CASE REPORT

Adult Vaginal Sarcoma: When a Rarity becomes Reality

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Abstract

Primary malignant diseases of the vagina are quite uncommon, particularly in adults. They constitute about 2% of gynecologic malignancies, the least common among them being sarcomas (3%). Very few cases have been reported in the literature due to the rarity of the disease.

A 36-year-old lady presented with complaints of irregular and excessive bleeding per vaginum of 3-months duration. There were no specific findings on general examination of the patient. The per-speculum examination revealed an ulcero-proliferative growth arising predominantly from the anterior vaginal wall extending from the anterior and right lateral fornices till introitus, filling the vaginal cavity. The cervix and all other fornices were free, so was the rectum. The MRI of the local part showed a 6 × 4 cm lesion arising from upper anterior vaginal wall causing obliteration of the vaginal cavity with extra serosal extension and abutment of base of bladder. The biopsy and the immunohistochemistry of the lesion were suggestive of high-grade sarcoma (Vimentin+, CD10+, p53+, ki 67–80%, SMA-, Desmin-, myogenin-, and Pan CK-)

The patient received 3 cycles of neoadjuvant chemotherapy (AIM regimen – Doxorubicin, Ifosfamide, and Mesna) due to the bulky nature of the disease. The response assessment MRI revealed 25% decrease in the size of the lesion with infiltration of the posterior wall of the bladder. The patient underwent laparoscopic anterior pelvic exenteration. The postoperative histopathology revealed the growth to be high-grade fibrosarcoma with focal undifferentiated pleomorphic sarcoma-like areas with bladder wall infiltration (pT3).

Sarcomas of the vagina are exceedingly rare. Surgery plays a major role in management. Postoperative radiation therapy may help in decreasing local recurrence. The benefits of chemotherapy, with its morbidity and possible mortality, remain unproven. Prognosis may be associated with tumor grade and stage. Owing to the rarity of the disease, treatment guidelines are still unclear and need further exploration.

Keywords: Mesenchymal tumor, Sarcoma, Soft-tissue sarcoma, Vaginal sarcoma, Visceral sarcoma.

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Introduction

Primary benign and malignant tumors of the vagina are quite uncommon, particularly in adults. The malignant ones constitute about 2% of gynecological malignancies.[1,2] The most common histological type is squamous cell carcinoma (75–90%), followed by adenocarcinoma (5–10%), melanoma (3%), and the least common being sarcomas (3%). Leiomyosarcomas, endometrial stromal sarcomas, malignant mixed Müllerian tumors, and rhabdomyosarcomas are the major types of primary vaginal sarcomas.[1] Very few cases have been reported in the literature due to the rarity of the disease.

The most common clinical presentation of vaginal sarcoma is similar to that of vaginal carcinomas, such as vaginal bleeding, mostly post coital or intermenstrual/post menopausal.[3,4] The other symptoms like low back pain and urinary and fecal disturbances develop as the disease progresses to involve adjacent structures.

Vaginal malignancies may invade locally and disseminate by several routes: direct extension to adjacent structures and pelvic soft-tissue structures, lymphatic and hematogenous dissemination. The prognosis is depending on the stage of the disease at presentation and histological type.[5] The global poor survival rates are probably because of presentation in a relatively advanced stage and the potential for treatment complications that prevent aggressive therapy.

Case Description

A 36-year-old lady presented with complaints of irregular and excessive bleeding per vaginum of 3-months duration. The Patient was P1L1 with a 6-year-old son without any family history of malignancies. There were no specific findings on general examination of the patient. The per-speculum examination revealed an ulcero-proliferative growth arising predominantly from the anterior vaginal wall extending from anterior and right lateral fornices till introitus, filling the vaginal cavity. The cervix and all other fornices were free. There was no involvement of rectal mucosa. The MRI of the local part showed a 6 × 4 cm lesion arising from upper anterior vaginal wall causing obliteration of the vaginal cavity with extra serosal extension and abutment of base of bladder (Fig. 1). The biopsy and the immunohistochemistry of the lesion were suggestive of high-grade sarcoma (Vimentin+, CD 10+, p53+, ki 67–80%, SMA-, Desmin-, myogenin-, and Pan CK-) (Figs 2 and 3).

