JOGECA Journal of Obstetrics and Gynaecology of Eastern and Central Africa

CASE REPORT

Gynecology

Vaginal leiomyosarcoma: A case report and literature review

Alinoor B.O[1*], Kibunja J.V.K[1], Mokomba A.M[2]

[1] Department of Obstetrics and Gynecology, University of Nairobi, Nairobi, Kenya.
[2] Department of Obstetrics and Gynecology, Kenyatta National Hospital, Nairobi, Kenya.

***Correspondence**: alinoorb@gmail.com

Received: 6 May 2021; Revised: 12 June 2021; Accepted: 18 June 2021; Available online: July 2021

Abstract

Background: Vaginal leiomyosarcomas are extremely rare. Approximately 288 cases have been reported in the literature.

Case presentation: A 60-year-old presented to the gynecological ward as a referral with a diagnosis of vaginal leiomyosarcoma for chemoradiation. She had a history of per vaginal discharge, bleeding, and a posterior vaginal wall mass. Wide local excision of the mass was done at the peripheral facility. The surgical margin was 4 mm and negative for malignant cells. The patient was started on paclitaxel and cisplatin. She underwent 25 sessions of external beam radiotherapy and two sessions of brachytherapy. She is currently on follow-up through the gynecological-oncology clinic.

Conclusion: Surgical excision is the primary mode of treatment for vaginal leiomyosarcoma. Adjuvant chemoradiation following surgery has a role in reducing local recurrence and systemic relapse but with no appreciable effect on advanced disease.

Keywords: vaginal leiomyosarcoma, surgery, chemotherapy, radiotherapy

Introduction

Vaginal carcinomas are sporadic, accounting for approximately 2% of gynecological malignancies (1). The common histological types include squamous cell carcinoma (75-90%), adenocarcinoma (5-10%), melanoma (3%), and sarcomas (3%). Leiomyosarcomas are the most common type of vaginal sarcomas in adult women (2), whereas rhabdomyosarcomas are the most common in children and adolescents (3). Leiomyosarcomas originate mainly from the posterior vaginal wall smooth muscle cells and occasionally from tissues near the vagina (4). A diagnosis of leiomyosarcoma is made histologically based on mitotic activity and cellular atypia (5). Nuclear atypia, mitotic index, and zonal necrosis are used to differentiate leiomyosarcomas from leiomyomas. African American women are twice as likely to develop leiomyosarcoma than Caucasian women (4). This is a case of a 60-year-old diagnosed with vaginal leiomyosarcoma. She was surgically managed and started on chemotherapy and radiotherapy.

Case presentation

A 60-year-old, Para 3+0, presented to the gynecological ward at the Kenyatta national hospital (KNH) as a referral with a diagnosis of vaginal leiomyosarcoma for chemoradiation. A total vaginal hysterectomy was done at a peripheral facility due to abnormal uterine bleeding 15 years ago. She had type 2 diabetes and was managed on metformin 500mg twice daily and glibenclamide 10mg twice daily with good glycemic control. She had a history of per vaginal discharge, bleeding, and a posterior vaginal wall mass. The mass was

exophytic, non-friable, bled easily on touch, and measured approximately 10cm in diameter with a pedicle attached to the posterior vaginal wall towards the fornix. Wide local excision (WLE) of the mass was done at the pedicle. The surgical margin was 4mm and negative for malignant cells. The tumor was heterogeneous with hyper and hypocellular areas and spindle cells exhibiting moderate to marked pleomorphism arranged in broad sweeping fascicles, with a mitotic index of >20/10 high power field (HPF), bizarre mitosis, and necrosis accounting for <50% without lymph vascular invasion. An immunochemical assay reported diffuse strong positivity for the smooth muscle actin (SMA), consistent with leiomyosarcoma. Other requested assays included S100 protein, melan-A, myogenin, and cytokeratin AE1/3; however, the results were not available.

On admission, the patient was in good performance status. Her blood pressure (BP) was 136/84 mmHg, pulse rate 71 beats per minute (BPM), respiratory rate (RR) 20 BPM, and temperature 36.6 °C. Her random blood sugar (RBS) was 5.4 mmol/l. The vaginal vault was normal. The total blood count, renal and liver function tests were normal. The vaginal vault pap smear was negative for intraepithelial neoplasm or malignancy. Her serum cancer antigen (CA-125) level was 7.09 U/mL. The abdominal and pelvic computerized tomography (CT) scan was negative for residual disease. The chest radiography was negative for lung metastasis. The patient was started on adjuvant chemoradiation. She was put on paclitaxel and cisplatin followed by radiotherapy due to the prohibitive cost of the standard of care. At the writing of this case report, she had completed six cycles of chemotherapy, 25 sessions of external beam radiotherapy, and two sessions of brachytherapy, with a good response. She is currently on follow-up through the gynecological-oncology clinic.

Discussion

Smooth muscle tumors are the most common benign and malignant mesenchymal tumors among adult women (2). Vaginal sarcomas arise mainly submucosal and can be from any part of the vagina. The benign and malignant tumors are primarily seen in the anterior and posterior vaginal walls, respectively (1). The primary sarcomas of the vagina include rhabdomyosarcoma, leiomyosarcoma, malignant fibrous histiocytoma, hemangiopericytoma, malignant schwannoma, endometrial stromal sarcoma, fibrosarcoma, alveolar soft part sarcoma, and angiosarcoma (7). Vaginal leiomyosarcomas are extremely rare. Approximately 288 cases have been reported in the literature (1,2,7). Vaginal leiomyosarcomas may present with vaginal bleeding or discharge, vaginal mass, dyspareunia, perineal pain, and difficulty in micturition (2). There may be a delay in its diagnosis, especially when the malignancy involves distal parts of the vagina or vulva resembling a Bartholin cyst (4), Gartner's duct cyst, granuloma, or epithelial inclusion cysts (5).

