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Vaginal Small Cell Neuroendocrine Carcinoma: A Case Report

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Abstract We report a case of small cell neuroendocrine carcinoma of the vagina treated with primary resection with positive margins and subsequent chemoradiation. This rare case highlights the importance of a thorough gynecologic exam, full evaluation for any and all abnormal findings, and a comprehensive care team.

Keywords: *Vaginal tumor, small cell carcinoma, vaginal mass*

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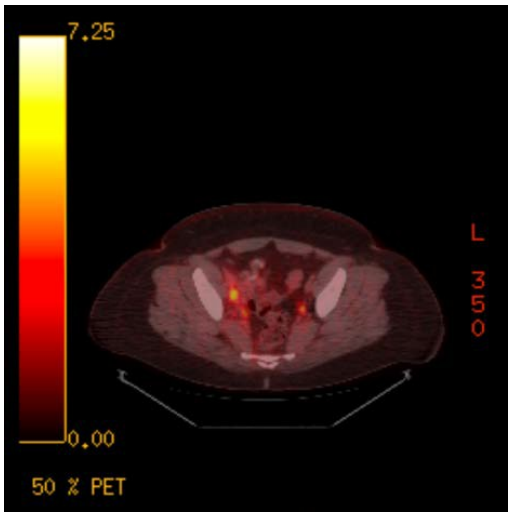
1. Introduction

Vaginal neuroendocrine tumors, including both small and large cell forms, are rare, with limited understanding of risk factors and treatment recommendations. There is also limited evidence for how to evaluate disease burden and response to treatment, although fluoro-2-deoxy-D-glucose (FDG) PET/CT scans have demonstrated efficacy for detecting lymph node involvement or other spread of disease, as well as to detect local recurrence or response to treatment. Disease outcomes are poor, as the tumors tend to be aggressive with early spread and poor response to treatment. We report a case of small cell neuroendocrine carcinoma of the vagina treated with primary resection with positive margins and subsequent chemoradiation. We explore the importance of thorough gynecologic exam, full evaluation for any and all abnormal findings, and a comprehensive care team.

2. Case Report

Patient is a 34yo G0 with history of HPV-negative atypical squamous cells of uncertain significance (ASCUS), fibroids, primary ovarian insufficiency, obesity, and hypertension presented to her primary gynecologist for abnormal uterine bleeding. Exam at the time was unremarkable and the patient opted to return if a second episode occurred. She returned 6 months later for a second abnormal cycle. Further workup was performed including repeat Pap smear, which was significant for high-risk HPV-positive atypical squamous cells, cannot exclude high grade squamous epithelial lesion (ASC-H). Colposcopy and endocervical biopsy were performed and within normal limits. She was recommended to undergo LEEP at this time but deferred. No lesion was noted at either of those visits or procedures.

She followed up 1 year after her initial visit for secondary amenorrhea. On physical examination at that time, there was noted to be a soft, ballotable lesion arising from the right lateral vaginal sidewall, concerning for a Gartner's duct cyst. She subsequently underwent hysteroscopy, dilation and curettage (D&C), loop electrosurgical excision procedure (LEEP), and excision of vaginal lesion the following month. Pathology of the LEEP demonstrated scant detached atypical cells and squamous metaplasia. D&C pathology demonstrated endometrial polyps with proliferative endometrium and a benign endocervix.