The patient received 3 cycles of neoadjuvant chemotherapy (AIM regimen – Doxorubicin, Ifosfamide, and Mesna) due to the bulky nature of the disease. The response assessment MRI revealed 25% decrease in the size of the lesion with infiltration of the posterior wall of the bladder. The patient underwent laparoscopic anterior pelvic exenteration. The postoperative histopathology

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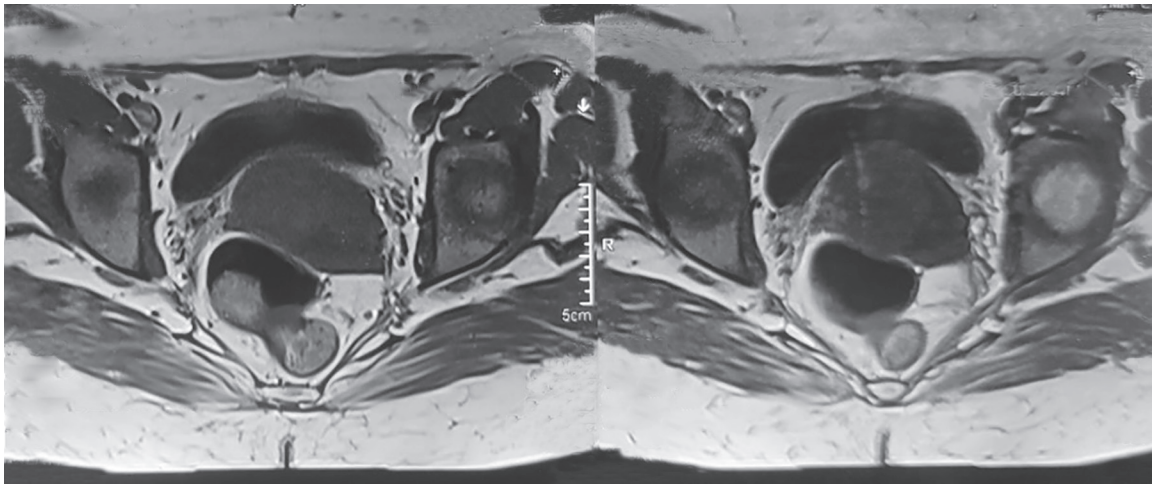

Fig. 1: Post-neoadjuvant chemotherapy MRI showing residual disease

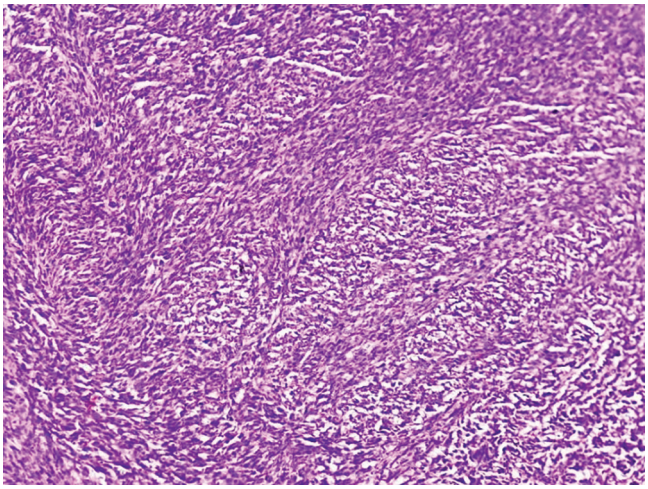

Fig. 2: Microscopic appearance

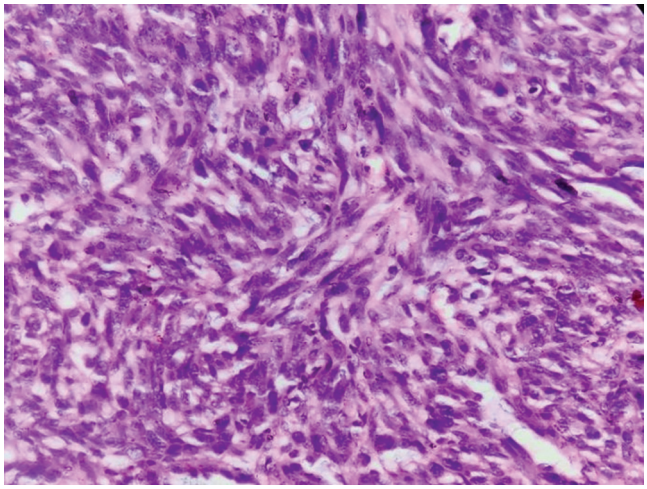

Fig. 3: Microscopic appearance

revealed the growth to be high-grade fibrosarcoma with focal undifferentiated pleomorphic sarcoma-like areas with bladder wall infiltration (pT3).

