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Case Report

Recurrent vaginal discharge: an unusual presentation of embryonal rhabdomyosarcoma of uterine cervix in an adolescent girl

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ABSTRACT

Embryonal rhabdomyosarcoma (ERMS) is a rare tumor of the female genital tract. It tends to occur during childhood in the vagina and rarely it can arise in the uterine cervix, with a peak incidence in the second decade. We report a case of 15 year old adolescent girl who presented with recurrent vaginal discharge not responding to medical treatment. Examination under anesthesia showed friable growth arising from the cervix. Histopathological examination revealed embryonal rhabdomyosarcoma (botyroid variant) of the cervix. Patient underwent local excision of growth followed by IRS-4 protocol based chemotherapy and now patient is under follow up at our side and pediatric oncology and doing well. Young girls presenting with recurrent vaginal discharge not responding to medical treatment must undergo proper clinical examination and EUA and any suspicious lesions should be examined so as to avoid missing rare but aggressive etiology like rhabdomyosarcoma. Due to the young age of affected patients, embryonal rhabdomyosarcoma (sarcoma botyroides) poses a management challenge as the preservation of hormonal, sexual and reproductive function is essential. Awareness of such as uncommon lesion and its clinical implications is important for the counseling and management of the patient.

Keywords: Embryonal rhabdomyosarcoma, IRS-4 protocol, Sarcoma botyroides, Uterine cervix neoplasm

INTRODUCTION

Embryonal rhabdomyosarcoma (ERMS) is a highly malignant tumor arising from embryonal mesenchymal cells committed to develop into striated muscles and is the most frequent soft tissue sarcoma in childhood and young adults, accounting for 4-6% of all malignancies in this age group.[1] The head and neck were the most frequent sites of origin (35%), followed by the genitourinary tract (25%). One of the least common sites for rhabdomyosarcoma in the genitourinary tract is the uterine cervix. 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 active reproductive stage and in the corpus uterus for postmenopausal patients. Although vaginal tumors are five times more common than the cervical type, the latter appears to have a better prognosis than the former.

Rhabdomyosarcoma (RMS) is classified by World Health Organization (2013) into embryonal RMS (inducing botyroid, anaplastic), alveolar RMS (including solid, anaplastic), pleomorphic RMS and spindle cell / sclerosarcoma RMS.[2] Currently, there are no specific factors identified for embryonal rhabdomyosarcoma of uterine cervix. However, RMS may be associated with certain genetic disorders including Beckwith-Wiedemann syndrome, Costello syndrome and neurofibromatosis type 1 and others.

We report a case of sarcoma botyroides of the cervix presenting as recurrent - vaginal discharge in a 15 year

old adolescent girl, treated with conservative surgery and adjuvant chemotherapy.

CASE REPORT

Authors report a case of 15 year old adolescent girl who presented with the chief complaint of discharge per vaginum not responding to medications as prescribed from various hospitals. Patient attained menarche at age 12, subsequent cycles regular for rhythm, quantity and length; the patient never had sexual activity. Family history was not significant. With proper consent patient was subjected to clinical examination. General examination was within normal limits. On per abdomen examination a suprapubic mass of ≈14-16 weeks size felt with regular margins and restricted mobility, mass non-tender, lower pole not reached.

