International Journal of Research in Medical Sciences
Mehra R et al. Int J Res Med Sci. 2015 Jan;3(1):320-324
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: 10.5455/2320-6012.ijrms20150161

Case Report

A rare case of leiomyosarcoma of cervix and challenges in its management: a case report and brief review of literature

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Received: 4 November 2014
Accepted: 3 December 2014

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ABSTRACT

Leiomyosarcoma of cervix is an aggressive and rare tumour of the cervix with poor prognosis. Management plan differs greatly from other common cervical malignancies. A 38 year old multiparous lady presented with a large circumferential cervical tumour which was diagnosed as leiomyosarcoma of cervix on cervical biopsy. It was a surgical challenge. Patient underwent pre-operative ureteric stenting followed by exploratory laparotomy with hysterectomy with bilateral salphingo-oophorectomy. Surgical difficulties were encountered due to deep impaction of tumour in pelvis and altered anatomy due to the bulk of tumour. Retrograde approach was taken and pouch of Douglas was opened before clamping the mackenrodts and the uterus was bisected and debulking of tumour done simultaneously to facilitate the surgery. Post operatively patient received chemotherapy (Vincristine, adriamycin and cyclophosphamide) followed by radiotherapy. Patient developed vesicovaginal fistula and liver metastasis nine months post- surgery. She received second line of chemotherapy (gemcitabine and docetaxel) after which she was lost to follow up.

Keywords: Leiomyosarcoma, Retrograde approach, Retrograde hysterectomy

INTRODUCTION

Leiomyosarcoma of cervix is a rare cervical tumour with poor prognosis. These are more aggressive than other cervical malignancies. Our knowledge on management of cervical leiomyosarcoma is restricted due to less number of cases and hence inability to conduct clinical trials or chart optimal management plan. Due to rarity of this tumour and deep impaction in pelvic cavity, different surgical challenges have been encountered and reported in literature. We are reporting a rare case of leiomyosarcoma of cervix where difficulties were encountered during it's management. Also the surgical technique of hysterectomy used in this patient was unique and designed in response to difficulties encountered intra-op.

Compilation of data of difficulties encountered in such cases, management followed, and thus charting outcome may help in formulating better management plans and protocols in future.

CASE REPORT

A 38 year old multiparous lady presented to a tertiary care hospital in Chandigarh with four month history of progressively increasing pain in lower abdomen, perineal pressure and increased frequency of micturition. She had no chronic medical illness and no history of any

malignancy in family. Previously, she had a visit to another hospital where a cervical biopsy was taken which was suggestive of leiomyosarcoma. Her general physical examination was within normal limits. On palpation abdomen was soft and non-tender.

Vaginal examination revealed a firm, non-tender, non-fragile circumferential growth arising from anterior lip of cervix, expanding anteriorly and laterally up to lateral pelvic wall (Figure 1). Per rectal examination revealed same firm mass with free rectal mucosa.

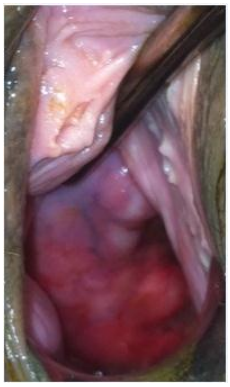

Figure 1: Visualization of cervix on per speculum examination.

Investigations

Patient's haemoglobin level was 9.8 mg/dl revealing mild anaemia. Rest of the blood biochemistry reports were within normal limits. Chest X-ray revealed no abnormality. Ultrasound revealed a cervical mass.

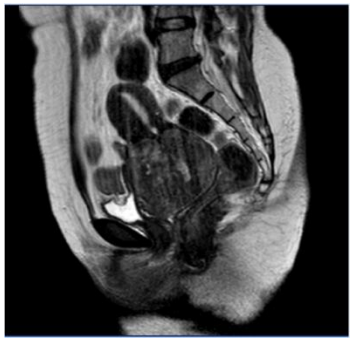

Figure 2: MRI scan showing hyper intense cervical lesion on T2 weighted image with circumferential wall thickening of cervix more on right lateral and anterior part of cervix. Note normal size uterus with no evidence of extension of lesion into uterine body or endometrial cavity.

