

Spindle Cell Sarcoma of Vagina: A Rare Case Report and Review of Literature

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Abstract

Primary malignancy of the vagina is unusual and sarcoma as the primary is rarer. The majority of sarcomas are diagnosed at an advanced stage. Histopathological grade is the most important factor to predict the outcome. Surgical resection is the main stay of treatment. The role of adjuvant radiotherapy and chemotherapy is controversial. An adjuvant radiotherapy should be given in the case of high-grade tumors, and post-operative surgical margin positive cases. Neoadjuvant chemotherapy should be given in locally advanced cases to decrease the tumor burden to make a possible wide local excision with wider margin. We reported a case of spindle cell sarcoma of the vagina in a 57-year-old woman who was treated with 3 cycles of neoadjuvant chemotherapy followed by surgical resection with adequate margin, and doing well on 6 months follow-up.

Key words: Neoadjuvant chemotherapy, Spindle cell sarcoma, Vagina, Wide local excision

INTRODUCTION

Malignant tumors of the vagina are extremely rare and accounts for 2% of all gynecological malignancies.¹ One of the rare varieties is spindle cell sarcoma of vagina having a poor prognosis. Newer diagnostic techniques, like immunohistochemistry, are now available to pick up even submicroscopic diagnosis of many pathological lesions. We report a case of spindle cell sarcoma which was successfully treated with neoadjuvant chemotherapy due to large size followed by wide local excision of the lesion, and adjuvant radiotherapy and chemotherapy.

CASE REPORT

A 56 years postmenopausal female presented as something coming out of vagina and pain in the periurethral region for last 2 months. The patient had history of undergoing

vaginal hysterectomy 10 years back for abnormal uterine bleeding. She had all vaginal deliveries with bilateral tubal ligation done 30 years back after her last childbirth. On examination, the patient was average built with stable vitals. Per abdomen examination found no abnormality. Per speculum examination showed a nodular growth of 2 cm × 2 cm in the sub urethral area in anterior wall of the vagina, firm in consistency, non-ulcerated, well-defined margin, not fixed to base, did not bleed on touch (Figure 1). On vaginal examination vault was healthy. Per rectal examination revealed soft pelvis.

Routine blood counts, urine examination, and chest X-ray findings were normal. Cytology from the vault lesion revealed inflammation. Ultrasound of abdomen and pelvis showed thickened urinary bladder wall with 117 ml residual urine. The transvaginal sonogram showed a hypoechoic mass of size 20 mm × 15 mm in anterior wall of the lower vagina and on Doppler the mass showed minimal vascularity of low resistance pattern (PS = 27.7 cm/s, ED = 6.1 cm/s) radiologically suggestive of malignancy. A biopsy was taken from growth and the histopathological study revealed features of high grade spindle cell squamous cell carcinoma of vagina with a differential diagnosis of spindle cell sarcoma of vagina (Figure 2). Immunohistochemistry was performed and revealed pan cytokeratin negative,

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vimentin strongly positive, desmin negative confirming diagnosis of spindle cell sarcoma (Figure 3). Further investigations ruled out distant metastasis. She was planned for surgery in the form of wide local excision with adequate margin. The patient did not come for further treatment and after 6 months of lost treatment, she reported with larger sized mass than the previous one. The treatment modality was changed, and the patient had received 3 cycles of neoadjuvant chemotherapy in the form ifosfamide and Adriamycin followed by wide local excision with adequate margins. The patient had received adjuvant radiotherapy followed by 3 cycles of adjuvant chemotherapy of the same regimen. Radiotherapy was given as standard field pelvic radiation with vaginal high dose radiation for a total dose of 66 GY. The patient was under regular follow-up since 9 months without disease recurrence or distant metastasis.

DISCUSSION

Primary cancer of vagina is rare and represents 1-4% of all genital malignancies. 75-90% is squamous cell type, 5-10% is adenocarcinoma, 3% are melanoma, and 3% are sarcomas.² Sarcomas are rare mesenchymal neoplasm that arise in soft tissues and bone. Primary sarcomas of the vagina are rhabdomyosarcoma, leiomyosarcoma, malignant fibrous histiocytoma, hemangiopericytoma, malignant schwannoma, endometrial stromal sarcoma, and fibrosarcoma. Up to 1985, only 68 cases of primary sarcoma of the vagina were reported. Vaginal sarcomas should be distinguished among themselves as to their precise pathogenetic origin using special stains, electron microscopy, and immunohistochemistry. Various immunohistochemical markers are used for confirmation of a given phenotype or differentiation or histogenesis. Sarcomas of the elderly are leiomyosarcoma, angiosarcoma, spindle cell sarcoma, alveolar soft part sarcoma, fibrosarcoma, and mixed mesodermal tumors of the vagina. Leiomyosarcoma is the most common type of sarcoma in the vagina.