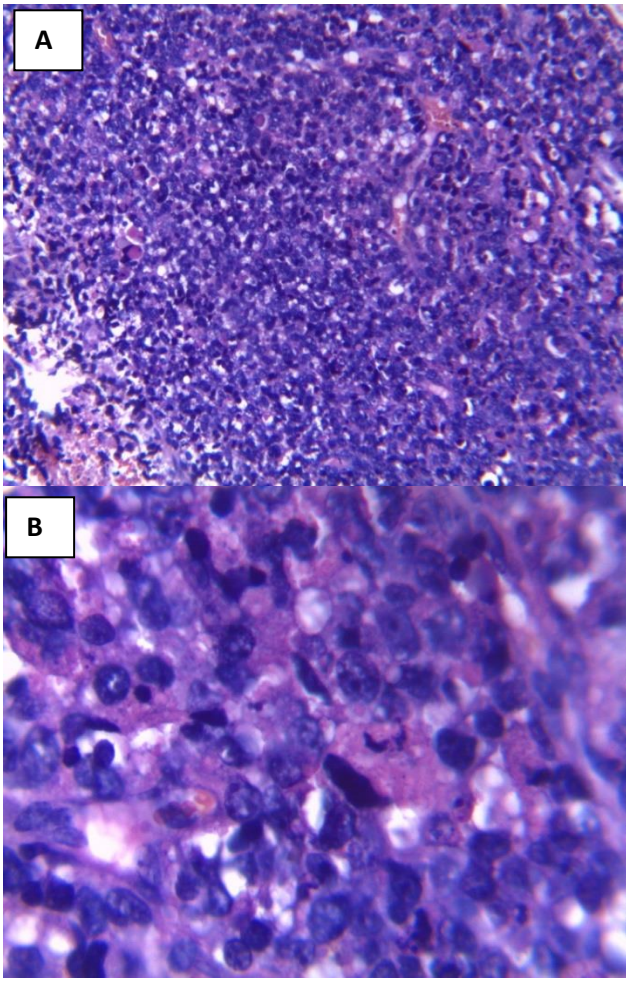

Figure 1: Microscopic section shows vimentin positive tumor cells with (A) dense mononuclear cell infiltrate all over the oedematosis. (B) loosely arranged oedematous stromal tissue admixed with atypical undifferentiated round to spindle cells.

Local examination showed dirty blood mixed discharge coming through vagina. Per rectal examination was done and an irregular mass of ≈7x6cm to felt seems to be arising from cervix, rectal mucosa free, bilateral parametrium free. All baseline blood investigations were done (hemogram, blood group, LFT, KFT, PT/INR). USG- whole abdomen and pelvis was advised which shows bulky uterus with large heterogeneously enhancing SOL in lower uterine segment involving cervix. Patient underwent examination under anesthesia (EUA) during which a friable growth of 5x6cm seen arising from infravaginal part of post lip of cervix from which biopsy was taken and sent for histopathological examination.

Microscopic examination revealed spindle mesenchymal cells of average and small size, surrounded by a myxoid stroma. Immunohistochemical staining highlighted positive cells for vimentin and myogenin and negative cells for estrogen and progesterone receptors [Figure 1 (A and B)]. These findings were consistent with ERMs (Sarcoma botyroides) of the uterine cervix.

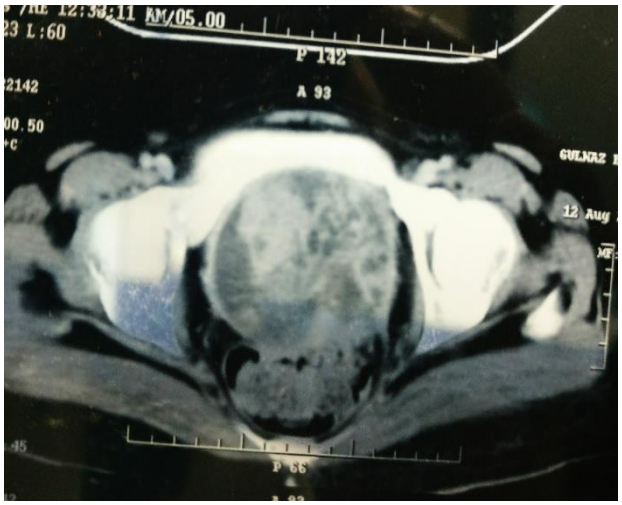

Figure 2: Computed Tomography of pelvis showing enlarged uterus with ill-defined inhomogenously enhancing soft tissue lesion in lower uterine segment.

Following this, computed tomography of her thorax abdomen and pelvis were performed which revealed enlarged uterus with ill-defined inhomogenously enhancing soft tissue lesion with multiple necrotic areas of around 9x8cm in lower uterine segment abutting base of urinary bladder and bowel loops (Figure 2).

