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Spindle Cell Sarcoma of the Uterine Cervix: A Case Report and Review of Literature

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Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Introduction: Squamous cell carcinoma is the most common histology of the uterine cervix. Sarcomatoid variety of the cervix cancer is a rare morphologic entity. The prognosis for women with cervical sarcoma tends to be worse than that of squamous histology.
Case Presentation: We are reporting a case of a 57 years old female with primary malignant cervical tumour showing spindle cell tumour in histopathology and immunohistochemistry positive for cytokeratin and vimentin.
Interventions: Patient underwent abdominal hysterectomy and bilateral Salphingoophorectomy and post op event was uneventful. The final diagnosis of Spindle cell sarcoma was confirmed by pathological and immunohistochemical examination.
Conclusion: Spindle cell Sarcoma of the cervix is a rare pathology. High grade Spindle cell tumours continue to have a poor prognosis due to its highly malignant, aggressive, and refractory nature to local treatment which necessitates the early intervention to improve the survival.

Keywords: Spindle cell sarcoma; squamous cell carcinoma cervix; sarcoma cervix.

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1. INTRODUCTION

Cervical cancer is one of the most common forms of cancer in women worldwide, too are on a fast and steady rise, accounting for more deaths in women than any other cancer in the developing world [1]. Most common histologic subtype of cervical cancer is squamous cell Carcinoma followed by adenocarcinoma, whereas cervical cancer is the second most common malignancy in women worldwide. Other rare pathologies are melanomas, sarcomas, lymphomas, and metastatic tumours [2].

Sarcoma of the Cervix is a very rare malignancy. Cervical sarcomas constitute less than 1% of all cervical malignancies [3]. Of the sarcomas, rhabdomyosarcomas, the most common of the embryonal subtype, are the most frequently reported [4]. It differs from squamous cell carcinomas of the cervix in terms of having a poorer prognosis, an aggressive nature with short disease-free survival. The tumour usually is at an advanced Stage at presentation and is characterized by early recurrence after treatment. The tumour has unusual metastatic sites, such as the peritoneum, kidney, and subcutaneous tissues. Because of the rarity of the disease, no standard diagnosis and treatment approach are available. It is usually managed like squamous carcinoma of the cervix [5].

2. CASE REPORT

A 57yr old female patient presented to our hospital with complains of postmenopausal bleeding per vagina since 2 months and was also associated with foul smelling white discharge per vagina. She is a known case of hypertension and hypothyroidism of regular treatment. No other significant comorbidities. On gynecological examination, cervix was replaced by ulcero-proliferative growth about 5x5cms, cervical os was completely destroyed, vaginal walls were free, rectal mucosa was normal, Bilateral Parametrium was free. Patient was clinically staged as FIGO stage IB2.

A biopsy excision from the tumour mass was done.Histopathology of the growth showed low grade spindle cell tumor. Her routine blood count, liver function test, renal function test were within normal limits. Chest X-ray showed no significant abnormality.

MRI pelvis was done which showed, 5x5x4 cms well defined smooth marginated mass lesion showing hyper intense signals on long TE sequences and isointense signals on T1W1 is seen in proximal vaginal lumen abutting external os, showing no distinct cleavage with posterior ectocervical wall. It is seen bulging into posterior fornix. No infiltration in adjacent structure and bilateral parametrium appear normal. No significant lymphadenopathy (Fig. 1).

After discussing the case in the institutional multidisciplinary committee, patient was planned for the surgery. Patient underwent abdominal hysterectomy and bilateral salphingo-oopherectomy and post-operative period was uneventful. Histopathological specimen shows tumor is composed of cellular areas and hyalinised areas. The tumor cells are spindle cells with thin linear wavy nuclei, and pump elliptical cells with blunt ended vesicular nuclei, cytoplasm is minimal. There is parallel streaming pattern of cells along with collagen fibers in interlacing and parallel bundles. The less cellular

