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LEIOMYOSARCOMA OF THE CERVIX - A RARE CASE REPORT

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ABSTRACT
Uterine cervix sarcomas are rare tumours forming less than 1% of all cervical malignancies. Leiomyosarcoma (LMS) of cervix is extremely uncommon. According to literature, only 22 cases have been reported of which only two cases have been reported in the Indian literature. The rarity of LMS is contributed to paucity of available data regarding the signs and symptoms, mode of management and prognosis of the patients. We report a rare case of leiomyosarcoma of uterine cervix in a 34 year premenopausal Indian female presenting with bulky cervical mass and bleeding per vaginam. Final diagnosis was made on the basis of histopathological and immunohistochemical findings.

INTRODUCTION

It is said that sarcomas comprise less than 1% of all cervical malignancies, of which leiomyosarcoma (LMS) is an extremely rare tumour with an aggressive course. Though leiomyosarcoma is one of the most common non-epithelial malignant neoplasms arising in soft tissue, incidences of the uterine cervix is extremely rare [1]. Most patients present with vaginal bleeding and a bulky cervical mass at the time of diagnosis. They commonly occur in the perimenopausal and postmenopausal population in their 4th–6th decades of life [1].

We present a case report of 34 year Indian origin female presenting with cervical LMS.

CASE REPORT

A 34 year old married lady (gravid 2, para 2) came with bleeding per vaginum and pain in abdomen since 2 months. Bleeding was on and off initially which later become progressive, persistent and was associated with dull aching, intermittent lower abdominal pain. Patient also experienced foul smelling vaginal discharge. Per vaginal examination revealed bulky cervix.

On speculum examination, a haemorrhagic mass was seen protruding from cervix. Abdominopelvic ultrasonography showed hypoechoic lesion arising from cervix suggestive of cervical fibroid.

MRI showed a large soft tissue mass of size 7.5 x 6.5 x 6.5cm arising from cervix. Simple hysterectomy was performed under the clinical impression of fibroid but later histopathological examination revealed leiomyosarcoma of uterine cervix hence total abdominal hysterectomy with bilateral salphino-oopherectomy was not performed.

Gross findings showed uterus along with cervix measuring 13.5 x 7 x 5.5cm. In Uterus, endometrium measured 4mm and was hemorrhagic and myometrium measured 2.4 cm and showed areas of haemorrhage. Cervical length was 9.5 cm. Cervix showed a large

proliferative growth protruding out of it, measuring 7.5 x 6.5 x 6.5cm whose cut surface was solid, fleshy, grey white with few areas of haemorrhage and necrosis (Figure 1)

Microscopic examination revealed a tumor mass arising from cervix which was composed of oval to spindle shaped cells arranged in fascicles and sheets, having pleomorphic, hyperchromatic blunt nuclei (cigar shaped), showing nuclear atypia with moderate amount of eosinophilic cytoplasm. Many bizarre shaped multinucleated cells were also noted. Malignant cells showing periangiomatous pattern were also seen. Mitotic activity of 7-9/10 hpf with atypical mitotic figures and areas of necrosis were also seen. No lymphovascular emboli was noted in cervical stroma (Figure 2, 3,4). Uterine endometrium showed endometrial glands and stroma in proliferative phase. Myometrium showed congested and dilated blood vessels .Uterus (endometrium and myometrium) was free of tumour. On immunohistochemical stains (IHC), neoplastic cells showed positivity for Smooth Muscle Actin (SMA) and caldesmon. On the basis of histopathological findings and IHC, final diagnosis made was Leiomyosarcoma of uterine cervix.

Fig 1. Gross image showing cut section of uterus and cervix with a large, solid, fleshy, grey white proliferative growth in the cervix.

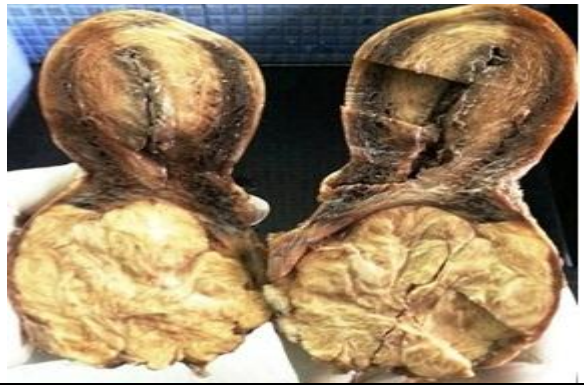


Fig 2. 10X, H&E, showing spindle shaped malignant tumour cells having pleomorphic, hyperchromatic blunt nuclei (cigar shaped).

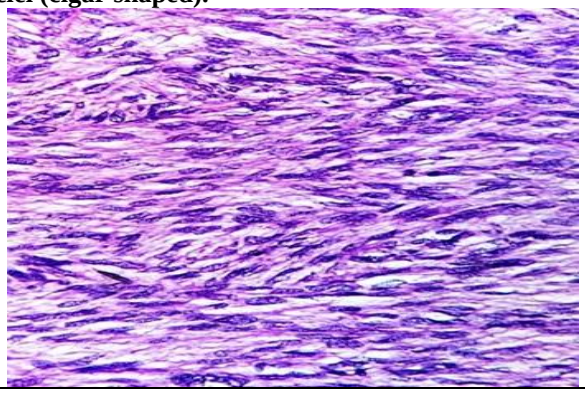


Fig 3. 40X, H&E ,showing malignant cells having nuclear atypia and atypical mitotic figures