No metastasis seen in any other viscerae. After multidisciplinary discussion including pediatrician, surgical oncologist and gynecologist, this tumor was staged as group IIIA and received IRS-IV (Intergroup rhabdomyosarcoma study- IV) protocol based neo-adjuvant chemotherapy in pediatric oncology consisting of 5 cycles of VAC pattern (vincristine 1.5mg/m^2, actinomycin-D 45u/kg, cyclophosphamide 1200mg/m^2) plus mesna and further 3 cycles of induction therapy with vincristine, ifosfamide and etoposide. Therafter patient underwent EUA for removal of residual growth. Following this patient received maintenance chemotherapy of VAC for further 8 cycles.

After completion of neo adjuvant chemotherapy, MRI shows no residual disease (Figure 3). Patient underwent laparoscopic oophoropexy prior to radiotherapy and received 25 cycles of radiotherapy. Till date, the patient is at 2nd year follow-up monthly, in complete remission without clinical signs of ovarian failure.

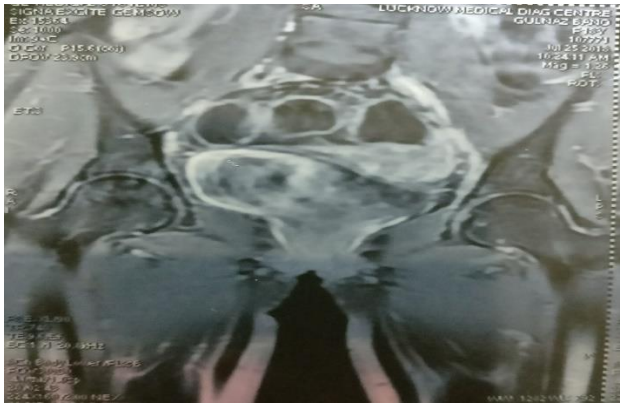

Figure 3: Post RT, MRI pelvis with contrast showing no obvious residual or recurrent lesion.

DISCUSSION

Sarcoma botryoides in the cervix is rare and most of our information is based on individual case reports. Currently the largest series consisting of original cases included 14 patients.[5] Furthermore, information regarding this tumor is difficult to obtain from the literature because the primary place of origin is not always known or described.

Earlier, radical surgery was considered the gold standard. However, after consecutive trials, the IRSG (Intergroup rhabdomyosarcoma study group) has modified the treatment protocols with the addition of multiple chemotherapeutic agents to surgery, greatly improving the prognosis and survival of this tumor and also allowed more conservative fertility-sparing surgeries in early stages of the disease.[3] Consequently, localized female genital tract disease normally is curable with combination of chemotherapy, a conservative surgical approach and the use of radiotherapy for selected patients4. Nevertheless the extent of surgical therapy and subsequent adjuvant therapy should be balanced between the patient's desire to maintain fertility and the presence of unfavorable prognostic variables, such as extensive uterine involvement and/or metastasis, deep myometrial invasion, lymphatic invasion and foci of alveolar subtype.[3,5,6] The presence of these factors should lead to discussion with the patient and eventually their parents or relatives to more aggressive treatment.[1,4,7]

In the case reported, after a multidisciplinary team evaluation, we have decided to perform fertility-sparing surgery followed by chemotherapy and radiotherapy for the treatment of this tumor.

More recent studies also report successful treatment of these tumors with fertility-sparing surgery with an innovating way. cervical conization or robotic-assisted radical trachelectomy in combination with multi-agent chemotherapy.[8,9]

Over recent years, the approach of ERMS, like other malignant diseases have been increasingly conservative in the sense of the preservation of fertility, given its prevalence in young nulliparous.[5,8,9] This case represents another example of successful conservative treatment of this kind of tumor.

CONCLUSION

Young girls presenting with recurrent vaginal discharge not responding to medical treatment must undergo proper clinical examination and EUA and any suspicious lesions should be examined so as to avoid missing rare but aggressive etiology like rhabdomyosarcoma. This is extremely important especially as an early disease stage at diagnosis is a highly favorable prognostic factor. Surgery and chemotherapy are the mainstays of treatment of cervical rhabdomyosarcoma, and the prognosis of patients treated with multimodal therapy is very good.

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