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Small Cell Neuroendocrine Carcinoma of the Cervix: A Striking Entity

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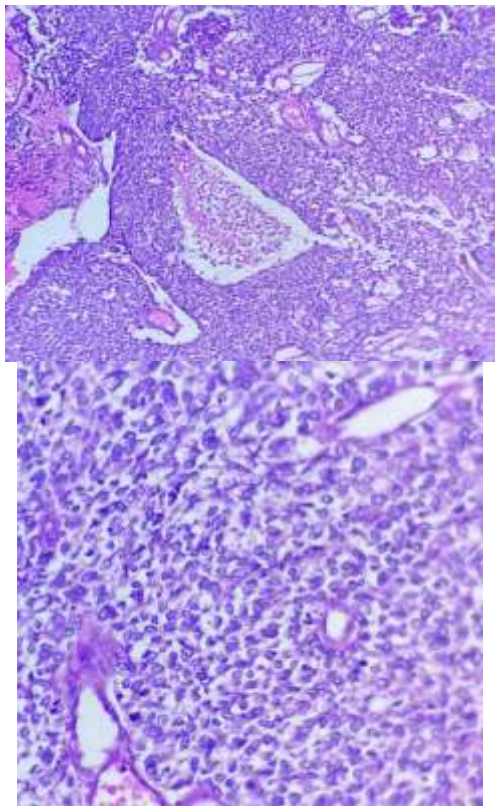
ABSTRACT- Small cell neuroendocrine carcinoma (SCENC) are high grade carcinoma composed of small to medium sized cells with scant cytoplasm and neuroendocrine differentiation. SCENC can occur anywhere in gynaecological tract, most commonly in the cervix. SCENC in cervix is a rare and a very aggressive tumour. Once being considered to be a rare type of squamous cell carcinoma, evidence has proven that most of the tumours express one or more markers of neuroendocrine differentiation. The behaviour of this rare malignancy is different from that of squamous cell carcinomas, with a high propensity for nodal and distant metastases. Hence, there is a need to highlight this histopathological entity.

I. INTRODUCTION-

Small cell carcinoma of the cervix (SCCC) is a rare neuroendocrine malignancy comprising less than 3–5% of cervical carcinomas . SCCC is a high-grade neuroendocrine tumor that shares numerous characteristics of small cell lung carcinoma (SCLC) and has often been considered an "extrapulmonary" small cell carcinoma.Along with shared morphological characteristics and architectural patterns, SCCC and SCLC sharemany immunohistochemistry markers, such as synaptophysin, chromograninA, and CD56. The diagnosis of primary SCCC is made by cervical biopsy noting histopathologic features characteristic of small cell carcinoma, along with exclusion of primary SCLC [1].Here we are reporting a case of Small cell carcinoma of the cervix (SCCC) to make pathologists and gynecologists aware about the occurrence of such rare and aggressive tumour entity.

II. CASE REPORT-

A 40 year old female presented with chief complaints of lower abdominal pain and increased vaginal bleeding since 2 years. On per vaginum examination cervix was erythematous with friable exophytic mass.On grosss examination severe hemorrhagic soft tissue piece of 2.5x2x0.8cm was found. Microscopy reveal diffuse growth pattern forming sheets with small acini resembling rosette. Perivascular growth pattern also seen. The tumor cells were round to oval,monomorphic with indistinct nucleoli. Mitotic activity and Foci of necrosis were also seen. On immunohistochemistry the neuroendocrine component exhibited intense membrane immunoreactivity for synaptophysin as well as granular, dot like perinuclear cytoplasmic positivity for chromogranin thus the tumor identified as Small cell carcinoma of the cervix (SCCC) on histopathology.



Small cell neuroendocrine carcinoma cervix H&E showing small "blue"cells with hyperchromatic nuclei and scant cytoplasm (10X,40X)

