

Small Cell Carcinoma of Cervix: Case Report of Rare Cervical Cancer Subtype

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ABSTRACT

Small cell carcinoma of the cervix is a rare and aggressive type of cancer that presents with early metastasis and poor prognosis. There are no specific features on imaging to distinguish it from the more common types, like squamous cell carcinoma. As the management differs from common subtypes, proper identification is essential for better prognosis. We present a case of a 42-year-old female with small cell carcinoma of the cervix with parametrial spread, vaginal involvement, and metastatic iliac lymphadenopathy. She was treated with sequential chemotherapy and radiation as per standard guidelines.

Keywords: Cervix Uteri; Lymphadenopathy; Small Cell Carcinoma

INTRODUCTION

Small Cell carcinoma of cervix (SCCC), also known as small cell neuroendocrine carcinoma of the cervix, is a rare, highly aggressive, and distinct subtype of cervical cancer. It represents a significant clinical challenge due to its rapid growth, early metastasis, and poor overall prognosis compared to the much more common squamous cell carcinoma and adenocarcinoma of the cervix. These may present with bulky or small lesions with greater local involvement and metastatic lymphadenopathy. MRI is the ideal non-invasive imaging modality for evaluating tumor size and local extent, though there are no specific imaging features to distinguish it from the more common cervical cancers. The gold

standard for diagnosis is histopathological and immunohistochemistry examination.¹

CASE REPORT

A 42-year-old P2 L2 lady presented in the emergency with a 2-month history of abnormal vaginal bleeding and lower abdominal pain without any other gastrointestinal and urinary symptoms or significant medical comorbidities. She had no family history of cancer. On per speculum examination, there was a 3 x 4 cm cauliflower-like growth present in the cervix. The uterus was bulky, around 8 weeks in size, and the anterior two-thirds of the vagina was involved without parametrial involvement, on per vaginal

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examination. On per rectal examination, the rectal mucosa was free. Cervical biopsy was done, and immunohistochemistry showed synaptophysin negative, chromogranin and CD56 positive, suggesting neuroendocrine carcinoma, favoring the diagnosis of small cell carcinoma.

Multiplanar MRI of pelvis was done in 1.5 T scanner and showed a large heterogeneously enhancing T2 iso to hyperintense cervical mass, measuring about 7.4 x 7 x 5.4 cm, showing restricted diffusion, with disruption of the stromal outline in the left posterolateral aspect

and parametrial extension towards the left side, abutting the internal iliac vessels and invading the upper two-thirds of the vagina, consistent with cervical malignancy (Figure 1). Additionally, there were two conglomerated left internal iliac lymph nodes, suggestive of metastasis. Chest X Ray, CT scan of chest, and whole-body bone scan were unremarkable. Final diagnosis of stage T2bN1MO small cell cervical cancer was made. The patient was treated with six cycles of chemotherapy (etoposide and cisplatin) followed by radiotherapy.

