his tracheloplasty technique. To perform the Sturmdorf technique, a mattress suture is placed both anteriorly and posteriorly to pull the lateral cervical mucosa (Fig. 5).^{11,12}

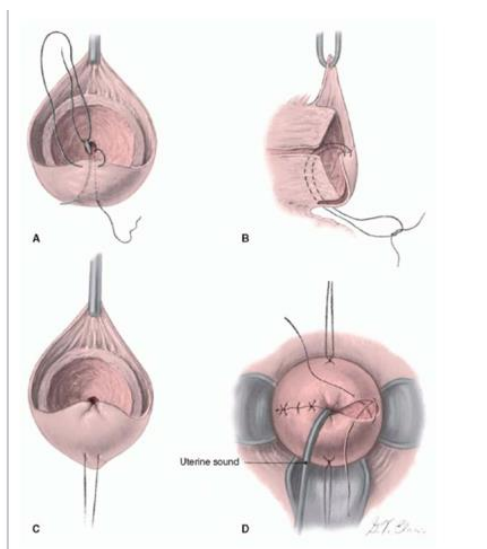


Figure 5. Sturmdorf suture technique¹²

Similar cases have been reported, one from our centre, recurrent cervical sarcoma botryoides in an infant, a wide excision of tumor was done followed by adjuvant chemotherapy.⁹ Other cases were about our patient's age, the tumor presented as cervical polyp, histological examination revealed a embryonal rhabdomyosarcoma, a polypectomy was performed and multi-agent chemotherapy administered after surgery.^{7,13}

The general management principles include biopsy and staging followed by chemotherapy as directed by pretreatment stage and clinical group. There is no role for initial management with radical surgery such as vaginectomy or hysterectomy.¹⁴ Vaginectomy and hysterectomy are performed only for persistent or recurrent disease.⁶ If unresponsive to chemotherapy, primary uterine tumors require hysterectomy with preservation of the distal vagina and ovaries. Lymph node involvement is very rare (5%) and thus, pelvic lymph node dissection is not indicated.¹⁵

There are different approaches in the surgical management of this tumor, from simple excision to extensive radical procedures.³ The use of radical trachelectomy in patients with large embryonal rhabdomyosarcoma of the cervix to achieve negative resection margins and preserve fertility have been reported.¹⁶ Surgical approaches need to place local control of disease as a primary goal. However, although minimally invasive procedures such as simple polypectomy for an exophytic, sarcoma botryoides have been reported, they carry the hazard of leaving behind residual gross or microscopic disease. Most recurrences of rhabdomyosarcoma of the female genital tract are locoregional.¹⁰ Patients are followed with routine abdominal and pelvic MRI to determine tumor response and detect recurrence.² In addition, surgical complications may include rectovaginal fistula, vesicovaginal fistula, and urinary incontinence all of which are associated with

significant morbidity.⁶ Sarcoma botryoides has a much improved prognosis with multiagent adjuvant chemotherapy than other rhabdomyosarcoma tumors when resected properly and embryonal cells are shown in histopathology examination.¹⁷

In our case, vaginal examination at postoperative week 12 demonstrated a neocervix that was pink and healthy in appearance, patient resumed in normal menstrual flow, it demonstrated the patency. Importantly, our patient had a fertility preservation treatment despite having a large cervical mass at presentation. She has no evidence of disease 3 months following diagnosis.

Conclusion

Sarcoma botryoides of cervix is a rare malignancy. Clinicians must thoroughly examine any polyp or mass protrudes from vagina. Early diagnosis and treatment are essential for better prognosis. Sarcoma botryoides has a much improved prognosis with multimodality treatment. To determine the long-term survival and future potential fertility outcome in this case, larger study and longer follow-up are needed.

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