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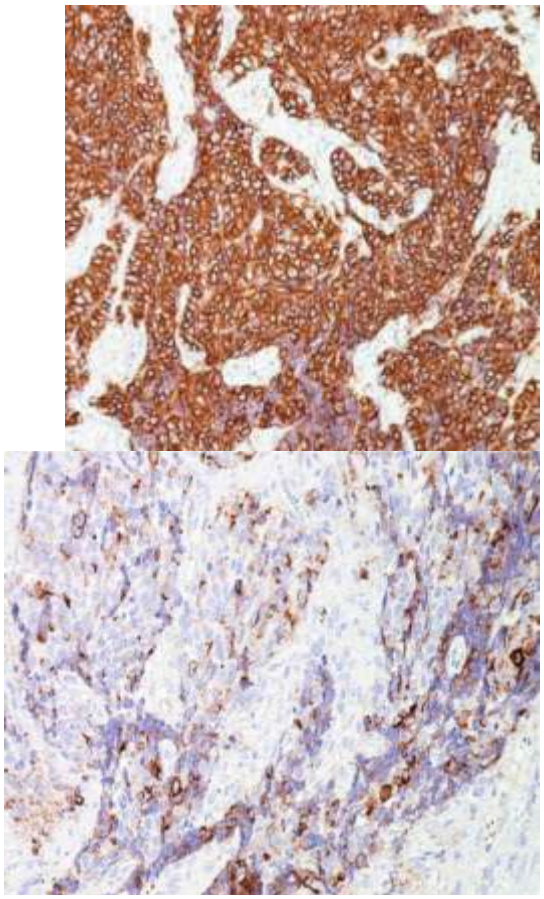

Intense membrane immunoreactivity forsynaptophysin
Granular perinuclear cytoplasmic positivity forchromogranin

III. DISCUSSION-

SCCC has historically been regarded an extrapulmonary variant of SCLC, recent advances have indicated that small cell carcinomas in different anatomic sites carry distinct genetic markers and are unique diseases[1].

SCCC of the female genital tract is a subtype of neuroendocrine tumors (NETs); the incidence of NET, particularly SCCC, is very rare, accounting for 1–2% (range, 0.5–5%) of all cervical malignancies. The presenting symptom is most commonly vaginal bleeding which was also present in this case. The precursor lesion is often nonspecific, and the clinical manifestations and predilection site of SCCC are similar to those in cervical cancer patients SCCC is difficult to distinguish from other small round-cell tumors, such as lymphoma, primary PNET, and poorly differentiated carcinoma, based on the morphological criteria [2].

The exact origin of the NENs of the cervix is unknsown. However, argyrophilic cells in ectocervix and endocervix epithelium are considered a potential precursor for NENs. Among NENs of the cervix, SCNEC is the most common (80%)variant, and it constitutes <1% of all female genital tract malignancies[3].

Diagnosis of SCCC requires tissue biopsy for routine H&E staining, immunohistochemistry for neuroendocrine markers, and systemic imaging of the chest, abdomen, and pelvis to rule out the possibility that this represents a metastasis from another site. SCCC exhibits various degrees of neuroendocrine differentiation, as detected using conventional morphological, ultrastructural, histochemical, and/or immunohistochemical criteria[2].

Diagnostic accuracy in detecting SCNECs by Pap smear is very low. Histomorphology includes various differential diagnoses such as poorly differentiated squamous carcinomawith small cells, poorly differentiated adenocarcinoma, low-grade endometrial stromal sarcoma, lymphoma,rhabdomyosarcoma, melanoma, myeloid sarcoma, and primitive neuroectodermal tumor (PNET).While in IHC synaptophysin and CD56 are the most sensitive, while chromogranin is themost specific NE immunohistochemical marker.NECs spread by lymphatic and hematogenous routes even if the tumor is clinically confined to thecervix. [3].

Multiple studies have demonstrated the presence of Human Papilloma Virus type 18 DNA or messenger RNA in almost two thirds of cases and perhaps more often in tumours in which a neuroendocrine differentiation has been shown. Patients with small cell neuroendocrine cervical cancer have a poor prognosis and a predilection for nodal and distal metastases is very high[4].

Advanced FIGO stage, larger tumor size, lymph nodemetastasis (LNM) (+), lymphovascular space involvement (LVSI) (+), parametrial involvement(PI) (+), depth of stromal invasion (DSI) > 2/3and radiation therapy (RT)were associated were associated with poor survival.However, Radicalsurgery combined with a chemotherapy regimen similar to that of small cell lung cancer maybe a potential therapeutic approach for SCNEC[5].

IV. CONCLUSION-

Small cell carcinoma of the cervix is a rare and a very aggressive tumour. Once being considered to be a rare type of squamous cell carcinoma, evidence has proven that most of the tumours express one or more markers of neuroendocrine differentiation. The behaviour of this rare malignancy is different from that of

squamous cell carcinomas, with a high propensity for nodal and distant metastases. This case has been highlighted for us to be familiarized with the histopathological differentials of small cell tumours of the cervix and to stress on the poor prognosis and aggressive nature of the tumour.

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