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Case Report

Sarcoma botryoides in a 14 year old girl: a rare case

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ABSTRACT

Embryonal rhabdomyosarcoma (Sarcoma botryoides) of the uterine cervix is an uncommon entity. Because of extreme rarity its discussion has mainly been in the light of individual case reports. We report a case of a 14-year old female who presented with irregular vaginal bleeding and cervical polyp. Her biopsy specimen confirmed sarcoma botryoides and she underwent abdominal hysterectomy with wide excision of vaginal cuff after a multidisciplinary consultation.

Keywords: Embryonal rhabdomyosarcoma, Sarcoma botryoides

INTRODUCTION

Embryonal rhabdomyosarcoma (ERM) originates from embryonal mesoderm and arises under the mucosal surface of body orifices such as the vagina, bladder and cervix. ERM is the most common soft tissue sarcoma in children and young adults and accounts for 4% to 6% of all malignancies in this age group.[1,2] While affecting the genitourinary tract, the ERM is most commonly manifested in the vagina,[3] and it usually presents before the age of 4 years.[2,4] Only 0.5% of ERM in females affect the cervix,[4] and is usually occurs in the second decade of life as seen in our patient, however it has also been reported in babies of 5 months age.[5] Few cases of ERM have been reported in the women older than 40 years.[6-9] A subset of ERM are grossly polypoid and are descriptively referred to as botryoid (grape like), which was a salient feature of our case too.

CASE REPORT

We report here a case of 14 year old school student who had attained menarche 3 months back, and had been experiencing irregular bleeding per vagnium since then. She also felt some mass coming out through her introitus on straining, for the same duration. The mass was fleshy, gradually increasing in size and often associated with foul smelling mucoid discharge. There was no significant past medical or surgical history and no family history of any malignancy. On examination, patient had mild pallor and there was no lymphadenopathy. Her secondary sexual characters were normally developed. On local examination, multiple grape like lesions were protruding through the introitus. The lesions had smooth glistening surface smeared with mucosanguinous secretion. Per vaginal examination revealed a spacious vagina occupied by the lesion, however the vaginal mucosa was free. A normal sized uterus with normal adnexa was palpated and the growth seemed to arise from the cervix. Rectal mucosa was free on per rectal examination. In the course of local examination few fragments from the growth got detached and these were submitted for histopathological examination. Ultrasonography of the patient showed utero cervical growth and CECT abdomen revealed a well-defined 8.2 x 8 x 7 cm mass lesion in the lower part of body of uterus and cervix. There was no evidence of pelvic lymphadenopathy and parametrium was reported normal. The histological examination of the lesion was consistent with an embryonal rhabdomyosarcoma (botryoide type). After a multidisciplinary consultation

and counseling the patient and her family, abdominal hysterectomy with wide excision of vaginal cuff along with omental and pelvic lymph node sampling was performed. Later the specimen report showed disease free surgical margins and lymph nodes. Her post-operative period was uneventful and patient was referred to regional cancer centre for chemotherapy.

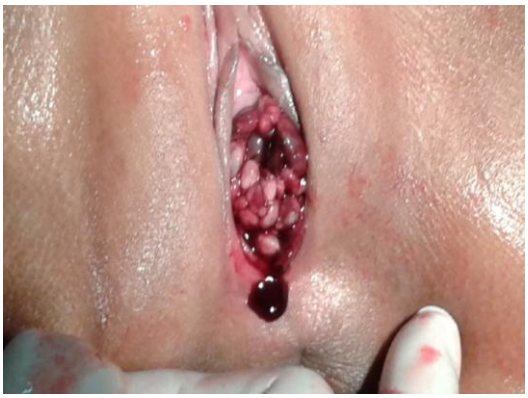

Figure 1: Grape like mass protruding through introitus.

