

Vaginal Sarcoma: A Rare Case Report and Review of Literature from Tertiary Care Hospital of Northern India

INTRODUCTION

Primary vaginal cancer is a rare entity, representing 1-2% of female genital neoplasia, since 80-90% vaginal neoplasia are metastasis from other organs or direct extension from uterus or cervix (1). Vaginal sarcomas constitute only 3% of the primary vaginal cancers and are most common in adults with leiomyosarcoma representing 50-60 % of vaginal sarcomas and mixed mullerian tumours, angiosarcoma and endometrial stromal sarcoma being less prevalent (1). Rhabdomyosarcoma or sarcoma botryoides is a rare tumour of paediatric age group. These tumours are known for their very aggressive nature and distant metastasis primarily by hematogenous spread. Herein we present another case of vaginal sarcoma in a 21 year old woman to add to the sparse literature.

Key words: vaginal cancer, Vaginal Sarcoma, genital neoplasia

CASE REPORT

A 21 year old unmarried nulliparous woman presented to the Gynecology OPD of GGSMC&H two months back with sensation of pressure in vagina for since four months accompanied by discharge for previous two months which was pinkish in colour, copious in amount that the patient had to use 2-3 vulval pads per day and had difficulty in micturition. She had normal menstrual cycle. There was no history of any chronic disease or previous medication intake. Family history was insignificant. She was sexually active for last 2 years. On examination, her general condition and vitals were stable. On local examination, a 4*4cm pedunculated mass was felt in the vagina arising from right anterolateral vaginal wall and there was pus like discharge. The mass was freely mobile and not fixed to adjacent structures. No pelvic or inguinal lymph nodes were palpable. Rectal examination did not reveal any abnormality. An Ultrasonography reported a 55x50 mm heterogeneous lesion in the vagina with query of cervical or vaginal polyp or cystic mass. The absence of distant disease was confirmed with a PET-CT. A provisional diagnosis of vaginal fibroid polyp was considered for further treatment. Her Hb was 13gm, TLC 8.5/cumm, platelets 208/cumm and renal and liver function tests were within normal limits. After routine investigations, she was undertaken for examination under anaesthesia which confirmed a pedunculated mass 4*4 cm arising from anterior vaginal wall by a thick stalk for which polypectomy with excision at the base was done with 2mm margin of normal vaginal tissue. The cervix appeared healthy with no apparent growth or stenosis. Cervical smear was taken for

cytology. The anterior vaginal wall was repaired and packing done. The histopathology report was suggestive of sarcoma. Surgical margins were disease free. The cervical smear was negative for dysplasia. Immunohistochemistry was strongly positive for Caldesmon and Smooth muscle actin.

DISCUSSION

Sarcoma of the vagina is rare, recurrent malignancy with aggressive hematogenous spread. Prior pelvic radiotherapy appears to be a risk factor (1). Most patients with vaginal sarcoma present with vaginal asymptomatic mass with bleeding or discharge or rarely dyspareunia. Most sarcomas arise de novo with malignant transformation of the benign mesenchymal tumour being a very rare event. Nuclear atypia, mitotic index, and zonal necrosis are parameters used in the differentiation from leiomyomas (2). It is generally accepted that the presence of 10 or more mitoses per 10 high power fields indicates malignancy (2). Cytological atypia and necrosis further bolster the diagnosis (2). Further, if either nuclear atypia or epithelioid cells are present, 5 mitoses per 10 high power fields are sufficient for the diagnosis of leiomyosarcoma. Early stage at diagnosis and radical surgical resection with wide disease free margins offer best chance of cure (3). The role of adjuvant radiotherapy and chemotherapy is not clearly defined in vaginal sarcomas owing to the rare incidence. While surgery is considered the mainstay of treatment,

Ciarvino et al showed that there was no difference in survival between patients who had surgery alone and those who had surgery followed by adjuvant radiotherapy or chemotherapy (4). Same has been claimed by Ngan et al that neither chemotherapy nor radiotherapy were of use in the treatment of late-stage disease (5). At the same time Peters et al has emphasized on the benefit of radical procedures that have shown better long-term survival and recurrence free period (6), same has been supported in recent studies (7,8).

Histopathological grade is the best predictor of outcome. Early stage at diagnosis, young age and radical surgical resection with wide disease-free margins offer best chance of cure. MRI is a useful tool to follow up patients for recurrence and complications.

CONCLUSION

Sarcoma of the vagina is a rare but recurrent malignancy that has been sparsely defined. It is important that all cases be reported so that effective treatment modalities may be recognized for future. Role of adjuvant therapy is clearly indicated in young patients but data on long term survival is not available. Also more of prospective studies are needed to guide research for patients who may desire fertility.

Consent

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

Ethical Approval:

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

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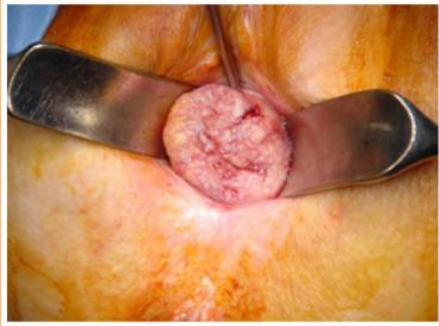


Fig 1:Gross image of mass

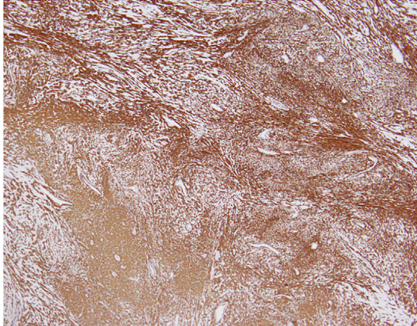


Fig: 2: Diffuse strong immunostaining of tumour cells with caldesmon

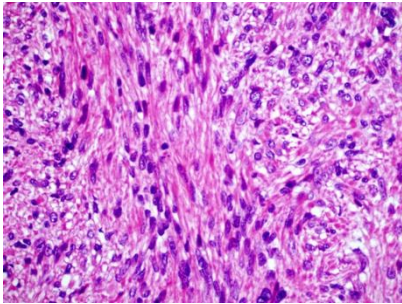


Fig: 3 Spindle to epithelioid morphology with marked pleomorphism and hyperchromatism