

## A Very Rare Case of a Three-Year-Old Girl with Rhabdomyosarcoma of the Vagina

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### Abstract

### Case Report

**Background:** Botryoid rhabdomyosarcoma is a rare tumor of soft tissue. It arises almost in the vaginal or vesical wall and accounts for 3 to 4% of all cases of childhood cancer. **Case report:** we report a case of 36-months-old presented with bleeding and pelvic pain. Physical examination revealed a vaginal mass with grape-like feature protruding from the vagina. A biopsy was done and the anatomopathological exam revealed an embryonal botryoid type rhabdomyosarcoma. The staging workup including thoraco-abdomino-pelvic computed tomography scan and abdomino-pelvic magnetic resonance imaging didn't show any metastatic lesion. The girl undergone neoadjuvant chemotherapy followed by conservative surgery and adjuvant external radiotherapy. Currently, the girl is disease-free with 5 years of regular follow up. **Conclusion:** the rarity of rhabdomyosarcoma in childhood leads to a lack of randomized controlled trials to guide the appropriate treatment. The new therapies conduct to increase the lifespan in those children. Actually, we tend to conservative treatments to preserve the quality of life.

**Keywords:** Embryonal rhabdomyosarcoma, sarcoma botryoid, vaginal malignancies, children, chemotherapy, prognosis.

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## INTRODUCTION

Children's gynecologic neoplasms are very rare. Only less than 5% of all pediatric tumors arise from the gynecologic tract [1].

Rhabdomyosarcoma (RMS) is a very aggressive soft tissue sarcoma. It originates from myogenic progenitor cells [2]. It is the most common sarcoma in childhood, with approximately 3 to 4% of all cases of pediatric cancers [3].

The most frequent sites of origin were the head and neck (35-40%), followed by the genitourinary tract, trunk, extremities, retroperitoneum, and uncommon regions (e.g. intrathoracic, GI tract, perianal and anal regions) [4].

We identified five major rhabdomyosarcoma histological subtypes: embryonic, alveolar, embryonal botryoid, embryonal spindle cell, and anaplastic [5].

The most common subtype in childhood is the embryonal subtype. In the walls of hollow, mucosal lined structures such as the vagina, bladder nasopharynx, and rarely cervix and uterine fundus,

Botryoid embryonal rhabdomyosarcoma is observed [6]. It's looked like a bunch of grapes. Sarcoma botryoid has usually been found in children below the age of 8 years [6, 7].

RMS had a poor prognosis. We present a case of a young girl who had RMS of the vagina, with a good outcome.

## CASE PRESENTATION

A girl of 36 months-old with no history of medical poisoning during pregnancy and no remarkable family history, was admitted into our institute, because of vaginal bleeding.

On physical examination, the baby was so active and appeared well. We found on the physical exam a polypoid mass prolapsing through the vaginal, reminding grapes. This mass was located in the anterior lateral vaginal wall, no palpable inguinal lymph node, no hepato-splenomegaly.

The digital rectal examination found a palpable unfixed mass that pushes the rectum. Other exams were in the normal course.

A biopsy was done and the anatomopathological exam revealed an embryonal botryoid type rhabdomyosarcoma.

The thoraco-abdomino-pelvic computed tomography (CT) scan showed a mass located in the uterine cavity without any suspicious lymph node in the pelvic area and or distant metastasis. The abdomino-pelvic magnetic resonance imaging (MRI) revealed a heterogeneous mass located in the anterior lateral vaginal wall measuring 35x21x16.5 mm, extended to the distal third of the vagina and infiltrated the rectovaginal fat with extension to the parametrium.

Patient referred to pediatric oncology, where she received 9 cycles of neoadjuvant chemotherapy using ifosfamide, vincristine, and dactinomycin (IVA) followed by 4 cycles of Vincristine only.

After chemotherapy, the patient underwent an evaluation with an examination under anesthesia, where we didn't found a clinical response of tumor for chemotherapy.

The patient underwent a conservative surgery based on local excision of the mass and ovarian transposition. The histological specimen concluded to a viable RMS with involved margins.

The patient was proposed for brachytherapy, but due to the lack of experience for pediatric brachytherapy, the patient underwent external beam radiotherapy. Before radiotherapy, the patient underwent an evaluation under anesthesia, where we found a vaginal mass that we biopsied. The histological exam showed RMS. The total dose of radiotherapy was 41.4 Gy. Post-treatment MRI showed no evidence of distant disease or local recurrence. The biopsy of the vaginal scar was free of tumor. The patient stills following up regularly by clinical and radiological examination without any metastasis or recurrence with 5 years of regular follow up.

## DISCUSSION

Rhabdomyosarcoma (RMS) is a malignant mesenchymal tumor that represents the most common soft tissue tumor of childhood, and responsible for approximately one-half of all soft tissue sarcomas [8].

RMS is classified by the World Health Organization in 2013 into embryonal (including botryoid, anaplastic), alveolar RMS (including solid, anaplastic), pleomorphic and spindle cell/sclerosing RMS [5].

The exact cause of the formation of embryonal Rhabdomyosarcoma stills unknown (8). It located mostly in the neck and head region, followed by the genitourinary tract in about 3% of all the cases. Cervix

and uterus are the most frequently affected sites in adolescent but rarely in the child. It mostly occurs in young children less than three years old, the average age is around 1.8 years (0-4 years) [8,9].

The most revealing symptoms are vaginal bleeding and pelvic pain, sometimes the tumor can protrude in the vaginal canal due to the large size like in our case. Sometimes, other clinical presentation can be observed as leukorrhea and malodorous discharge, rarely the diagnosis is made in stage of metastases in less than 10%. The common sites of metastases are lungs and/or bones [8,9].

The diagnosis is not always established by the first biopsy specimen. Serum AFP can be helpful for the diagnosis, especially in the evaluation of treatment response and the detection recurrence [10].

Historically, in the 1960s, the treatment was based on vaginectomy to total pelvic exenteration because RMS is a very aggressive tumor, but this aggressive surgery is no longer used since 1970. A new tendency towards to limit surgery and to preserve both the bladder and the rectum due to various combination chemotherapy that improved survival [11].

Recently, the development of chemotherapy conducted to a conservative therapy to maintain sexual and reproductive function. So since then the multimodality therapy, including combination chemotherapy with or without radiotherapy with less radical surgery such as local excision, polypectomy, or cervical conization, was adopted and has significantly improved survival. Local excision should be considered only when complete resection can be achieved without important functional impairment [10]. In our case, we performed a local excision, but the margins were involved, so we performed external beam radiotherapy. The treatment outcomes were excellent with no local recurrence. That results lead to consider radiotherapy as a cornerstone for the treatment of RMS.

At present, the indication of pelvic exenteration in the management of patients with vaginal RMS is limited to patients with extensive local recurrence or after the failure of standard therapies [8].

RMS is sensitive to chemotherapy, which has an important role in local control, and eradication of micrometastatic. The primary therapy for small and localized RMS is radiotherapy and neoadjuvant chemotherapy based on vincristine, dactinomycin and cyclophosphamide protocol that is the most used in RMS [7,8,10].

After chemotherapy, patients should undergo an evaluation with examination under anesthesia and resection of the residual mass, or biopsy of the tumor site [10,12].

RMS has a great response to chemotherapy than 90%. The neoadjuvant chemotherapy followed by local excision resulted in a 79% disease-free survival rate [8].

## CONCLUSION

RMS in childhood is a rare and aggressive disease. The management of RMS tends to be more conservative with the combination of chemotherapy, surgery, and radiotherapy. That leads to improve the quality of life for those young patients.

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