MRI revealed normal size uterus with altered signal lesion with circumferential wall thickening involving more so right lateral and anterior part of cervix. Endocervical cavity displaced to left side. Lesion heterogeneously hyper intense on T2W and hypo intense on T1W. Lesion projecting into vagina and causing indentation of its wall. Left side subtle thickening with mild stranding seen adjoining fat plane extending up to left levator ani. (Mass closely abutting & indenting posterior wall of bladder with T2 hypo intense line of posterior wall of bladder attenuated at places. Fat planes were well defined with rectum & adjoining bowel loops). No evidence of extension into uterine body or endometrial cavity (Figure 2).

Cervical biopsy report (done outside) revealed smooth muscle neoplasm with mitotic index of 7-10/high power field, with diffuse moderate atypia with no coagulative necrosis. It was reported as atypical smooth muscle tumour with features suggestive of leiomyosarcoma.

Differential diagnosis

A patient presenting with perineal pressure and pain with little or no change in bleeding pattern with a non-fragile cervical growth, these symptoms point towards a cervical fibroid. Invasive cervical cancers have different presentation with irregular vaginal bleeding as the chief complaint. In case of fast growing tumours, cervical sarcomas may be considered. Among cervical sarcomas the commonly found subtype is rhabdomyosarcomas. Other less common tumours are lipsarcoma, leiomyosarcomas, Ewing sarcoma and malignant nerve sheath tumour.[1] In such cases, a biopsy from tumour along with endometrial sampling may be diagnostic. Immunohistochemical stains like Desmin, cytokeratin and SMA are used to differentiate from rhabdomyosarcomas, melanomas and other exophytic cervical cancers.

Treatment

Preoperatively, right ureteric stenting was done. Left ureteric stenting was not possible due to tumour indenting posterior bladder surface.

On exploratory laparotomy, normal size uterus on a ballooned up cervix approximately 10 x 8 cm size was noted. Bilateral round ligaments, infundibulopelvic ligaments and bilateral uterine vessels were clamped, cut and ligated in usual manner. Bladder was pushed down as far as possible.

As cervix was expanded anteriorly and laterally towards left by the mass, it was difficult to reach lower edge of tumour. The uterus was bisected in an attempt to debulk the tumour centrally, to facilitate lateral application of clamps. We did not get much space laterally even after the debulking. However, vision of the area became better. Then the pouch of Douglas was opened and a posterior approach was taken. Transverse cervical ligaments and uterosacral ligaments were clamped successively from below upwards (posterior to anterior) along with an attempt to debulk the tumour centrally.

Total abdominal hysterectomy with bilateral salphingo oophorectomy was done in this unique method. Left ureteric exploration was done and it was found to be intact. Exploration of abdominal cavity and palpation of lymph nodes revealed no other abnormality.

Outcome and follow up

Patient's immediate post-operative period was uneventful.

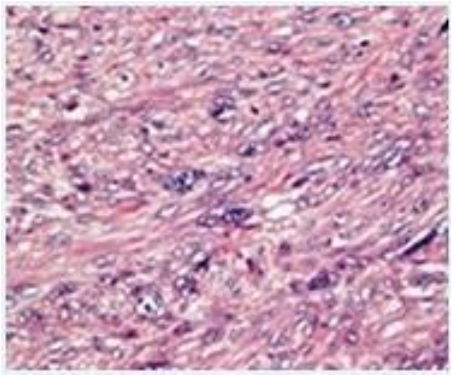

Figure 3: H&E stained slide with 400x magnification showing many bizarre forms and multinucleate tumour cells.