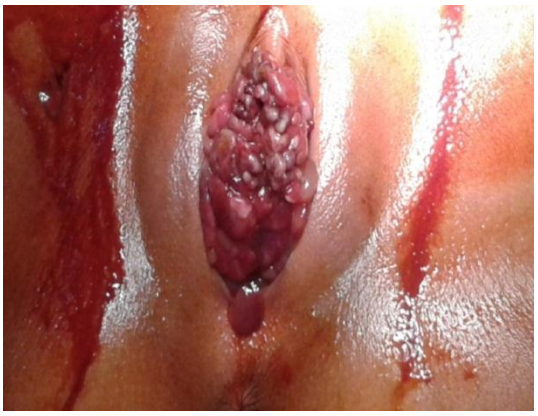

Figure 2: Examination under anaesthesia revealing extensive growth.

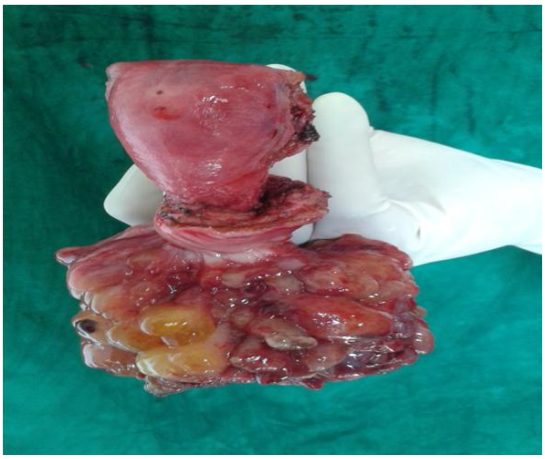

Figure 3: Hysterectomised specimen showing uterus along with sarcoma botryoides of the cervix.

DISCUSSION

Sarcoma botryoides in the cervix is rare and accounts for about 10% of all RMS cases.[10] Case reports have been the major source of information regarding this topic. It typically occurs in young females and most commonly affected age group is between 12-26 years.[11,12] Clinically sarcoma botryoides can present as abnormal vaginal bleeding, prolapsing mass per vaginum or an abdominopelvic mass. Histological features include undifferentiated mesodermal and striated muscle cells with fibromyxomatous stroma and a distinct "cambium layer" beneath the epithelium which is characteristic of sarcoma botryoides.[13]

The available treatment options for sarcoma botryoides include radical surgery, fertility sparing surgery, chemoradiation and combined approach. The optimal management of these tumors is still not codified owing to their rarity; however, combined approach to treatment may result in better outcome.[14] According to some authors, the prognosis of cervical sarcoma botryoid is more favorable than other genital RMS, particularly when the tumor arises as a single polypoid lesion and the polyp is completely removed.[15,16] Similarly in another study Daya and Scully reported favorable outcome for cervical botryoid in comparison to the vaginal counterpart and demonstrated that 3 of the 13 patients treated with fertility-sparing surgery followed by chemotherapy had results comparable to those treated with more radical surgery with or without chemotherapy.[17] However a contrast report was shown by Gruessner et al. in their study, stating higher survival rates and better prognosis for vaginal lesions. They demonstrated 96% survival rate for vaginal lesions versus 60% for cervical botryoid.[18] Regarding chemotherapy, the most widely used regimen is vincristine, d-actinomycin and cyclophosphamide (VAC). Gordon et al reported that between 6-12 cycles of VAC there was good chance of return of menstruation and reproductive function.[19]

Considering our patient, following a multidisciplinary consultation, abdominal hysterectomy with wide excision of vaginal cuff was performed. The decision for surgery was taken keeping in mind the poor patient profile likely to be lost to follow up. Literature too has stated a similar case of a 16 year old Moroccan girl[11] with sarcoma botryoides where the lesion persisted even after chemotherapy and subsequently she had to undergo surgery. The risk of recurrence of the disease, poor patient compliance and follow up, and the patient party consenting for surgery were the main factors which prompted us to take a surgical approach. The post-operative period of the patient was uneventful and she was discharged on 10th day following surgery.

CONCLUSION

Sarcoma botryoides of the cervix is a rare disease and it presents as a cervicovaginal polyp in the 2nd decade of

life. Histopathology is diagnostic but the treatment protocol is not codified due to its rarity. Surgery along with chemo radiation may results in prolonged survival, but requires more evidence.

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