Case Report

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A RARE CASE REPORT OF ALVEOLAR RHABDOMYOSARCOMA IN VAGINA OF YOUNG FEMALES

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ABSTRACT

Alveolar rhabdomyosarcoma in a rare aggressive tumour which manifests in children and young adults. The site of alveolar rhabdomyosarcoma in vagina is also extremely rare. We presented a case report of 38 yrs.lady who was having complain of intermenstrual bleeding & discharge per vaginum from last 6 months. A large fleshy infiltrative bluish friable mass (3*3cm) present at the junction of middle and upper 1/3rd and upper 1/3rd of posterior vaginal wall . Another similar mass (2* 2cm) was also present in the lateral vaginal wall. Wide excision of both growth was done & postoperatively chemotherapy was given.

KEYWORDS: Rhabdomyosarcoma, Alveolar , Chemotherapy ,wide excision .

INTRODUCTION

Rhabdomyosarcoma of vagina is a very rare high-grade malignant tumor. It is a malignant mesenchymal tumor which arise from the embryonal muscle cells (rhabdomyoblasts) and it mostly affects children and young adults (1). It is very uncommon in patients over 20 years of age, as in our case.

There are 4 WHO (world health organisation) types of rhabdomyosarcoma which includes :

- Embryonal rhabdomyosarcoma 65% (most common variety) & best prognosis.
- Alveolar rhabdomyosarcoma 20-25% (2nd most common) & poor prognosis.
- Pleomorphic subtype (13%).
- Spindle cell/ sclerosing rhabdomyosarcoma .

The alveolar subtype mainly affects older children & young adults (peak age 10-25 yrs.) & most common site involved are the extremities & perineal site (2). The alveolar rhabdomyosarcomas which involves vagina, is extremely rare. It is also typically more aggressive than other subtypes.(3)

HISTORY OF PRESENT ILLNESS:

A 38 years old P1+2L1 presented to the out patient department of obstetrics and gynaecology at Era's Lucknow Medical College & hospital, Lucknow with complains of intermenstrual bleeding and discharge per vaginum from last 6months with history of hypertension -of duration 20 years, asthma and hypothyroidism both from last 10 years.

MENSTRUAL HISTORY: Regular menstrual cycles, average amount of blood flow, duration-4 to 5 days.

OBSTETRICS HISTORY : $P_{1+2}L_1$ (just after marriage she conceived which result in two abortions at interval of 6 month & after 17 yrs gap, took treatment for secondary infertility & conceived by in vitro fertilisation following which a healthy male child was delivered by lower segment cesarean section).

CLINICAL EXAMINATION

PER ABDOMEN: soft, non-tender, no organomegaly

PER SPECULUM: A large fleshy infiltrative bluish friable mass (3X3cm) present at the junction of middle and upper 1/3rd and upper 1/3rd of posterior vaginal wall (Fig.1). Another similar mass (2X2cm) was also present in the lateral vaginal wall (Fig. 2). Cervix was healthy.

PER VAGINUM: Vaginal examination confirmed the presence of polypoidal growth in vagina . The polypoidal growth was firm, non-tender, friable and did not bleed on touch. Surrounding vagina was smooth, no sign of induration and no sign of inflammation present. Uterus was mobile, bilateral fornices were clear and non-tender growth (bluish friable mass).

On the basis of these finding our differential diagnosis is :

- Vaginal growth (benign/malignant)
- Choriocarcinoma
- Vaginal varicosities
- Hemangioma
- Rhabdomyosarcoma

INVESTIGATIONS

Beta HCG (human chorinicgonadotropin) on investigation is 0.1 µIU/ml, which was done to rule out choriocarcinoma.

USG of whole abdomen including Pelvis: shows normal finding.

All necessary pre procedural investigation for surgical procedure were carried and were within normal limits.

Wide excision biopsy of both vaginal growth were performed & sent for histopathological examination.

Endometrial sampling was done, to rule out any endometrial pathology.

MANAGEMENT

Under day care anaesthesia wide excisional biopsy of both vaginal growth was done. Endometrial biopsy was also done and tissue was sent for histopathological examination.

Histopatological examination report of excised growth showed tumour comprising of round to spindle shaped cell having eosinophilic cytoplasm with uniform nuclei, 1-2 nucleoli and showing mild pleomorphism and rare mitotic activity. At various places these cells are arranged in nest of variable size ,separated by fibrous septa. Cells are loosely arranged in centre (alveolar pattern) while adhere in single layer at periphery of nest.

Endometrial tissue was non secretory.

Patient was referred to surgical oncology centre for further management. Chemotherapy was given at oncology centre. Further management was not known as patient lost to follow up.