Several factors have been demonstrated to play a role in recurrence including tumor diameter, cytologic atypia, mitotic index, and infiltrating margin. The majority of the cases presents as asymptomatic vaginal mass. The patient may present as a lump near introitus which may cause problems with micturition, defecation or intercourse. Bleeding or discharges are late features. Complete history and physical examination should be performed including speculum and per vaginal examination, cervical cytologic examination, endometrial biopsy when indicated, colposcopy and biopsy of the vaginal tumor. Pre-treatment evaluation may include the following studies: Chest x-ray, intravenous pyelogram, cystoscopy, proctosigmoidoscopy, contrast enhanced CT and MRI scan of the abdomen and



Figure 1: Small suburethral nodule

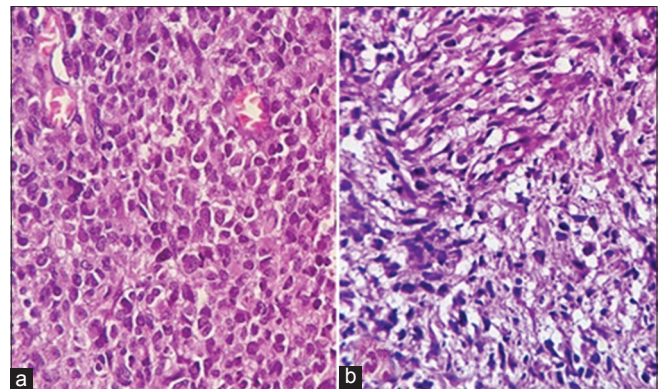


Figure 2: Histopathological examination (a) Malignant cells with nuclear atypia and high nuclear: Cytoplasmic ratio (b) Spindle shaped arrangement of cells. H and N $\times 400$

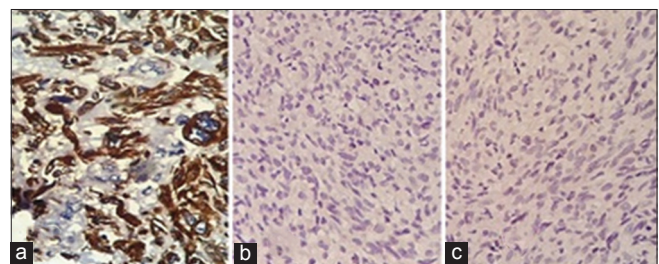


Figure 3: Immunohistochemistry study (a) Vimentin positive cells (b) Pan CK-negative cells, and (c) Desmin negative cells

pelvis.³ The treatment plan depends on the patient's age and general health state, the tumor location and size, the need to maintain the function of the vagina and stage of the disease. Prognosis is related to the stage of the disease (American Joint Commission on Cancer staging system for sarcomas).⁴ Although there are no specific treatment guidelines, the mainstay of therapy has been surgical excision followed by chemotherapy. Wide local excision with reconstructive surgery, vaginectomy with lymphadenectomy Radical hysterectomy, pelvic exenteration, and laser surgery⁵ are the different surgical options. Neoadjuvant chemotherapy

has been used in patients who have bulky tumors in which surgical debulking had not been optimal. Currently, some believe that neoadjuvant chemotherapy can also be used as primary therapy followed by surgical debulking regardless of the tumor size. It is thought that this treatment strategy can optimize surgical excision, giving rise to less morbidity, and a longer disease-free interval.⁶

The role of adjuvant radiotherapy and chemotherapy in primary vaginal sarcomas is unclear due to paucity of data. According to Hensley, surgical resection plus adjuvant radiotherapy should be given in the case of high-grade tumors and in the case of positive surgical margins to prevent local recurrence and also requiring chemotherapy in persistent or recurrent disease.⁷ Due to high risk of systemic relapse, chemotherapy has been utilized. A meta-analysis showed ifosfamide and Adriamycin based combination chemotherapy resulted in a reduction of death risk from 41% to 30%.⁸ Marginal efficacy of chemotherapy in terms of local recurrence, distant metastasis and overall survival found in localized respectable soft tissue sarcoma. A study showed both chemotherapy and radiotherapy had no effect on outcome in late or recurrent disease.⁹

CONCLUSION

Spindle cell sarcomas are rare malignant tumors of the vagina with poor prognosis. Most of the cases present with lump in the vagina with various dysfunctions. Recently modalities like electron microscopy and immunohistochemistry are most important tools used to

reach at a submicroscopic diagnosis. Due to paucity of the data, there is no standard treatment guidelines. Surgery with or without radiotherapy has resulted in improved outcome. Neoadjuvant chemotherapy followed by wide local excision is reserved for bulky lesions to facilitate surgery to get a good margin. A large number of case data in series or study with a longer duration of observation are necessary to draw a standard treatment guideline.

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