Case Report

An unusual case of cervical leiomyosarcoma in an adolescent girl

Sangeeta Pahwa*, Arshdeep Kaur, Preet K. Kaur, Tavleen Kaur

Department of Obstetrics and Gynecology, Sri Guru Ramdass Institute of Medical Sciences and Research, Amritsar, Punjab, India

Received: 31 August 2020
Accepted: 09 October 2020

*Correspondence:
Dr. Sangeeta Pahwa,
E-mail: sangeetapahwa139@gmail.com

Keywords: Cervical leiomyosarcoma, Adolescent age group, Management, Reproductive outcome, Ovarian preservation, Uterine transplant

ABSTRACT

Primary leiomyosarcoma of the uterine cervix is an exceedingly rare tumor. It is malignant mesenchymal tumour of smooth muscle origin. We report a case of 15-year-old adolescent girl who presented with complain of excessive bleeding per vaginum off and on since 3-4 months and was diagnosed as leiomyosarcoma of cervix. Patient was advised further management but father refused as according to them when bleeding has stopped completely then what is the need of further treatment. In this article we want to highlight the points: what is prognosis and survival of such patient whose mass was removed completely and patient is not willing for further treatment inspite of knowing the aggressive nature of tumor and if hysterectomy is the answer what about the reproductive outcome in such young girls and what is the prospect of uterine transplant surgery.

INTRODUCTION

Leiomyosarcoma is one of the rare histopathological variants of tumours of the cervix comprising nearly 1% of overall tumours of the cervix. The most common symptom is discharge per vaginum, bleeding per vaginum, pain in lower abdomen. The tendency to metastasize is high especially to lungs, peritoneum, bones, liver. It is aggressive tumor with poor prognosis. Gross appearance is a large poorly circumscribed mass often protruding from the cervical canal expanding it circumferentially. Microscopy shows presence of tumor cells arranged in sheets and intersecting fascicles.

The cells are oval to spindle shaped with hyperchromatic, pleomorphic nuclei with eosinophilic cytoplasm and increased mitotic activity. Management plans differ, according to presentation, histopathology and extent of metastasis.

CASE REPORT

A 15-year-old girl presented with complaint of excessive bleeding per vaginum off and on since 3-4 months. Patient sought treatment elsewhere where symptomatic treatment was given but bleeding did not stop.

MRI of pelvis done which showed polypoidal mixed intensity soft tissue mass with cystic areas of approximate size 85x66x54 mm seen in vagina. A T2 hypointense soft tissue intensity stalk like structure noted extending from posterior lip of cervix into the vaginal mass (Figure 1).

As patient was anemic 2 packed red blood cells were transfused. Her other routine investigations and chest Xray was normal. Examination under anaesthesia done, 11x10 cm degenerated, edematous and necrotic mass seen arising from the posterior lip of cervix and sent for histopathology (Figure 2).
A polypoidal mixed intensity mass with cystic areas within it.

A pedicle on the posterior lip of cervix noted, removed and bleeding points cauterized. Her uterus was nulliparous and vaginal fornices were clear. Histopathology report showed leiomyosarcoma of cervix (Figure 3).

Figure 1: A polypoidal mixed intensity mass with cystic areas within it.

Figure 2: Degenerataed, edematous, necrotic mass removed from posterior lip of cervix.

Figure 3: Tumour cells arranged in sheets and intersecting fascicles. These cells are oval to spindle with hyperchromatic, pleomorphic nuclei and moderate amount of eosinophilic cytoplasm.

After going through the literature about the retrospective study done by Jayaram VK, Parikshith J, Narayanan GS et al multimodality management of leiomyosarcoma of the cervix, 2018; further treatment was advised but father refused and lost to follow up.

DISCUSSION

Leiomyosarcoma is an uncommon malignancy accounting for approximately 1% of uterine cancer with an estimated annual incidence of 0.64 per 10,000 women. Although leiomyosarcoma can occur anywhere in pelvis including cervix and urinary bladder, it is more commonly found in uterus. It represents 0.21% among all invasive tumours of the uterine cervix. Primary leiomyosarcoma from uterine myometrium extending to the cervix has to be excluded. Diagnosis before surgery is a rare occurrence. But in our case whole of the mass was removed vaginally and definitive diagnosis was made on histopathology. Differentiation from cervical fibroid is difficult unless supported by histopathology and immunohistochemical marker. Various morphological varieties of leiomyosarcoma of the cervix myxoid type, epithelioid type, conventional type and certain variants contain abundant xanthomatous cells. Cervical leiomyosarcoma are aggressive tumour with high rates of recurrences. Local therapy consists of total hysterectomy. Casanova et al reported a case of localised leiomyosarcoma of the cervix in a 63-year-old woman who presented with lung metastasis after 1 year of diagnosis. There are various prognostic factor including tumour stage, grade, mitotic index, tumour size, age and menopausal status. Amongst these factors, stage, grade and mitotic index were more significant prognostic factors.

An 18-year-old female gravida 3 para 1011 presented to an outside clinic with several months of vaginal bleeding and abdominal pain. She was diagnosed with an intrauterine pregnancy with an estimated gestational age (EGA) of 27 weeks based on dating by ultrasound. She was also noted to have an exophytic mass protruding from the endocervical canal. Subsequent examination under anesthesia revealed a 4.2x3.9 cm, pedunculated mass, arising from the endocervical canal. The mass was resected and pathology was consistent with a leiomyosarcoma.

Signs and symptoms are bleeding per vagina, discharge per vagina, pain and fullness in the pelvic and abdominal region. Most of the tumours are bulky at the time of presentation ranging from 10 to 12 cm in size. Pressure symptoms such as increased frequency of micturition and sometimes urinary retention can also be seen.

Since it is a rare disease, there is no definitive guideline for management of leiomyosarcoma of the cervix. Most recommended treatment is multimodality comprising surgery, total abdominal hysterectomy with bilateral salpingo-oophorectomy in patients having disease confined to the cervix, followed by post-operative radiation therapy to reduce the incidence of local recurrences.
recurrence. In view of the low incidence of lymph node involvement in leiomyosarcoma of the cervix, the role of prophylactic pelvic lymph node dissection along with primary surgery is debatable, unless there is gross pelvic lymph node involvement. The role of chemotherapy has been extrapolated from sarcomas of the uterine corpus; most commonly used drugs including doxorubicin and ifosfamide. This regimen has an overall response rate of 30% in advanced and metastatic forms of disease. Other chemotherapy drugs used are cyclophosphamide, gemcitabine and docetaxel used with response rates of approximately 30%.

Ovarian involvement in sarcomas of the cervix, in particular to leiomyosarcoma of the cervix, is a very rare mode of presentation. In view of the rarity of ovarian involvement, the role of oopherectomy in sarcomas of the uterine cervix is a debatable issue. As similar to the other histological variants of sarcoma with some exceptions, the mode of spread to distant sites is through the blood. Lung is the most common site of metastasis.

**CONCLUSION**

In view of rarity of leiomyosarcoma cervix there is lack of guidelines for its management. Surgery is the main modality of treatment. Radiotherapy in the form of external beam irradiation and intravaginal brachytherapy improves the locoregional control via decreasing the local recurrence. But we are worried about the life expectancy and prognosis of patient as being asymptomatic afterwards and parents are illiterate not willing for further treatment. If hysterectomy is the modality of choice then we can think for about the of ovarian preservation and uterine transplant. If radiotherapy is treatment of choice for local recurrence then we can go for oocyte retrieval before radiotherapy.

**Funding:** No funding sources  
**Conflict of interest:** None declared  
**Ethical approval:** Not required

**REFERENCES**