The majority, 80-90%, of primary vaginal cancers, result from metastasis, mainly from the cervix and vulva. Therefore, to diagnose primary vaginal cancer, the absence of clinical and histological diagnosis of cervical and valvular malignancy within the last 5-10 years is required (3). Other cancers that metastasize to the vagina include gestational trophoblastic neoplasia and endometrial adenocarcinoma (5). Vaginal leiomyosarcomas spread via local invasion and lymphatic or hematogenous route, and therefore, are associated with both local recurrence and distant metastasis (8). Tumors containing >5 mitoses/10 HPF are locally aggressive and occasionally give rise to distant metastasis (7). In this case, the mitotic index was >20/10 HPF though no evidence of metastasis was demonstrated from the workup done.

The treatment of vaginal leiomyosarcomas depends on the age and health status of the patient, the location and size of the tumor, disease stage, and the need to maintain the function of the vagina (7). The primary mode of treatment for an early lesion is surgical excision ensuring disease-free margins, followed by chemotherapy and radiotherapy (8). Other surgical options include wide local excision with reconstructive surgery, vaginectomy with lymphadenectomy, and laser surgery (7). The predictors of survival are the degree of tumor differentiation, the patient's age, positive surgical margins, and lymph node involvement. Higher clinical stage at diagnosis, poor differentiation, lymph node involvement, and positive margins are associated with poor prognosis (9). Early recurrence of cancer is seen in cases with a tumor size ≥3 cm, irregular tumor contour, cellular atypia, ≥5 mitoses/10 HPF, and poorly differentiated lesions (3). A younger age, low stage at diagnosis, surgical resection are associated with a better prognosis than chemotherapy or radiotherapy (2).

The role of adjuvant radiotherapy and chemotherapy is not well defined due to the paucity of data and rarity of the condition. However, adjuvant radiotherapy and chemotherapy are frequently used to prevent local recurrence and systemic relapse in patients with high-grade vaginal leiomyosarcomas, low-grade recurrent tumors, and those whose tumors extend beyond the surgical margins (2). Radiotherapy and chemotherapy may not improve the survival outcome and have no appreciable effect on

advanced disease (10). The standard adjuvant chemotherapy regimen for sarcomas include docetaxel plus gemcitabine, doxorubicin, dacarbazine, doxorubicin plus dacarbazine, doxorubicin plus ifosamide, gemcitabine plus Dacarbazine and gemcitabine and Vinorelbine (6). The gynecological tumor boards are essential in rare conditions like vaginal leiomyosarcoma (9,11). The tumor board discussed the patient and was recommended for chemotherapy followed by radiotherapy.

Conclusion

Surgical excision is the primary mode of treatment for vaginal leiomyosarcoma. Adjuvant chemoradiation following surgery has a role in reducing local recurrence and systemic relapse but with no appreciable effect on advanced disease.

Consent for publication

Informed consent for publication was obtained from the patient.

Acknowledgement

The authors acknowledge the patient for consenting to the publication of this case report.

Declarations

Conflict of interests

The authors declare no conflicts of interest.

Funding

None

References

1. Yordanov A. A Rare Case of Vaginal Leiomyosarcoma. J Tumor Med Prev. 2018;2(1):2–5.
2. Khosla D, Kumar R, Nijhawan R, Patel F, Sharma S, Gowda K. Leiomyosarcoma of the vagina: A rare entity with a comprehensive review of the literature. Int J Appl Basic Med Res. 2014;4(2):128.
3. Okunowo AA, Ugwu AO, Owie E, Kolawole HF, Adebayo LA, Kusamotu OA, et al. Primary vaginal leiomyosarcoma: A case report of a rare gynaecological malignancy and diagnostic challenge in a resource-constraint setting. Oxford Med Case Reports. 2020;2020(9):341–3.
4. Keller NA, Godoy H. Leiomyosarcoma of the Vagina: An Exceedingly Rare Diagnosis. Case Rep Obstet Gynecol. 2015;2015(3):1–4.
5. Ahram J, Lemus R, Schiavello HJ. Leiomyosarcoma of the vagina: Case report and literature review. Int J Gynecol Cancer. 2006;16(2):884–91.
6. Köhler G. Uterine sarcoma treatment. Pathologe. 2009;30(4):304–12.
7. Parikh, A., Suryanarayana, U., Agrawal, P. and Koladiya, J., 2018. A CASE REPORT ON SPINDLE CELL SARCOMA OF VAGINA. PARIPEX - INDIAN JOURNAL OF RESEARCH, 7(3), pp.218-219.
8. Zakashansky K, Peiretti M, Mahdavi A, Chun JK, Nezhat F. Combined laparoscopic and radical vaginal treatment of primary vaginal leiomyosarcoma in a patient with a unicornuate uterus and pelvic kidney. J Minim Invasive Gynecol. 2007;14(4):518–20.
9. Patel D, Handorf E, Von Mehren M, Martin L, Movva S. Adjuvant chemotherapy in uterine leiomyosarcoma: Trends and factors impacting usage. Sarcoma. 2019;2019.
10. Li Y, Ren H, Wang J. Outcome of adjuvant radiotherapy after total hysterectomy in patients with uterine leiomyosarcoma or carcinosarcoma: A SEER-based study. BMC Cancer. 2019;19(1):1–9.
11. Specchia ML, Frisicale EM, Carini E, Di Pilla A, Cappa D, Barbara A, et al. The impact of tumor board on cancer care: Evidence from an umbrella review. BMC Health Serv Res. 2020;20(1):1–14.