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Case Study

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NEUROENDOCRINE TUMOR OF VAGINAL VAULT: A RARE PRESENTATION

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ABSTRACT

In about 2% of malignancies, small cell carcinoma is diagnosed in female genital tract.[1] Cervical small cell carcinoma and ovarian carcinoids are the most prevalent gynaecological NE tumors.[2] Small cell nueroendocrine carcinoma (SCNET) of female genital tract is aggressive in nature and is similar to cancer arising from the bronchus We report a very rare case of 63-year-old hysterectomized woman, who presented to gynecology OPD at Dr. RPGMC Tanda, with complain of postmenopausal bleeding, hematuria, and dyschezia. She had undergone total abdominal hysterectomy with bilateral salpingophorectomy for AUB 11 yr back at our institute. Diagnosis of small cell neuroendocrine ca of vault was made with tissue biopsy and IHC. Imaging studies were done to asses the extent of disease. The patient underwent six cycles of chemotherapy and 16 cycles of external radiotherapy, leading to complete regression of the lesion. To the best of our knowledge, very few NE tumor of the vault has been reported.

INTRODUCTION

Small-cell neuroendocrine tumors (SCNETs) of the female genital tract are rare and aggressive tumors that are characterized by a high rate of recurrence and poor prognosis.[3] They can arise from various sites within the female genital tract, including the cervix, endometrium, ovary, fallopian tube, vagina, and vulva. They are composed of cells with neuroendocrine features, such as the ability to produce and secrete hormones and peptides, and a high mitotic rate.[4] The diagnosis of NE tumors may be challenging. The differentiation of small cell NE carcinoma from poorly diffrentiated squamous cell carcinoma with NE features may be especially difficult.

Immunohistochemical staining plays a crucial role to both confirm the diagnosis as well as ascertainthe primary site of origin of tumor. On IHC neuroendocrine markers, such aschromogranin A, synaptophysin, and CD56, aid in the diagnosis of these tumors[5] CECT ofchest abdomen and pelvis can help define the regions of disese involvement. Imaging modality of choice is PET- CT as it shows the of stage of metabolicallyavid disease and is important in follow up of the patient.[6] Chemotherapyand radiotherapy are more effective treatment choices rather than surgery.

CASE REPORTA

63 year old womanpresented to gynaecology OPD of Dr RPGMC Tanda, with complain of lower abdominal pain radiating to back, postmenopausal bleeding hematuria and dyschasia since 15 days. Postmenopausal bleeding was mild, with sokage of only 1 pad per day. She had undergone abdominal hystrectomy with bilateral salpingophorectomy for AUB 11 years back at our institute, but no recordsof operative findings and histopathology examination were available. There wasno postoperative complication. She was hypertensive and diabetic from 15 years, on medication for the same.

Per speculum examination revealed a well-defined mass of approx. 4* 4 cm projecting from the vault. On per vaginal examination a fraible growth was arising from vault around 4*4 cm which bleeds on touch. Colposcopic examination was also done (Figure: 1). Onroutine investigations CBC, LFT, RFT were in normal limits and there were nored blood cells in urine routine microscopy.

USG pelvic organ revealeda well-defined 4.9* 5.4 cm solid hypoechoic lesion with few small cystic areasin between. CECT abdomen and pelvis (Figure: 2) showed large ill-defined heterogeneously enhancing necrotic mass in pelvis showing internal air foci, invasioninto posterior wall of urinary bladder and vaginal vault, abutting the distalrectum and anorectal region. With multiple heterogeneously enhancing pelvic andretroperitoneal necrotic lymph node suggestive of neoplastic etiology.

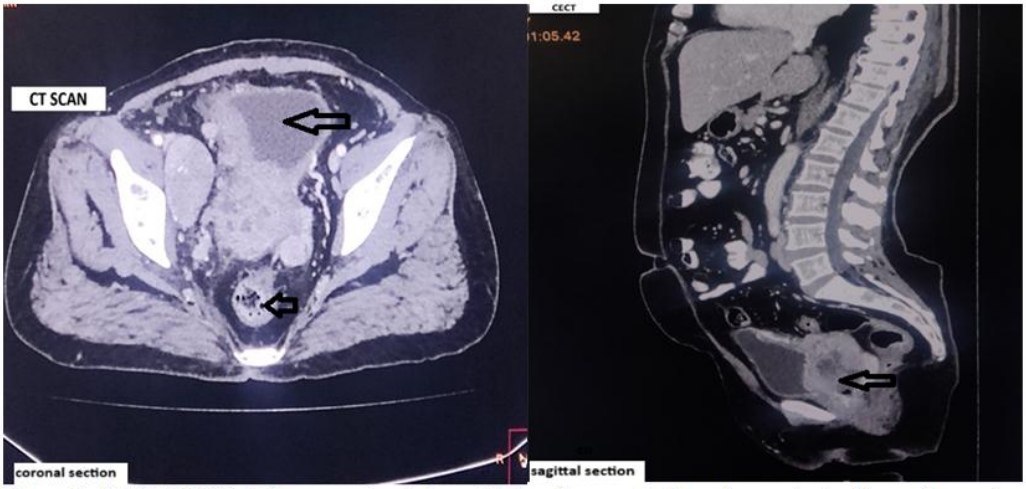

Figure 2: Initial CECT abdomen and pelvis sagittal and coronal views demonstrate (shown by ⇦) a large ill-defined heterogeneously enhancing necrotic mass in pelvis showing internal air foci, invasion into posterior wall of urinary bladder and vaginal vault, abutting the distal rectum and anorectal region.

