ORIGINAL RESEARCH PAPER

Oncology

KEY WORDS:

A CASE REPORT ON SPINDLE CELL SARCOMA OF VAGINA

Dr Ankita Parikh Gujarat Cancer & Research Institute, Ahmedabad.

Dr U.Suryanarayana* Gujarat Cancer & Research Institute, Ahmedabad. *Corresponding Author

Dr. Prerak Agrawal Gujarat Cancer & Research Institute, Ahmedabad.

Dr.Saheli Saha Gujarat Cancer & Research Institute, Ahmedabad.

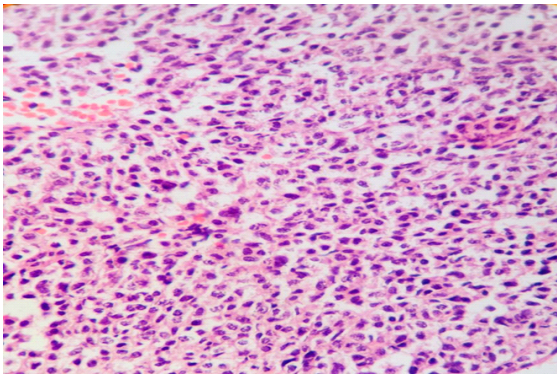
Dr.Jagruti Koladiya Gujarat Cancer & Research Institute, Ahmedabad.

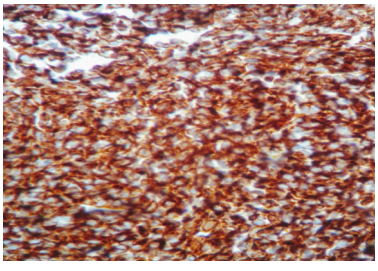
Primary cancer of vagina is rare and represents 1-2% of all genital malignancies and vaginal sarcomas are even rarer. Sarcomas account for only 3.1% of all vaginal malignancies . Primary vaginal sarcomas are aggressive neoplasms with different presenting characteristics and inferior survival out- comes when compared to epithelial vaginal tumors. We report a case of spindle cell sarcoma of vagina due to its rarity

Case report

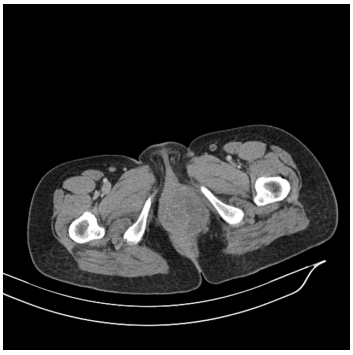
A 45year old female presented to our outpatient department after being referred from another hospital. She had a swelling in her vulva for which she was provisionally diagnosed as having bartholin cyst and underwent excision of the lesion at that hospital, one month before and subsequent histopathological examination and Immunohistochemistry was diagnostic of undifferentiated high-grade sarcoma. On presentation, she had the complaint of swelling on left side of vulva. General condition was fair with stable vitals.

On examination, no nodes were palpable. Per abdomen examination revealed no abnormality. On per vaginal examination a mass was palpable in left paravaginal space extending till left fornix. On per rectal examination left parametrium was involved upto one finger short of lateral pelvic wall. Immuno histochemistry at our institute was diagnostic of spindle cell sarcoma [AE1 / EMA / Desmin -negative, vimentin- positive].





CT scan of abdomen & pelvis was showing 6.2 5.4 7.5 cm3 lesion left lateral vaginal wall with extension into ischiorectal fossa and left parametrial region and abuts anal canal and levator ani muscle. Few small inguinal nodes [largest 1.3 1.1 cm], multiple lung metastases were also seen. Baseline blood investigations were normal. Decision was made to start with chemotherapy first.



After 2 cycles of Ifosfamide & Adriamycin, stable disease was evident radiologically. Hence further 2 cycles of Ifosfamide & Adriamycin were given. Post chemo CT scan showed 4.5 3.5 4.8 cm3 lesion of left lateral vaginal wall with extension to ischiorectal fossa and abutting anal canal and left levator ani muscle. B/L few small inguinal nodes [largest 1.2 0.8 cm] and lung metastasis were present. As per surgical oncology opinion, patient was clinically inoperable. Hence, she was given palliative radiotherapy 30Gy/10fractions. As good clinical response was seen, she was further given an additional radiation of 20 Gy/10 fractions. On clinical examination after external radiation there was smooth growth over left posterolateral vaginal wall upto 2 cm from introitus. Cervix/ fornices and upper vagina were free. On per rectal examination both parametrium were free. USG abd/pelvis showed 5.4 3.6 cm metastatic lesion in liver, 4.2 2.2 cm2 mass lesion left lateral vaginal wall. Hence decision was made to give best supportive care to the patient and she expired at home 4 months later.

