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Rhabdomyosarcoma of the Cervix, Local Excision or Radical Surgery (Report of Two Cases and Review of the Literature)

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Abstract

Introduction: Embryonal (Botryoid) Rhabdomyosarcoma (RMS) is an aggressive malignancy that arises from embryonal rhabdomyoblasts. It is commonly seen in the genital tract of female infants and young children. The primary site of these tumors is closely related to the age of the patient. Embryonal Rhabdomyosarcoma has a marked tendency for local recurrence after excision. Due to young age of affected patients who desire fertility, the management of this rapidly growing malignancy is very critical and poses challenges.

Case Presentation: We report on two cases embryonal rhabdomyosarcoma of uterine cervix, who were referred to Imam Khomeini hospital during year 2014. Both of them were young virgin females. The presenting symptom for both was vaginal bleeding and protrusion of polypoid mass from the hymen. After neoadjuvant chemotherapy, radical hysterectomy was offered to them. One of them refused, thus local excision was done. Both patients received adjuvant chemotherapy yet in the patient with local excision, the tumor recurred with multiple metastases.

Conclusions: There are several methods of surgical approach and variation in adjuvant therapy in the management of embryonal rhabdomyosarcoma. If we choose a conservative approach for surgery of early stage, surgical margin should be negative and in other cases doing radical surgery is the best.

Keywords: Embryonal Rhabdomyosarcoma, Cervix, Surgery

1. Introduction

Embryonal (Botryoid) rhabdomyosarcomas are soft tissue tumors occurring most commonly in young patients (1, 2). These tumors arise from embryonal rhabdomyoblast and are approximately 3% of all rhabdomyosarcomas. The primary site of these tumors is closely related to the age of the patient; it is found in the vagina during infancy and early childhood, in the cervix during the reproductive age, and in the corpus uteri in post-menopausal patients (3, 4).

Because of the invading nature and tendency to local recurrence of these tumors, appropriate surgical treatment with adjuvant therapy should be done. Especially when the patient is young and saving the fertility is the request of the patient, optimal management is challenging (5, 6).

2. Case Presentation

Here, we report on two cases of embryonal rhabdomyosarcoma of uterine cervix, who referred to our gyneco-oncology center in Imam Khomeini hospital during year 2014. Both of them were young virgin females and were 23 and 25 years old, respectively. The presenting symptom for both was vaginal bleeding and protrusion of polypoid mass from the hymen. After excisional biopsy and confirmation of diagnosis with pathology, these patients received three cycles of neo-adjuvant chemotherapy with Vincristine, Adriamycin and Cyclophosphamide (VAC regimen) and after reducing the size of tumor, we suggested radical hysterectomy for both of them. One accepted and radical hysterectomy was performed, but the other refused and local excision of the mass and cervix was done. We took informed consent from both patients for selected surgical approach and its outcomes and also for using these data in clinical research. The final pathology in this patient revealed positive margin in one section. Both patients received three cycles of VAC regimen after surgery. In the one-year follow-up, the patient, who had a radical hysterectomy was tumor free, but in the patient with local excision, three months after the end of chemotherapy, multiple metastatic masses was found in the pelvic and abdomen. She received two cycles of chemotherapy with Adriamycin and Cisplatin and then incomplete debulking surgery was done, due to her request but she died with obstruction and renal failure presentation due to multiple

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massive metastatic tumors in abdomen and pelvic areas.

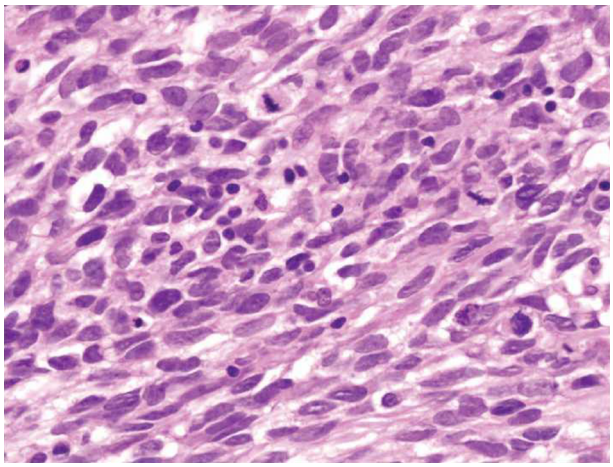


Figure 1. Microscopic Feature of Embryonal Rhabdomyosarcoma of the Cervix (Magnification x400)

3. Discussion

Embryonal rhabdomyosarcoma of uterine cervix is an uncommon presentation of the most common soft tissue sarcoma in young females (7). There are a few case reports and case series regarding the management of embryonal rhabdomyosarcoma of female genital tract in the literature. Because of the rare incidence of this malignancy, there are limited evidences for optimal treatment of this tumor and there is not any consensus for its management, aggressive radical surgery gradually reduced from exentration procedure to simple local excision.

This tumor is mostly treated with surgery and adjuvant chemotherapy (8). There has been no agreement on chemotherapy regimen but the VAC regimen was most widely used. There are reports of using neoadjuvant chemotherapy (NACT) to shrink large tumor before surgery. Nowadays, radiotherapy is not used as an adjuvant treatment because this tumor is not radiosensitive.

Copeland et al. reported a case series of embryonal rhabdomyosarcoma of female genital tract with 14 patients and showed the evolution of treatment over a 30-year period (9). Daya et al. reported a series of these patients with conservative approach and favorable outcome (10).

Fatal recurrence with poor outcome was reported with the use of conservative surgery alone. Overall survival has been reported as 37% to 79% in patients treated with surgery and adjuvant chemotherapy. There are case reports of unfavorable outcome despite an adequate surgical excision and chemotherapy. The risk of recurrence and metastatic spread, especially in patients treated with conservative approach, is a concern. Fertility-sparing surgery should not be considered with the presence of extensive uterine involvement or metastases (11). We suggest that, if negative surgical margin cannot be achieved with conservative surgery, radical hysterectomy should be performed.

Footnote

Conflict of Interest: The authors declare that there were no conflicts of interest.

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