DISCUSSION

Adult alveolar rhabdomyosarcoma is extremely rare . Site of gynecological origin is also less common. Only 7 cases of adult alveolar rhabdomyosarcoma in the vulva was reported (4) . There are early involvement of lymphatics and hematogenous tissue in alveolar rhabdomyosarcoma and the most common sites of metastasis are lung &bones. A similar case report of alveolar rhabdomyosarcoma was reported in 20 yrs old lady in which right side of vulva is involved and tumor metastasized to the bone within a few months (4).

- According to Intergroup Rhabdomyosarcoma study incidence of known lymph nodes metastasis is 26%.
- There is no standard treatment strategy established yet. In most cases of adults, multimodality treatment is given in which surgery is performed & chemotherapy & ssradiotherapy are used as adjunct.
- Two main chemotherapy regimens used in the treatment of rhabdomyosarcoma include a) VAC regimen which consist of vincristine, actinomycin D and Cyclophosphamide. B) IVA regimen ,which consist of ifosfamide ,vincistine & actinomycin-D .These drugs are administered upto 15 cycle depending on disease progression (5)
- In alveolar rhabdomyosarcoma of young patient, the 5 year survival rate after surgery is 29%+_10%(4).

CONCLUSION

Rhabdomyosarcoma is a rapidly growing malignancy. The prognosis of rhabdomyosarcoma depends on the the anatomic location at the time of presentation, patient's age, extent of removal of mass, extent of metastasic disease and tumor histology . Early consultation with doctors and aggressive surgery in combination with multidrug chemotherapy may improved the outcome.

CONFLICT OF INTEREST: None

FINANCIAL CONSIDERATION : None

ETHICAL CONSIDERATION: Patient identifiers have been anonymised

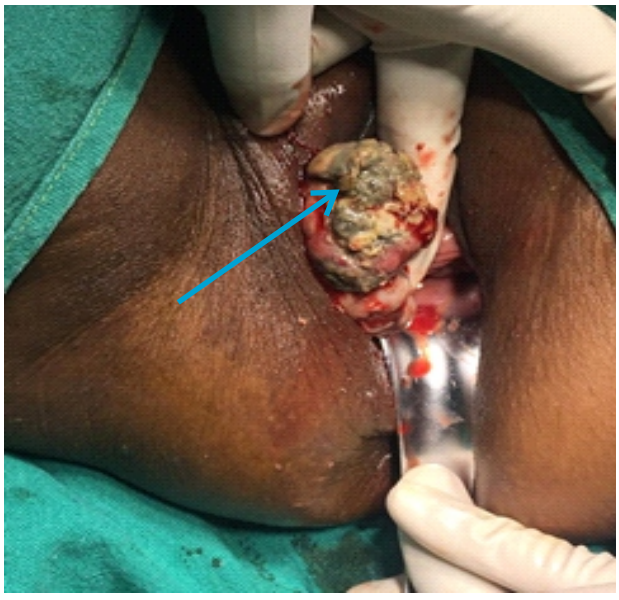

Fig. 1: Showing Site of Origin of Vaginal Growth

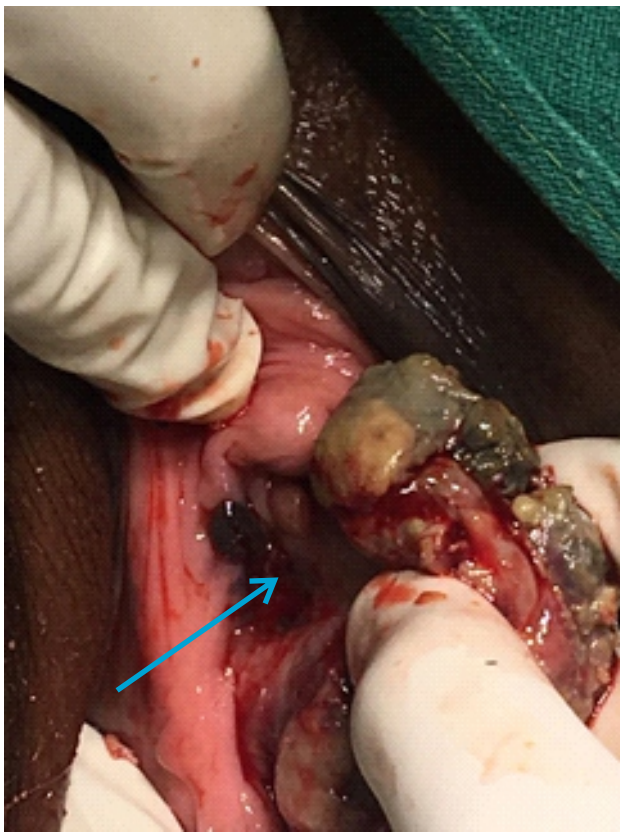

Fig. 1: Showing Another Similar Mass in Vagina

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