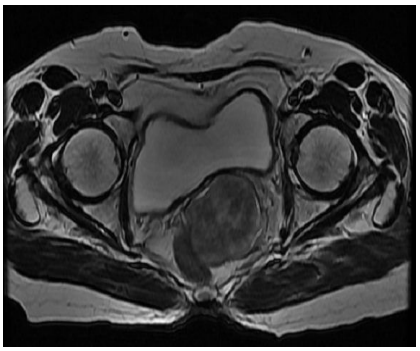

Fig. 1. MRI image showing the lesion

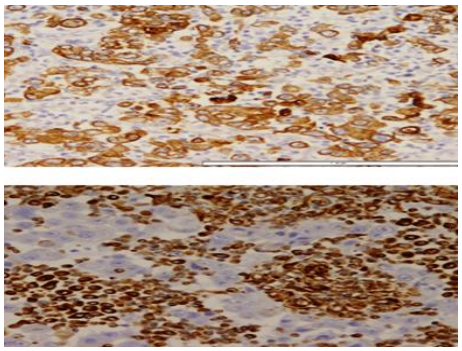

Fig. 2. Pan Cytokeratin, Vimentin positive on IHC

areas have more prominent collagen fibers. Hyalinised areas are devoid of nuclei. Mitotic figures are 2-10/hpf. Impression was made as low grade spindle cell stromal tumor. Immunohistochemistry showed positivity to Pan-cytokeratin and Vimentin and negative to S100 and CD34 (Fig. 2). Postoperative period was uneventful and patient did not develop any symptoms. Since the Postoperative histopathology showed Low grade spindle cell tumor, patient did not undergo any adjuvant treatment and was kept on regular follow-up. After one-year follow-up, patient is free from the disease without any symptoms and on imaging studies there was no relapse or recurrence.

3. DISCUSSION

Squamous cell carcinoma is the most common histology of cervical cancer pathology; other types are adenocarcinoma, carcinosarcoma, lymphoma, and sarcoma [2]. Sarcomatoid carcinoma is a rare entity of the female genital tract and even more rare in the cervix. It has been described more frequently in the aero-digestive tracts and skin. As in prior studies, carcinosarcomas were the most common histologic variant and accounted for 40% of the cervical sarcomas [6,7,8].

Sarcomas of the uterine cervix are uncommon and constitute less than 1% of all cervical cancers. 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 small case series. To date, the largest series of cervical sarcomas identified 323 cases among 33,074 patients with cervical cancer treated over nearly 17 years for an incidence of 1% [9]. Among 1583 patients with cervical cancer treated over nearly 17 years, wright et al reported only 8 cases of cervical sarcoma for an incidence of 0.5% [10].

Primary cervical sarcomas are exceedingly rare neoplasms. Most patients present with vaginal bleeding and a bulky pelvic mass at the time of diagnosis. The diagnosis of sarcomatoid carcinoma is based on histopathology and immunohistochemistry findings. The immunohistochemistry of sarcomatoid carcinoma is indicated to distinguish the epithelial differentiation from sarcoma. The causes associated with the development and the behaviour of sarcomatoid carcinomas is not well understood. The theories which have been described for the development of sarcomatoid carcinomas are a stem cell monoclonal origin which is capable of divergent differentiation or a multiclonal derivation from two or more independent type of cells. Several studies of sarcomatoid carcinomas in different organs showed an evidence for the sarcomatoid transformation from the epithelial cells [11-14].

A case reported by Lin et al of sarcomatoid squamous cell carcinoma of the cervix which is HPV-16 positive, and a lack of p53 overexpression in the components of both spindle cells and squamous cells, suggested that tumour cells with spindle cell morphology are derived from the squamous cells. Also tumour had an overexpression of the Retinoblastoma protein and decrease in apoptosis compared with the squamous cell cancer cases of the same institution. This data may suggest the aggressiveness of the clinical behaviour of the disease. As this type of cervical cancer is rare, it is difficult to draw conclusions on behaviour of the disease, methods for diagnosis, treatment protocols and outcome of the disease [15].

A series of 9 cases of sarcomatoid carcinomas of the cervix was reported by Brown et al found that patients have a median disease-free interval of 4.9 months and one patient had a survival of 40 months. In this study, although all patients had a complete response to initially, more than half of the patients had recurrences of the disease in less than 5 months after initial treatments. None of patients responded well to a second-line treatment [16].

The standard of treatment is Surgery and radiotherapy, but chemotherapy (Doxorubicin based regimen) has shown good response either as adjuvant therapy or as a palliative therapy in metastatic disease. Hysterectomy with bilateral salphingo-oophorectomy is the main modality. Surgery is used to diagnose stage and treat uterine sarcoma. After surgery, adjuvant radiotherapy is indicated if high risk factors such as the extra capsular extension, positive lymph nodes and positive margins are present. In metastatic disease palliative chemotherapy or radiotherapy may be given. Nodal disease was rare in women with cervical adenosarcomas and occurred in only 3% [17]. Given the low incidence of lymph node metastases for uterine and soft tissue sarcomas the value of lymphadenectomy for cervical sarcomas has been questioned [18,19].

4. CONCLUSION

Sarcomas of the uterine cervix are uncommon and constitute less than 1% of all cervical cancers and they have a more aggressive behaviour with a very short interval for disease progression after initial therapy and failure to respond to second line treatment, In comparison to squamous cell carcinomas of the cervix, cervical sarcoma should be considered as a high-grade tumours and an aggressive approach at the presentation in the form of multimodality therapy should be considered.

CONSENT AND ETHICAL APPROVAL

This study was approved by the Institutional Ethical Committee and Consent from the patient has been taken for Publication.

CONFLICT OF INTEREST

The author declared no potential conflict of interest with respect to the research authorship, and or publication of the article.

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