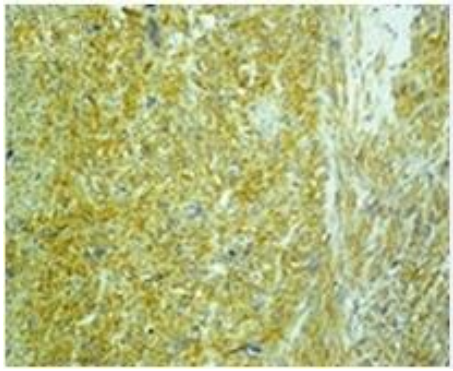

Figure 4: Immunohistochemical study showing positivity of tumour cells for desmin.

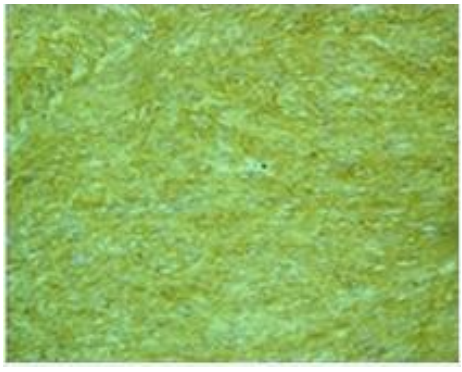

Figure 5: Immunohistochemical study showing positivity of tumour cells for Smooth muscle antigen (SMA).

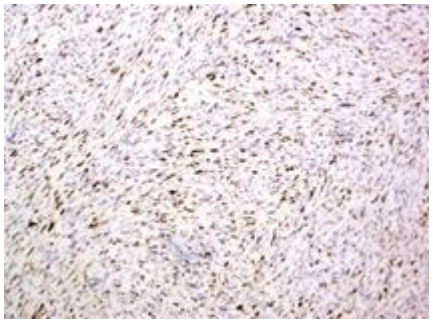

Figure 6: Immunohistochemical study showing positivity of tumour cells for Ki 67.

Histopathological examination of specimen showed a 9 x 5 x 3 cm cervical mass. Microscopic examination revealed a spindle cell tumour with high mitotic activity with presence of bizarre forms and multinucleate tumour cells, with areas of necrosis and hyaline change.

IHC staining showed tumour cells positive for desmin, SMA and Ki 67 suggestive of leiomyosarcoma of cervix (Figure 3, 4, 5, 6).

Four weeks post-surgery, patient was given vincristine 2 mg, cyclophosphamide 870 mg and adriamycin 85 mg 3 weekly for 6 weeks. Later on she received radiotherapy (46 Gy/23 fractions/5 weeks).

Post radiotherapy, three months after surgery, the patient was diagnosed to have a vesicovaginal fistula for which she was catherized and was partially relieved of continuous incontinence suggesting a high vesicovaginal fistula .

Liver metastases were found nine months post-surgery. Metastatic deposits were confirmed by USG guided fine needle aspiration cytology.

Patient received six cycles of gemcitabine (1 gm/m^2) on day 1 and day 8 with docitaxel (100 mg/m^2) on day 8 after which she was lost to follow up.

DISCUSSION

Leiomyosarcomas of cervix are rare tumours occupying a small fraction (approximately 13%) of all reported cervical sarcomas which themselves are rare tumours (less than 1% of all cervical malignancies).[1]

Diagnosis before surgery is a rare occurrence. Differentiation from cervical fibroid is difficult unless supported by histopathology and immunohistochemical marker. Stage, menopausal status, mitotic count, tumour size, age are known to affect the prognosis of uterine leiomyosarcoma,[2,3] however there are too few cases to elaborate the criteria for predicting prognosis in cervical leiomyosarcoma.

Reviewing the cases reported in literature[4-8] it is evident that management plans regarding cervical leiomyosarcoma are mostly extrapolated from studies on uterine leiomyosarcomas.