Vault biopsy was performed, microscopic sections (Figure: 3) showed lobules of monomorphic cells having high N/C ratio and frequent atypical mitotic figures, suggestive of poorly differentiated carcinoma. Immunohistochemistry confirmed the diagnosis of small cell neuroendocrine carcinoma (SNEC) i.e. synaptophysin immunoreactive in 76-100% cells, INSM 1 positive in 51-75% cells and negative for CD 45 and DESMIN.

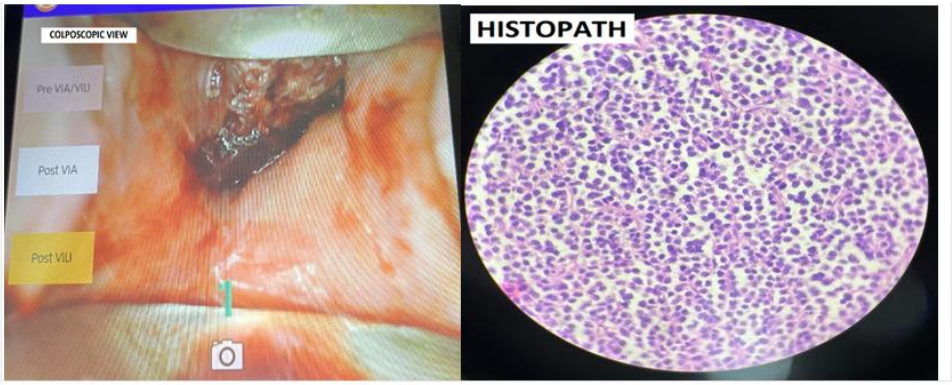

Figure 1: Coloposcopic image showing friable growth
Figure 3: Histopathology of the vaginal vault growth showing showed lobules of monomorphic cells having high N/C ratio and frequent atypical mitotic figures

Volumetric modulated arc therapy technique of radiotherapy and 3 cycles of brachytherapy. Follow up CECT at 6 months revealed complete regression of lesion with few enlarged lymph nodes in pararectal and bilateral iliac region. After follow up of 1.5 years there is no locoregional recurrence or distant metastasis.

DISCUSSION

Tumor arising from the diffuse neuroendocrine cell system comprise NET tumors, these tumors are commonly identified in the gastrointestinal tract, pancreas, lung and thymus. Small cell carcinomas (Sm CCs) arise in the lung and only 5% are extrapulmonary.[7] Cervical small cell carcinoma and ovarian carcinoids are the most prevalent gynaecological NE tumors.[8] SmCC predominantly affects postmenopausal women, typically between the ages of 38 and 74. It presents with a wide range of symptoms, from no symptoms at all to more noticeable signs like painful intercourse (dyspareunia), burning during urination, postmenopausal bleeding, and the presence of a vaginal mass, as in our case. Early detection is crucial for better outcomes, as these symptoms can sometimes be mistaken for less serious conditions. The family of well differentiated neoplasms (i.e. carcinoid and atypical carcinoid) is morphologically and clinically distinct from high-grade neuroendocrine carcinoma (i.e., small cell and large cell). This latter entity is closely related to pulmonary small-cell carcinoma, is highly aggressive, and is generally managed with a multimodality approach including platinum based chemotherapy. Well and poorly differentiated NE tumors are grouped together only because of generic neuroendocrine marker expression (i.e. expression of the markers synaptophysin and chromogranin detected by immunohistochemistry). Due to increased diagnostic recognition and use of IHC there

has been an increased reported incidence of NE tumors. These rare tumors should be reported as there is limited data to guide decisions in case of NET. Small cell carcinomas are characterized by high mitotic rate, extensive necrosis, frequent lymphovascular space involvement (LVSI) and a strong association with HPV 18.[8] Use of chemotherapy and radiotherapy have shown promising results in management of SCC NET as reported by Hoskins et al. Hoskin et al managed series of 31 patients using paclitaxel, cisplatin and etoposide followed by EBRT, 3 year failure free survival for patients with early stage disease was 80%.[9] Sevin et al manged patients with surgery and radiation the 5 year survival rate was 33.5 %.[10] In our case after chemotherapy and radiotherapy patient had complete regression of lesion and no clinical symptoms after 1 year follow up.

CONCLUSION

This case highlights the importance of early recognition, accurate diagnosis, and tailored treatment approaches in the management of vault carcinoma, particularly in the setting of rare histological subtypes such as SCC NET. Continued efforts to enhance our understanding of this disease entity are essential for improving outcomes and quality of life for affected individuals.

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