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CASE REPORT

Minimal invasive treatment of cervical rhabdomyosarcoma in an adolescent girl

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Summary

Rhabdomyosarcomas are the most common soft tissue sarcomas in childhood and adolescence. In cases of minimal cervical invasion, less invasive local excision in combination with adjuvant chemotherapy has resulted in excellent survival rates with complete preservation of the bladder, rectum, uterus and vagina. Herein we present the case of an adolescent girl with a large rhabdomyosarcoma of the uterine cervix treated with minimal invasive therapy.

Key words: adolescence, minimal invasive surgery, rhabdomyosarcoma, vaginoscopy

Introduction

Rhabdomyosarcomas are the most common soft tissue sarcomas in childhood and adolescence. Genitourinary sites comprise 17% of the primary location of these tumors [1]. The botryoid variant arises in infancy from the lower vagina or urinary bladder [2]. Cervical botryoid sarcoma is usually found in older girls with peak incidence in the second decade of life. The tumor usually appears as a mass protruding from the introitus of the vagina or with bloody discharge or irregular bleeding. Historically, exenteration was the treatment of choice. Evidence from previous Intergroup Rhabdomyosarcoma studies has established the superior effect of combining chemotherapy and radiation therapy, with less radical surgery in low-risk patients [3,4].

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Herein we present a case of a large cervical botryoid rhabdomyosarcoma in a 14-year-old girl treated with minimal invasive therapy.

Case presentation

A 14-year-old, previously healthy girl, was referred to our Institute for irregular menstrual cycles and anemia. Her menarche occurred when she was 11 years old and she had not been sexually active. In the previous 3 months her local gynecologists performed 2 sonographic examinations and prescribed progesterone p.o. The initial diagnosis was dysfunctional uterine bleeding, but vaginal bleeding continued almost every day despite the therapy given. Laboratory investigations showed low red blood cell count (2.9×10^{12}/l), hemoglobin 8.5 g/l and hematocrit 27%.

Physical examination showed normal abdomen, but rectal bimanual palpation revealed a pelvic tumor. Abdominal ultrasound revealed a huge solid tumor probably arising from the vagina. Doppler flow examination showed vascularization near the mass only in a small part close to the cervix. Computed tomographic (CT) scan revealed a solid vaginal tumor with possible involvement of the uterus and urinary bladder, with a volume of 115 cm^3 (Figure 1). Tumor markers, including α-fetoprotein, β-HCG and LDH were normal.



Figure 1. Computed tomographic scan shows a solid vaginal tumor (arrow) with possible involvement of the uterus and urinary bladder.

Biopsy of the mass was performed and frozen sections were sent for histological diagnosis which showed botryoid rhabdomyosarcoma. It was then decided to explore the vaginal cavity. The tumor distended the whole vagina but without involvement of the vaginal walls. Vaginoscopic resection of the tumor was performed and, firstly, one third of the tumor was excised. We then noticed that the tumor actually originated from a 2×2 mm pedicle at the posterior part of the uterine cervix (Figure 2). The operation ended with total tumor excision along with a small part of the cervix. The final pathology report revealed embryonal rhabdomyosarcoma without tumor cells in the excised part of the cervix. The patient received postoperative adjuvant chemotherapy with vincristine, actinomycin-D and cyclophosphamide (VAC) every 3 weeks for 4 cycles.

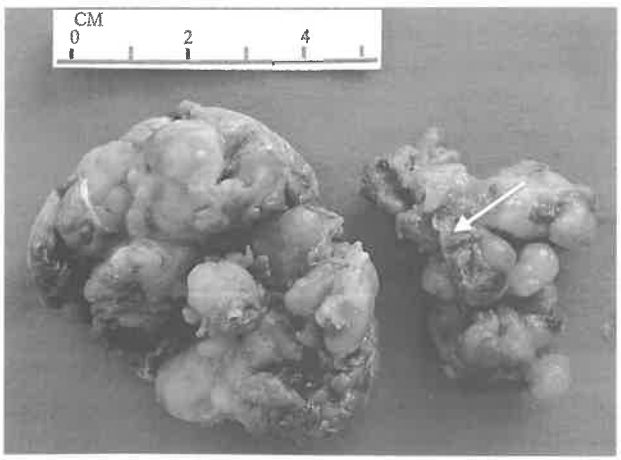


Figure 2. Macroscopic appearance of botryoid rhabdomyosarcoma and calcifications surrounding the central tumor vessel (arrow).