Discussion

Primary cancer of vagina is rare and represents 1-2% of all genital malignancies. 82.6 are squamous cell type, 9.6% are adenocarcinoma 3.1% are sarcomas. Sarcomas are rare mesenchymal neoplasms that arise in soft tissues and bone. These tumors are usually of mesodermal origin, a few are derived from neuroectoderm. Primary sarcomas of the vagina are Rhabdomyosarcoma, Leiomyosarcoma, Malignant fibrous histiocytoma, Haemangiopericytoma, Malignant schwannoma, endometrial stromal sarcoma, Fibrosarcoma, Alveolar soft part sarcoma, and Angiosarcoma'. Most of the cases present as asymptomatic vaginal mass. When sarcomas occur in Bartholin's

gland area, they can be mistaken for Bartholin's gland cyst, delaying the diagnosis and worsening the prognosis .The patient may present as a lump near introitus which may cause problems with micturition, defecation or intercourse. Abnormal vaginal Bleeding or discharge 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 tumour–. Vaginal sarcomas should be distinguished among themselves as to their precise pathogenetic origin by using special stains, electron microscopy, and immunohistochemistry. Various immunohistochemical markers are used for confirmation of a given phenotype or differentiation or histogenesis. Tumors containing >5 mitosis/10HPF are locally aggressive and occasionally give rise to distant metastasis. Pretreatment evaluation may include the following studies: chest x-ray, intravenous pyelogram, cystoscopy, procto-sigmoidoscopy and CT, MRI scan of the abdomen and pelvis.

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). Although there are no specifc treatment guidelines, the mainstay of therapy has been surgical excision followed by chemotherapy and/or radiotherapy. Wide local excision with reconstructive surgery, vaginectomy with lymphadenectomy Radical hysterectomy, pelvic exenteration, and laser surgery are the different surgical options. Neoadjuvant chemotherapy

may be considered in patients with unresectable tumors .
Primary chemotherapy followed by surgical debulking regardless of the tumor size has also been tried. The role of adjuvant radiotherapy and chemotherapy in primary vaginal sarcomas is unclear due to paucity of data. 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. Due to high risk of systemic relapse, chemotherapy has been utilized. Ifosfamide and Adriamycin based combination chemotherapy resulted in a reduction of death risk from 41% to 30% . How-ever survival outcomes still remains poor.

Conclusion

Vaginal spindle cell sarcoma is a rare tumour with poor prognosis. Due to paucity of the data, there is no standard treatment guidelines. A large number of case data in series or study with a longer duration of observation are necessary to draw a standard treatment guideline.

References

1. Berek JS, Hacker NF, editors. Berek & Hacker's gynecologic oncology. Sixth edition. Philadelphia: Wolters Kluwer; 2015. 960 p.
2. Mills SE. The vulva and the vagina. Mills SE,editors. Sternberg's Diagnostic surgical pathology.Vol-1. 4th ed.Newyork. Lippincot:cl.1992.P.2364-5
3. Vulvar leiomyosarcoma in Bartholin's gland area: a case report and literature review F. González-Bugatto M. J. Añón-Requena M. A. López-Guerrero J. M. Báez-Perea J. L. Bartha B. Hervías-Vivancos
4. Sankareswari R. SPINDLE CELL SARCOMA OF VAGINA-A CASE REPORT. J Evol Med Dent Sci [Internet]. 2013 [cited 2017 Sep 6];2(17):2882–2889. Available from: http://jemds.com/data_pdf/sankareshwari.pdf
5. Coppleson M.Gynaecological oncology.2nd ed .London: CL.1992.P.435-436.
6. Shreyaskumar R.Patel, Robuk. S. Benjamin. Softtissue and Bone sarcoma and Bone metastasis. Dan L.Lango,editor. Harrison's haematology and oncology. Newyork: Mc Graw Hill; 2010. p .540-3.
7. Collini P, Sorensen PHB, Patel S, Blay J-Y, Issels RD, Maki RG, et al. Sarcomas With Spindle Cell Morphology. Semin Oncol [Internet]. 2009 Aug [cited 2017 Sep 6];36(4):324–37.Availablefrom: 'http://linkinghub.elsevier.com/ retrieve/pii/S0093775409001079
8. Pervaiz N, Colterjohn N, Farrokhyar F, Tozer R, Figueredo A, Ghert M. A systematic meta-analysis of randomized controlled trials of adjuvant chemotherapy for localized resectable soft-tissue sarcoma. Cancer [Internet]. 2008 Aug 1 [cited 2017 Sep 6];113(3):573–81. Available from: http://doi.wiley.com/10.1002/cncr.23592