Case Report (Olgu Sunumu)

Botyroid Rhabdomyosarcoma of the Vagina in A Two-month-old Girl

İki Aylık Bir Kız Çocuğunda Vajina Botryoid Rabdomyosarkom

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Özet

Botryoid rabdomyosarkom çocukluk çağı ve infantil dönemde en sık görülen malign mesenkimal tümördür. Bu tümör vajina, mesane, biliyer system gibi içi boş organları örten rhabdomyoblast denilen immatür iskelet sistemi hücrelerinden orijin alır. Botryoid sarkom; erken çocukluk çağı ve infantlarda vajinada, reprodüktif çağda serviksde, postmenaposal yıllarda uterus korpusta bulunur. Tedavi cerrahinin adjuvant kemoterapi ve radyoterapi ile kombinasyonudur. Biz vajinal kanama ve vajinadan prolabe olan kitle ile başvuran 2 aylık kız çocuğu olgusu bildirdik. Difüzyon Manyetik Rezonans Görüntüleme (MRG) 'de rektovajinal septumda yaklaşık 6 cm çapında kitle görüldü. Vajinadan alınan eksizyonel biyopsi botryoid rabdomyosarkom tanısını doğruladı. Biyopsi sonrası hasta çoklu kemoterapi tedavisi aldı. Hasta şuan hala hastalıksız şekilde remisyonda takip edilmektedir.

Anahtar kelimeler: Botryoid, Rabdomyosarkom, Vajina, Tümör

Abstract

Botryoid rhabdomyosarcoma is the most common malignant mesenchymal tumor of the vagina in children and infants. It is a malignant tumor that arises from immature skeletal muscle cells (i.e., rhabdomyoblasts) lining the walls of hollow organs, including the vagina, bladder, and biliary tract. This cancer most commonly affects children under the age of 4, although it can sometimes present in older individuals. Botyroid sarcoma is usually found in the vagina during infancy and early childhood, in the cervix during reproductive years, and in the corpus uteri during postmenopausal years. Treatment is a combination of surgery with adjuvants chemotherapy and radiotherapy. We report a case of a two-month-old girl presented with a history of vaginal bleeding and a mass protruding from the vagina. Diffusion Magnetic Resonance Imaging (MRI) showed an approximately 6 cm mass in the rectovaginal septum. An excisional biopsy from the vagina confirmed the diagnosis of botyroid rhabdomyosarcoma. After the biopsy, the patient received multiple cycles of chemotherapy. The patient is disease-free and on remission at this time.

Keywords: Botyroid, Rhabdomyosarcoma, Vagina, Tumor

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Submission date: 16.12.2022

Acceptance date: 08.09.2023

DOI : 10.17517/ksutfd.1187920

INTRODUCTION

Gynecologic tumors are rare in childhood with %5 incidence of all pediatric neoplasms. Rhabdomyosarcoma (RMS) are malignant mesenchymal tumors originating from myogenic progenitor cells. They represent the most common soft tissue tumor in childhood. The head and neck region are the most affected tumor sites, followed by the genitourinary tract. Four major histologic subtypes of RMS are identified, Embryonal, Alveolar, Botryoid, pleomorphic/undifferentiated. Botryoid variant is a type of embryonal RMS arising within the wall of the bladder or vagina, can also occur in the cervix. This tumor is seen almost exclusively in infants, it is characterized by 'grape-like' appearance caused by polypoid mass arising in submucosal tissue (1). We report a case of two-month-old girl presented with a history of vaginal bleeding and a mass protruding from vagina.

CASE REPORT

A two-month infant female consulted to our hospital with fresh intermittant vaginal bleeding and mass protruding from vagina. No use of drugs and no history of disease was reported during pregnancy. There was no history of similar disease in family. The baby's vitals were stable.

Pelvic diffusion magnetic resonance imaging (MRI) of lower abdomen was performed. MRI showed a lesion near posterior vaginal wall and vulvar, anal channel with a diameter of approximately 6 cm. The lesion showed hyperintense in T2A images and heterogeneous intense contrast in contrasted series. MRI suggested a malign tumour of mesencyhmal origin and tissue biopsy was advised. Bladder wall and perivesical fat tissue showed normal signal intensity. Dynamic upper abdomen MRI was normal.

18F-FDG (fluorodeoxyglucose) PET/CT imaging of full body was performed. In pelvic region close to anal region a heterogeneous lesion with mild to moderate metabolic activity was observed. No metastases were detected. The excisional biopsy was done from vaginal tumor.