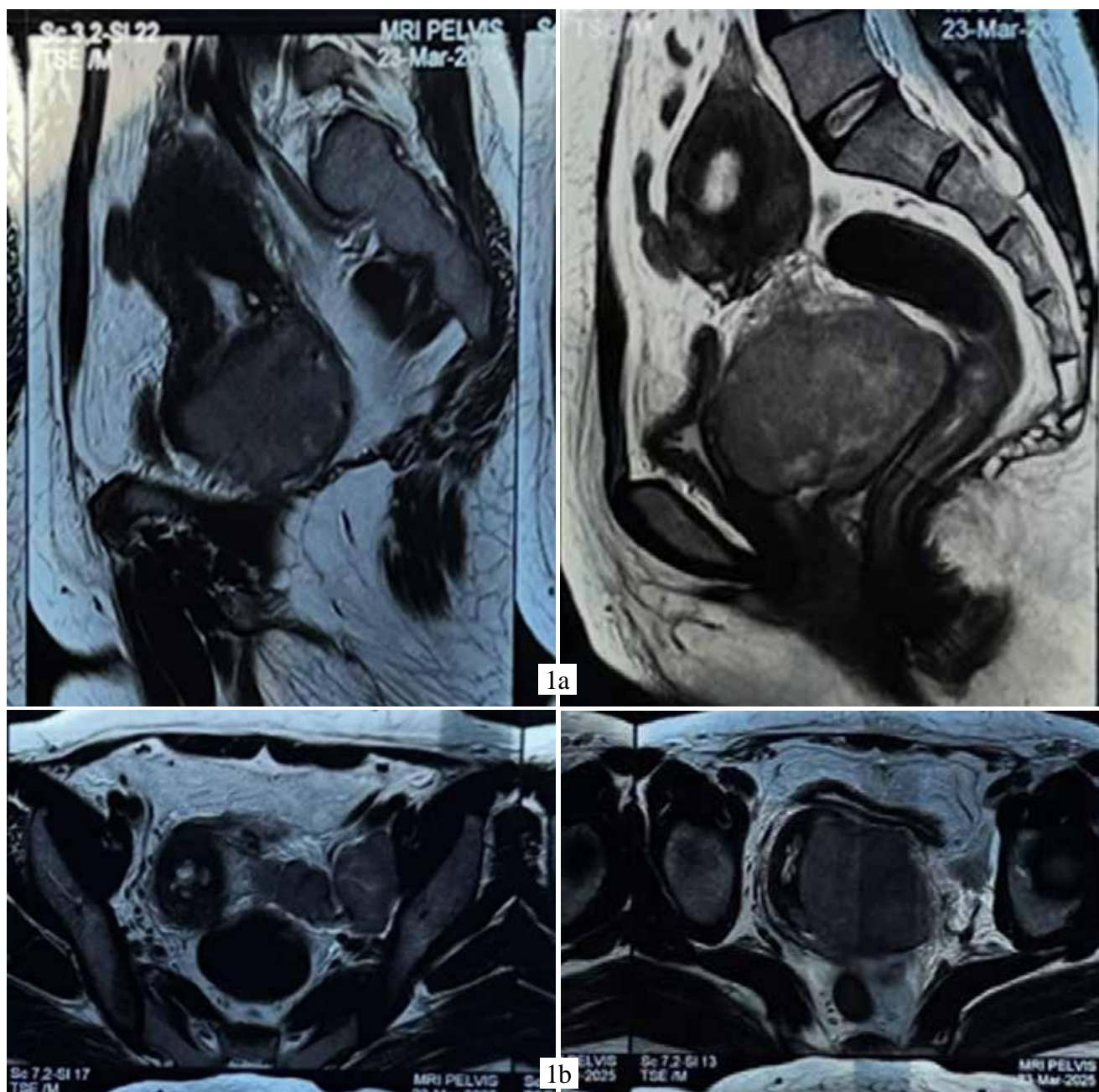




Figure 1: 1a. T2 sagittal, 1b. T2 axial images of the pelvis of a 42-year-old female, showing a large iso-hyperintense mass in the cervix with parametrial extension on the left side, with a conglomerated nodal mass in the left Iliac region. 1c. Post-contrast T1 coronal and axial images show heterogeneous enhancement.

DISCUSSION

Cervical cancer is the most common gynecological cancer in women in Nepal, according to GLOBOCAN 2022. Small cell carcinoma of the cervix (SCCC) is a heterogeneous group of tumors, representing 0.5-1% of all invasive cervical cancers. This cancer is linked to human papillomavirus (HPV), mainly types 16 and 18. It develops in the hormone-producing neuroendocrine cells and arises from the argyrophil cells or AUPD amine precursor uptake and decarboxylation (AUPD) cells in the cervix. MRI is the ideal initial imaging modality for evaluating the tumor size and local extent. However, this subtype is a pathological diagnosis with no specific imaging features to differentiate from the more common types of cervical carcinoma. Histopathologically, the tumor comprises round to spindle-shaped cells with an infiltrative growth pattern with hyperchromatic nuclei, lymphovascular space invasion, and necrotic areas. Immunohistochemical markers of neuroendocrine differentiation-including chromogranin, CD56, and synaptophysin, are usually positive (4,5).^{1,2,3,4,5}

Patients are usually younger and typically present with abnormal vaginal bleeding, pelvic pain, or discharge. Due to its aggressive nature, it often presents at an advanced stage with lymph node involvement and distant metastases at the time

of diagnosis, with a greater mortality (1.84-fold higher risk of death) compared with cervical SCC. Due to its low incidence, a standard treatment protocol for SCCC of the uterine cervix has not yet been established. Based on data from a limited retrospective analysis, the Society of Gynecologic Oncology recommends a multimodality therapeutic strategy and etoposide/platinum-based chemotherapies for SCCC.^{6,7}

The Gynecologic Cancer Inter Group published a consensus review on the treatment of SCCC. The treatment for patients with early-stage disease, up to FIGO stage IIA1, except IB3, consists of radical hysterectomy and lymphadenectomy followed by adjuvant chemotherapy. In advanced-stage disease, chemoradiation or systemic chemotherapy consisting of etoposide and cisplatin is recommended. In locally advanced disease (FIGO stage IB3 and IIA2 onwards) or nonoperative patients, combined chemotherapy and radiation therapy is recommended. Systemic chemotherapy with three-drug combinations (vincristine/doxorubicin/cyclophosphamide), alternating regimens (cisplatin/etoposide), and dose escalation regimens is the initial treatment of choice in patients with distant metastasis (FIGO stage IVB) (10).^{7,8,9,10}

CONCLUSION

SCCCs are extremely rare, aggressive types of cervical cancers with markedly worse prognosis as these present with locally extensive disease, lymphadenopathy, and distant metastasis in early stages. MRI is ideal for assessing the local extent of the disease, though there are no specific morphological features to differentiate it from other common types. Pathological diagnosis is the gold standard for early identification and optimal treatment.

CONFLICT OF INTEREST

None

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