Apart from routine laboratory investigations, pre-operative work up should include MRI of abdomen & pelvis for detailed assessment of tumour and its metastasis. Lung metastasis should be looked for along with evaluation for local spread. Also, due to proximity of this tumour to ureter, the surgical anatomy may be altered; hence intravenous pyelography or ureteric stenting seems prudent to reduce surgical morbidity. Immunohistochemical stains should be used to rule out the differential diagnosis.

Unlike bulky cervical cancers where radiotherapy is the mainstay of treatment, surgery is preferred in leiomyosarcomas. Sarcomas are radio-resistant and surgery is the first line of treatment, which is often difficult as was in our case. Surgery often includes total hysterectomy with bilateral salphingo-oophorectomy. However, removal of ovaries in premenopausal patients is still a controversial issue, especially in those with low grade tumour. It has been observed in uterine leiomyosarcomas that retention of ovaries in premenopausal women does not worsen the outcome.[3] In postmenopausal patients, hysterectomy with bilateral salphingo oophorectomy seems to be justified.

Most of the problems in surgically removing the tumour are due to its impaction in the narrow pelvic cavity and involvement of the paracervical tissues.

Various surgical techniques have been followed by different surgeons according to the need of the situation.

Grover et al reported use of enucleation of tumour as in myomectomy before doing hysterectomy.[4] Abell et al. have reported several cases with different treatment plans including polypectomy with excision of cervical lip in one patient and partial excision of the tumour followed by radiotherapy in another patient. One case was reported where removal of cervical stump, bladder excision and transplantation of ureters was done.[6] Surgical plans hence may differ in different cases.

Primary surgery has been done for most of the patients except in some cases where the disease was found to be too extensive to excise where primary radiation[6] or chemotherapy[9] has been tried.

Role of lymphadenectomy is controversial in uterine leiomyosarcomas.[3] So, lymph node sampling may be restricted to cases with suspicious lymph nodes in cases of cervical leiomyosarcomas also.

Evidence gathered from studies from uterine leiomyosarcoma shows that adjuvant chemotherapy with doxorubicin decreases recurrence rates but does not cause any significant change in survival rates.[10]

Recently, a phase II trial for adjuvant chemotherapy in high grade uterine leiomyosarcomas with gemcitabine and docetaxel followed by doxorubicin has showed that 78% patients were progression free at 2 years and 57% remained progression free at 3 years.[11]

Post-operative chemotherapy is believed to control systemic spread and radiotherapy prevents local recurrence. However, further input is needed in knowing the exact role of radiotherapy and risks versus benefit analysis in such tumours.

Due to variation in treatment strategies undertaken in many cases reported in literature, and lack of standard protocol for diagnosing and management of these patients, the true response to a standard treatment cannot be deduced from available data.

The prognosis of these tumours is poor, even for early stage disease. In the series reported by Abell et al., 6 out of 8 patients died of disease progression, pelvic recurrences or distant metastasis within two years.

Clearly, it is an aggressive tumour where all the three modalities of surgery, chemotherapy and radiotherapy are tried to achieve better patient survival. Because of the rarity of this tumour and problems encountered with management, a multidisciplinary team should chart treatment plan for patient and should include a pathologist, an oncosurgeon, and radiation & medical oncologists.

As these are rare tumours of cervix, every such patient should have a thorough follow up and the outcome should be charted and reported for contribution to further studies.

ACKNOWLEDGEMENTS

We would like to thank Dr. Anju Huria, Professor and Head of Department of Obstetrics and Gynaecology (Chandigarh, India) for her valuable support.

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

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DOI: 10.5455/2320-6012.ijrms20150161
Cite this article as: Mehra R, Sidhu SPK, Pandey AK, Garg S. A rare case of leiomyosarcoma of cervix and challenges in its management: a case report and brief review of literature. Int J Res Med Sci 2015;3:320-4.