

An Unusual Case of Cervical Leiomyosarcoma with Uterine Prolapse: A Case Report

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Abstract: Background: Cervical leiomyosarcoma is very uncommon when compared to uterine LMS. They comprise less than 1% of all cervical malignancies, thought to follow an aggressive course. Most patients present with vaginal bleeding and a bulky pelvic mass at the time of diagnosis. Macroscopically, the tumours are typically large and poorly circumscribed. Case Report: Here, we describe a case of a postmenopausal woman diagnosed with an early stage (2a) cervical leiomyosarcoma with acute onset uterine prolapse. Local examination revealed a necrotic gangrenous exophytic mass at vulva measuring 7*5*6 centimetres which was biopsied and sent for histopathology which revealed a spindle cell sarcoma suggestive of cervical leiomyosarcoma. Her LDH levels were elevated. Abdominal panhysterectomy was done for this patient. The growth which was arising from the posterior lip of cervix was excised vaginally to reduce the bulk and the rest of the procedure was completed abdominally. The frozen section report revealed a high grade leiomyosarcoma of the cervix. The postoperative recovery of the patient was uneventful and the patient was planned for combination chemo radiotherapy. Conclusion: MRI with LDH levels is 100% sensitive in diagnosing leiomyosarcoma. Combined modality of treatment including surgery, radiotherapy and chemotherapy improved the survival rate.

Keywords: spindle cell sarcoma, LDH, gangrenous mass

1. Introduction

Cervical sarcomas are rare neoplasms thought to follow an aggressive course. They comprise less than 1% of all cervical malignancies (1). Among the women with cervical sarcomas, carcinosarcoma was the most common (41%), while adenosarcoma and leiomyosarcoma accounted for 21% each (2). Most patients present with vaginal bleeding and a bulky pelvic mass at the time of diagnosis. Leiomyosarcoma of the cervix occur mostly in premenopausal women. In this case, we describe a postmenopausal woman diagnosed with an early stage (2a) acutely prolapsed cervical leiomyosarcoma.

2. Case

A 55 year old postmenopausal woman presented to our emergency room with complaints of postmenopausal bleeding for the past 1 month and something coming out per vaginum since 3 days. She had complaints of difficulty in passing urine since 3 days and was referred from GCRI with a urinary catheter in situ supposing it to be an acute uterine inversion.

Her physical examination was consistent with a normally developed elderly woman, normotensive with a BMI of 30. Pelvic examination revealed a soft abdomen. Per speculum revealed a necrotic gangrenous exophytic mass at vulva measuring 7*5*6 centimetres (fig 1). A biopsy was taken and sent for HPE. Per vaginal examination showed a uterus deviated to left and the rectal mucosa was free with no parametrial involvement on bimanual rectovaginal examination and was clinically staged as stage 2A according to the International federation of obstetrics and gynaecology.



Figure 1: A necrotic gangrenous mass at vulva measuring 7*5*6 centimetres with a urinary catheter in situ

Her USG revealed a bulky uterus measuring 12.2*6.8*7.2cm with a combined endometrial thickness of 8mm. A large predominantly necrotic mass lesion was noted in the uterine cervix extending to the posterior aspect of lower uterine segment measuring 7.7*6.2*7.2 cms. Her MRI revealed a large abnormal soft tissue mass noted in the cervix extending inferiorly into the upper half of vagina with no significant regional infiltration or parametrial invasion noted. No lymph nodes were noted with minimal free fluid in the peritoneal cavity, suggestive of malignant neoplasm carcinoma of cervix stage 2a disease.

Her biopsy report revealed predominantly haemorrhagic / necrotic tumour composed of fascicles of spindle cells with marked nuclear atypia s/o spindle cell sarcoma – leiomyosarcoma. Her CECT abdomen and pelvis revealed a 10.2*9*3.9cm sized, ill-defined heterogeneously enhancing soft tissue density lesion noted in the cervicovaginal region arising from the anterior wall the lesion seen protruding out from the vagina. Loss of fat plane between the lesion and the anterior wall of distal rectum & posterior wall of bladder and urethra s/o neoplastic etiology with no e/o metastasis.

The patient was posted for abdominal panhysterectomy by abdomino-perineo repair after getting an informed and written consent and a proper anaesthetic evaluation. Her hemoglobin was 9.3 g%, coagulation profile was normal. Her LDH levels were 354 U/L which was on the higher side and her chest radiograph showed no evidence of metastasis.

Differential Diagnosis: The clinical presentation often misleads with infected fibroid uterus or acute uterine inversion. The diagnosis of leiomyosarcoma cervix mainly depends on the pathological and immunohistochemical profile and includes squamous cell carcinoma producing similar clinical presentation and other sarcoma arising from cervix such as rhabdomyosarcoma, liposarcoma, primitive neuroectodermal tumour and malignant peripheral nerve sheath tumour(1).