Pathological images of tumor are given in **Figure 1**. Pathological examination of the tumor showed the embryonal rhabdomyosarcoma botyroides. The immunohistochemical staining analysis tumor cells showed intense positivity for desmin, intense positivity for Ki67 (over %80), focal positivity for myogenin and myoD1.

Since no metastases were detected, the girl received chemotherapy. The patient was treated with VAC (vincristine, dactinomycin, cyclophoshamide) chemotherapy for 42 weeks. Due to tumor location, tumor type and the good response to chemotherapy, radical surgery and radiotherapy were not performed. PET scan was used to monitor response to therapy. She has now been in complete remission with no complains for almost 1 year and is followed-up every 3 months via MRI.

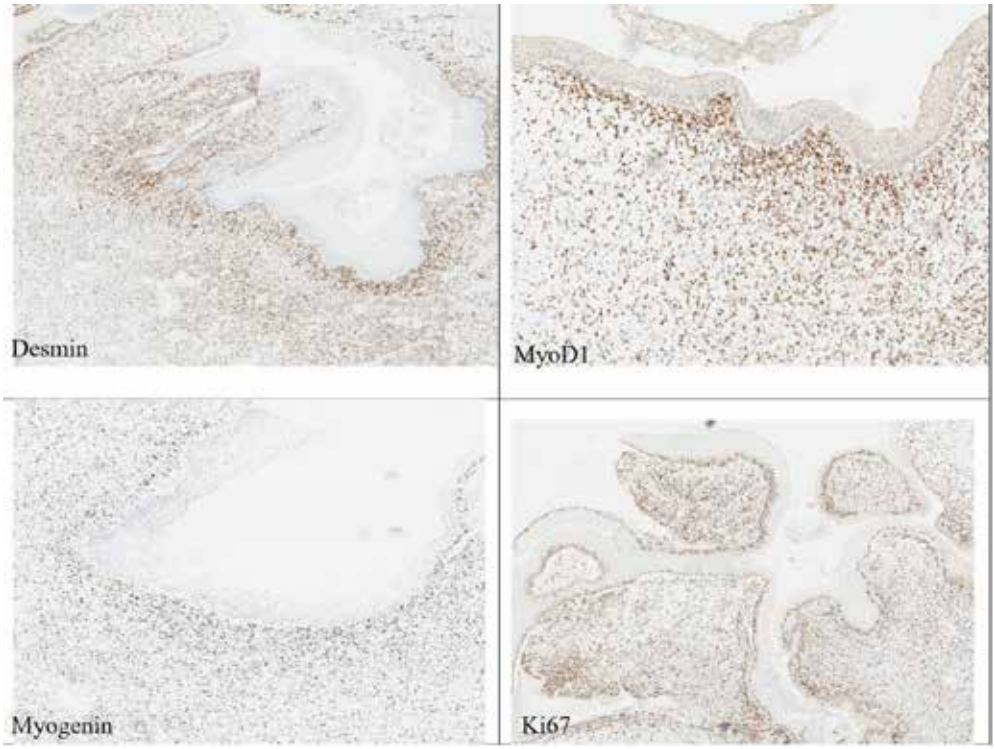

Figure 1. Pathological images of tumor

DISCUSSION

Rhabdomyosarcoma is the most common soft tissue sarcoma in childhood and adolescence and accounts for approximately 5% of all malignancies in children (1). It is a malignant tumor that arises from immature skeletal muscle cells (i.e., rhabdomyoblasts) lining the walls of hollow organs, including the vagina, bladder, and biliary tract. This cancer most commonly affects children under the age of 4, although it can sometimes present in older individuals (2,3). RMS may occur in many anatomic location of the body where there is skeletal muscle, as well as in sites where there are no skeletal muscles, such as urinary bladder and common bile duct. RMS children under 10 years of age generally involves the head and neck or genitourinary sites (2,4,5). The botryoid variant presents as a submucosal lesion with a typical 'grape bunch' appearence. Sarcoma botryoides which arise from the vagina are extremely malignant. This case report describes the importance of early recognition of the typical clinical symptoms of sarcoma botryoides, since a rapid diagnosis followed by treatment is necessary to prevent death (6).

CONCLUSION

We think that this case report of rhabdomyosarcoma, which is rarely seen in childhood, will contribute to the current literature.

Conflict of Interest and Funding Declaration: The authors declare that they have no conflict of interest.

Informed Consent: Informed written consent was obtained from the patient presented in this article.

Author Contribution: The authors declare that they have contributed equally to the article.

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