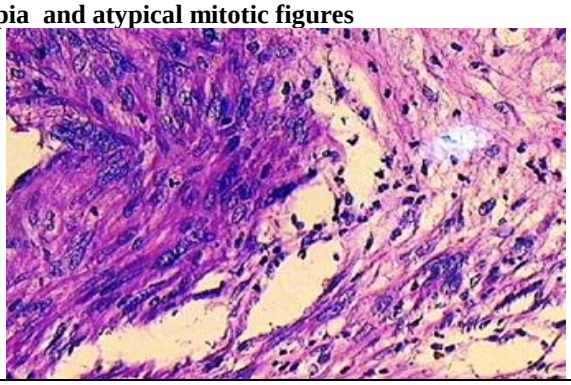
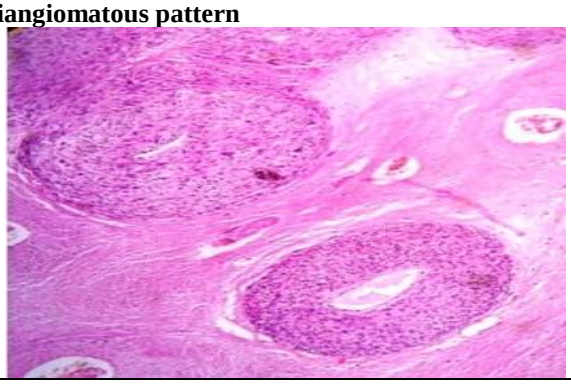


Fig 4. 10X, H&E, showing malignant cells having periangiomatous pattern



DISCUSSION

The most common histological subtype of cervical cancer is squamous cell carcinoma followed by adenocarcinoma, adenosquamous and neuroendocrine/ small cell carcinoma [2].

The mesenchymal tumors of the uterine cervix are heterogenous diseases and are distinctly uncommon. Cervical sarcomas on histopathology may exhibit smooth muscle or skeletal muscle, fibroblastic, nerve sheath, fatty, fibrohistiocytic, neuroectodermal component and tumors with uncertain differentiation. The seven most common types of cervical sarcomas, listed in descending order are embryonal rhabdomyosarcoma (ERMS); leiomyosarcoma (LMS); undifferentiated endocervical sarcoma (UES); alveolar soft part sarcoma (ASPS); Ewing's sarcoma/primitive neuroectodermal tumor (PNET); and liposarcoma [3].

Cervical leiomyosarcoma is very uncommon tumour, when compared to uterine LMS. Cervical LMS is usually seen in the perimenopausal age group. The average age of diagnosis is said to be 46 years. Abnormal vaginal bleeding is known as the most common presenting symptom [4]. Imaging techniques cannot give a reliable preoperative diagnosis and computed tomography is also not able to differentiate between different types of cervical pathology. The clinical presentation often misleads with fibroid of uterus.

The diagnosis is usually made on histology and immunohistochemisrty on basis of the criteria proposed by Norris and Taylor, which is similar to those for uterine LMS. Other criteria were proposed by Bell, Kempson and Hendrickson [4].

Grossly, the tumours are usually large (up to 12 cm) and not well circumscribed [5]. The tumour mass either protrudes from the cervical canal or thickens and expands it circumferentially.

On microscopy, leiomyosarcomas are malignant tumours comprising of cells having smooth muscle features [1]. The current method is to simplify diagnostic criteria from the corpus tumours and apply them to their cervical counterparts, in view of various combinations of cytological atypia, coagulative necrosis and mitotic activity to predict their malignant potential. On immunohistochemical stains (IHC), neoplastic cells showed positivity for desmin, smooth muscle actin (SMA), calponin and caldesmon.

Several prognostic factors are reported in patients with LMS, which has impact on patient survival rates. The premenopausal age group, low mitotic figures (< 10/ hpf) and lower grades (grade 1 and 2) are observed as favourable prognostic factors associated with a higher survival rate [5]. Giuntoli *et al.*, retrospectively evaluated that higher stage, older age (>51 years), postmenopausal status and larger tumour size (>5 cm) to be significantly associated with a reduced likelihood of survival in cervical leiomyosarcoma [6]. Being of premenopausal age, lower grade was the favourable prognostic factor in our patient.

Because of the rarity of this tumour and the paucity of literature on the subject, clinicians and pathologists should observe guidance to the current accepted standards for staging, grading and the management of uterine cervix LMS [4]. The best management of these tumours is uncertain owing to its rarity;however combined modality treatment can result in prolonged survival and cure. These includes total abdominal hysterectomy with bilateral salpinooophorectomy. However, if pelvic or retroperitoneal lymphnode are found to be involved, they should be removed. Adjuvant radiotherapy is required to decrease local recurrences. Adjuvant chemotherapy is needed for managing metastatic LMS [4]. The combination of doxorubicin and ifosfamide is given more commonly as first line therapy for females diagnosed with recurrent or advanced LMS [5].

In a review of literature, Salazar and Dunne observed a non-significant trend towards improved survival in stage-I uterine leiomyosarcoma who received postoperative pelvic radiotherapy [1].

CONCLUSION

Leiomyosarcoma of uterine cervix is a rare disease and diagnostic confirmation is based on pathological and immunohistochemical profile. Because of the small number of cases reported so far, the ideal approach for these malignant tumours is the team work between the surgical oncologist, the pathologist, the radiation and the medical oncologists for optimising the results for the best interest of the patient.

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CONFLICT OF INTEREST:

The authors declare that they have no conflict of interest.

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