Unfortunately, the patient succumbed to chemotherapy-related complications (acute kidney injury) while on adjuvant therapy after completing 2 cycles in the postoperative period.

Discussion

In the adult female, vaginal sarcoma is a very rare oncological diagnosis among vaginal malignancies accounting for 2%.[1] The retrospective studies show a wide range of ages from 17 to 60, but since squamous cell carcinoma of the vagina occurs at a relatively older age, sarcoma should be taken into account when a younger patient presents with vaginal mass.[6] The staging is clinical based on the findings from physical examination, colposcopy, cystoscopy, proctoscopy, and imaging, which includes MRI local part to evaluate the extent of the disease and a CT chest to rule out metastasis. Owing to the rarity of the disease, there is no consensus on metastatic workup, but 2021 update of FIGO on vaginal cancer recognizes PET CT to be the most sensitive in detecting nodal metastasis. It should also be kept in mind that these recommendations are primarily formed for vaginal squamous cell carcinomas as they account for 90% of vaginal cancers.[5,7] Once again because of the rarity of the disease, there is no common consensus on its treatment. The recommendations are mostly the extrapolation of treatment guidelines for uterine sarcomas and to some extent vaginal cancers. A review article of vaginal sarcomas in a single institution in China discussed 8 cases of primary vaginal sarcomas with the mainstay treatment being surgery.[6] In this cohort, there was significant variation in surgery (TAH BSO, local excision), chemotherapy (regimen), and radiotherapy (EBRT and Brachy). The authors recognized that the majority of patients had vaginal leiomyosarcoma and others were endometrial stroma sarcoma, undifferentiated sarcoma, and adenosarcoma.

Surgery is the main treatment for vaginal sarcoma.[6,8] Common surgical procedures ever reported include wide local excision and pelvic exenteration depending on tumor size and invasive depth. For tumors at the upper one-third of the vagina, a radical

hysterectomy may be necessary. Surgery was not an attractive option for a long time due to the proximity to bladder, urethra, and rectum requiring multivisceral resection. Tavassoli and Norris recommended aggressive surgery for tumors likely to recur, which may have the following risk factors: tumor size ≥3 cm, irregularity of contour, histologic cellular atypia, and ≥5 mitoses/10 HPF.[9] Peters et al. believed that only patients who underwent pelvic exenteration (including anterior and posterior exenteration) could achieve long-term survival.[8] With the improvement of surgical techniques, instruments, perioperative care, antibiotic availability, and patient selection, morbidity and perioperative mortality rates of pelvic exenteration have decreased over time, and now exenteration is considered as a suitable therapeutic option for young and fit patients.[8,10] The role of adjuvant chemotherapy and radiotherapy remains unclear. Many studies suggest that there is not much of survival benefit from adjuvant chemotherapy, while some show increase in local control with radiation but no effect on overall survival.[6] Hormonal therapy is reported to have a favorable survival outcome in endometrial stromal sarcomas if they express ER/PR.[11] The 5-year survival of vaginal sarcoma varies from 35% to 70%.[6] The factors that may predict recurrence and affect prognosis are tumor dimension greater than 5 cm, significant cytologic atypia, mitotic index > 10 figures per 10 HPFs, presence of atypical mitotic figures, presence of tumor cell necrosis, and pattern of tumor interface.[12]

Conclusion

Sarcomas of the vagina are exceedingly rare. Surgery plays a major role in management. Postoperative radiation therapy may help in decreasing local recurrence. The benefits of chemotherapy, with its morbidity and possible mortality, remain unproven. Prognosis may be associated with tumor grade and stage. Owing to the rarity of the disease, treatment guidelines are still unclear and need further exploration.

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