Discussion

The presentation of genital botryoid rhabdomyosarcoma is usually by a mass protruding from the vaginal introitus or with foul-smelling or bloody discharge in girls [2-5]. In the presented patient with abnormal bleeding the origin of the disease was not properly defined in the first 3 months. Most of the solid pediatric genital tumors have malignant biological potential and vascularization. An interesting part of the diagnostic process was the absence of Doppler flow during the sonographic exploration of the mass. We supposed that this was caused by the calcifications which surrounded the central tumor vessel. Staging requires evaluation of the disease extent, usually with CT of the chest and abdomen and bone scan and/or bone marrow biopsy. Ultrasound and magnetic resonance imaging (MRI) can assist in the identification of lymph node involvement, and vaginoscopy can delineate the extent of the disease and any extension into the cervix. According to the Intergroup classification [3], our patient had stage I disease.

In the past, treatment of uterine sarcomas involved total pelvic exenteration, which caused significant physical and psychological morbidity in these adolescent patients. In 1976, Mahesh Kumar and colleagues reported 92% 3-year disease-free survival rate in patients treated with chemotherapy and conservative surgery [6]. Conservative management has also been shown to be effective for botryoid sarcoma involving the cervix [7]. A report from the Intergroup Rhabdomyosarcoma Study Group showed that initial excision of ≥50% of the tumor may be associated with increased failure-free survival [8]. For the last 3.5 years our patient is free of disease.

We conclude that ultrasonographic examination for abnormal vaginal bleeding during adolescence should include evaluation of the vaginal appearance. Absence of Doppler flow in a vaginal tumor do not exclude presence of malignancy. Minor surgical approaches in combination with chemotherapy for early stage I cervical rhabdomyosarcoma in adolescent girls are recommended.

References

1. Bhurgri Y, Bhurgri A, Puri R et al. Rhabdomyosarcoma in Karachi 1998-2002. Asian Pac J Cancer Prev 2004; 5: 284-290.
2. Emans SJ. Vulvovaginal Problems in the Prepubertal Child. In: Emans SJ, Laufer MR, Goldstein DP (Eds): Pediatric and Adolescent Gynecology (5th edn). Philadelphia: Lippincott Williams and Wilkins, 2005, pp 83-119.

3. Andrassy RJ, Wiener ES, Raney RB et al. Progress in the surgical management of vaginal rhabdomyosarcoma: a 25-year review from the Intergroup Rhabdomyosarcoma Study Group. J Pediatr Surg 1999; 34: 731-715.
4. Solomon LA, Zuravin RK, Edwards CL. Vaginoscopic resection for rhabdomyosarcoma of the vagina: a case report and review of the literature. J Pediatr Adolesc Gynecol 2003; 16: 139-142.
5. Gruessner SE, Omwandho CO, Dreyer T et al. Management of stage I cervical sarcoma botryoides in childhood and adolescence. Eur J Pediatr 2004; 163: 452-456.
6. Mahesh Kumar AP, Wrenn EL Jr, Fleming ID, Omar Hustu H, Pratt CB. Combined therapy to prevent complete pelvic exenteration for rhabdomyosarcoma of the vagina or uterus. Cancer 1976; 37: 118-119.
7. Harting MT, Blakely ML, Andrassy RJ. Surgical management of gynecologic rhabdomyosarcoma. Curr Treat Options Oncol 2004; 5: 109-118.
8. Raney RB, Stoner JA, Walterhouse DO et al. Intergroup Rhabdomyosarcoma Study-IV, 1991-1997. Results of treatment of fifty-six patients with localized retroperitoneal and pelvic rhabdomyosarcoma: a report from the Intergroup Rhabdomyosarcoma Study-IV, 1991-97. Pediatr Blood Cancer 2004; 42: 618-625.