Surgery

After giving epidural + general anaesthesia, the patient's parts were painted and draped. A midline vertical incision of adequate length kept and abdomen was opened in layers. On entering the peritoneal cavity, the ascitic fluid of about 5 cc aspirated and sent for cytology. The uterus was everted and held with two Kocher's clamps and the bowel was packed. Subsequent clamps were applied till uterine and was cut and ligated. The ureters were secured with a tape and were checked after every clamp. At the vaginal end, the growth which was arising from the posterior lip of cervix was excised and the uterus was removed by the abdominal route after applying the uterosacral and the cervicovaginal clamps and was sent for frozen. The vault was closed in a continuous interlocking manner. Hemostasis was secured and abdomen was closed in layers. Intra op one pcv was transfused and patient was shifted to the post op ward. The frozen section report revealed a high grade leiomyosarcoma of the cervix.

Postoperative Period

As the patient presented with burst abdomen, secondary suturing was done. Local examination of the abdomen and pelvis revealed a surgical scar healed by secondary intention and per vaginal examination revealed post hysterectomy status. She was planned for 25 fractions of EBRT for one and half months followed by 6 cycles of Vincristine Adriamycin Cyclophosphamide as first line agents, doxorubicin and ifosfamide as second line agents and gemcitabine and paclitaxel as third line chemotherapeutic agents. The patient was discharged from our side and was given follow up through cobalt unit.

3. Discussion

Sarcomas of the uterine cervix are distinctly uncommon and constitute less than 1% of all cervical malignancies (1). Leiomyosarcomas are malignant tumours composed of cells showing smooth muscle features. Cervical leiomyosarcoma is very uncommon when compared to uterine LMS. They generally occur in the perimenopausal and postmenopausal population in the 4th – 6th decade of life. Most patients present with vaginal bleeding and a bulky pelvic mass at the time of diagnosis. Macroscopically, the tumours are typically large and poorly circumscribed.(3)Owing to the relative infrequency of the disease, most of the available

data on the natural history of cervical sarcomas are derived from case reports and series. To date the largest series of cervical sarcomas identified only 67 cases of cervical LMS among 33074 patients with cervical cancer treated over nearly 17 years for an incidence of 0.2% (2).Diagnosis before surgery is a rare occurrence. In leiomyosarcoma of the uterine corpus, involvement of the cervix is not uncommon, so for definitive diagnosis of primary cervical leiomyosarcoma, the bulk of the tumour should be on the cervix but not the isthmus (1).

The risk factors for cervical leiomyosarcoma include high dose radiation exposure to pelvis, those who are born with an abnormal copy of RB gene, immunocompromised patients infected with EBV and use of tamoxifen. (4)

The histologic diagnosis of cervical leiomyosarcoma is based on the bell's criteria which includes diffuse moderate to severe atypia, a mitotic count of more than 10 mitotic figures/10 high power field and Coagulative tumour cell necrosis (5). Immunostaining showed positivity for smooth muscle actin and muscle specific actin.Strong diffuse block staining for p16 and ki67 showed a higher proliferation rate (6). MRI with LDH is 100% sensitive in diagnosing leiomyosarcoma (7). Leiomyosarcomas typically metastasize via the hematogenous route, most commonly to the lungs. There have been no reported cases of cervical leiomyosarcoma with occult parametrial invasion and lymphatic metastasis is a rare event. Total abdominal hysterectomy with bilateral salpingo oophorectomy is the surgery of choice for early stage cervical leiomyosarcoma (6). There is a debate about oophorectomy in low grade tumour and premenopausal patients as ovarian involvement in sarcomas of the cervix, in particular to leiomyosarcoma of the cervix, is a very rare mode of presentation.

Most patients are considered to be candidates for chemotherapy owing to its tendency to metastasize hematogenously. The prognosis of cervical LMS is based on the tumour grade, tumour stage, age and menopausal status of the patient (8, 9). Giuntoli et al, retrospectively evaluated that stage, older age (>51 years), postmenopausal status and larger tumor size (>5m) to be significantly associated with a reduced likelihood of survival in cervical leiomyosarcoma (10). The five year survival for cervical sarcoma were 80%, 67%, 42% and 20% compared to 95%, 80%, 56% and 32% of cervical squamous cell carcinomas for stage 1A, 1B, II and III, respectively (2).

Doxorubicin and ifosfamide when used as first line chemotherapeutic agents showed the largest response rates(11).Other chemotherapeutic agents used with efficacy in treating advanced leiomyosarcoma include gemcitabine, docetaxel, doxorubicin and ifofamide. Radiation therapy is usually given in the adjuvant setting to prevent locoregional recurrence (11, 12). Combined modality treatment including surgery, radiotherapy and chemotherapy should be used to achieve better survival (13).

The surveillance recommendations as per NCCN guidelines (2015 guidelines) includes history and physical examination every 3 months for 2 years, then every 6-12 months, ct chest/abdomen and pelvis every 3-6 months for 2 to 3 years,

then every 6 months for next 2 years, then annually and education regarding the symptoms of potential recurrence (6).

4. Conclusion

Primary cervical sarcomas are exceedingly rare neoplasms. Given the rarity of these neoplasms management of patients with cervical sarcomas should be individualized based on clinical and pathologic characteristics. In this case of cervical LMS, she presented with acute pelvic organ prolapse and was diagnosed to have an early stage LMS with elevated levels of LDH which is a rare presentation and was treated with combined modality of treatment.

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