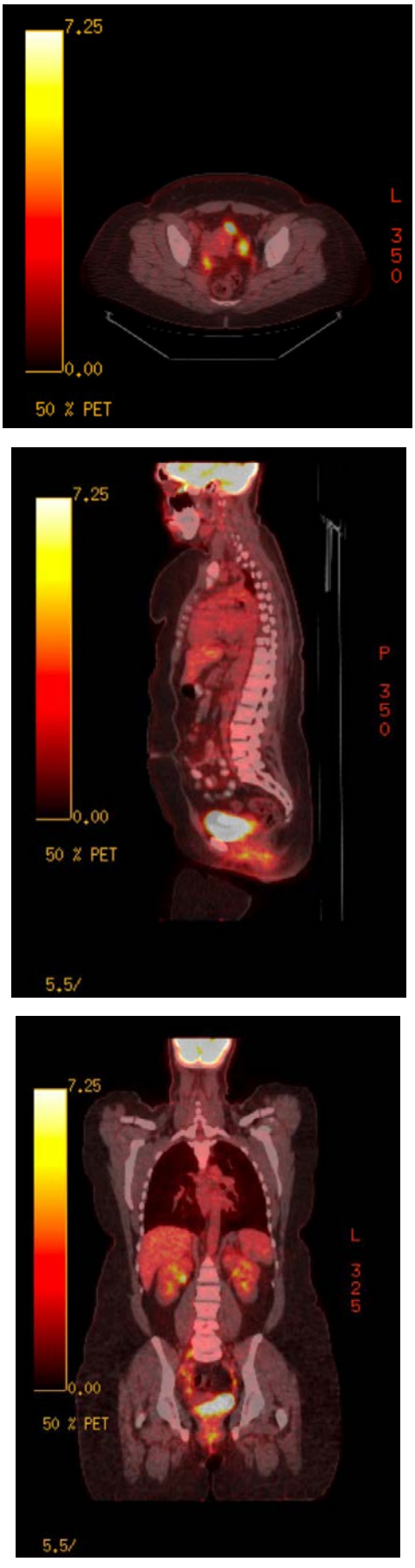


Figure 1. Multiple FDG avid enlarged lynmph nodes are seen in the right iliac and lower retroperitoneal lymph nodes

At surgery, the approximately 3cm lesion was noted to be firmer and pedunculated arising from the lateral vaginal right fornix adjacent to the junction of the cervix at 10 o'clock. It was resected at its base and felt as though it was removed en toto. Pathology of the vaginal lesion demonstrated high-grade neuroendocrine tumor with carcinoma extending to the biopsy margins. Immunostains demonstrated positive CD56, chromogranin, synaptophysin, and PCK. Stains for CK20, HMB45, and S-100 were negative. PET-CT performed the same month demonstrated fluorodeoxyglucose (FDG) avid enlarged right iliac and lower retroperitoneal lymph nodes consistent with lymph node metastases. No FDG avid distant disease was noted (Figure 1 and Figure 2).

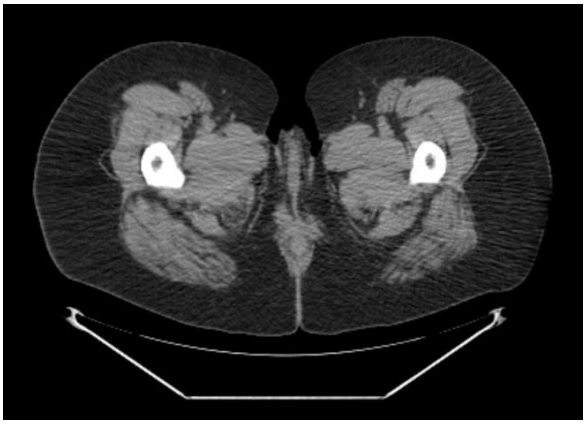


Figure 2. (A) CT showing mass in vagina

Patient was referred to oncology specialists at a tertiary care center including gynecologic oncology, medical oncology, and radiation oncology.

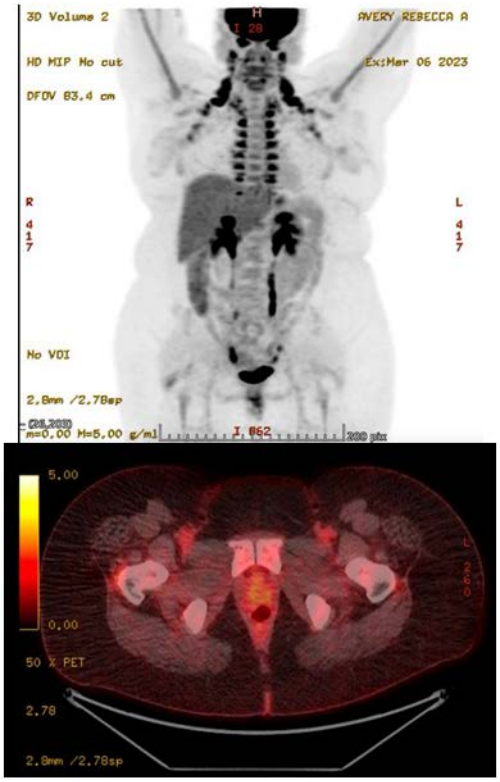


Figure 3. (A) 3/6/2023 Body FDG PET/CT: Resolved FDG avid retroperitoneal and pelvic nodal metastases. No new FDG avid metastatic disease seen. (B). No suspicious uptake in the vagina

Patient was diagnosed with stage IIIc2 small cell carcinoma of the vagina, which was determined to not be

amenable to surgical resection. She was treated with chemoradiotherapy starting 14 months after her initial visit. Treatment included 25 treatments with external beam radiation, as well as high-dose rate interstitial brachytherapy with 4 fractions (2400cGY). She underwent 5 cycles of cisplatin/etoposide. At this time, 19 months after her first visit and 6 months after initial resection, she was noted to have non-avid PET-CT consistent with successful treatment (Figure 3 and Figure 4).

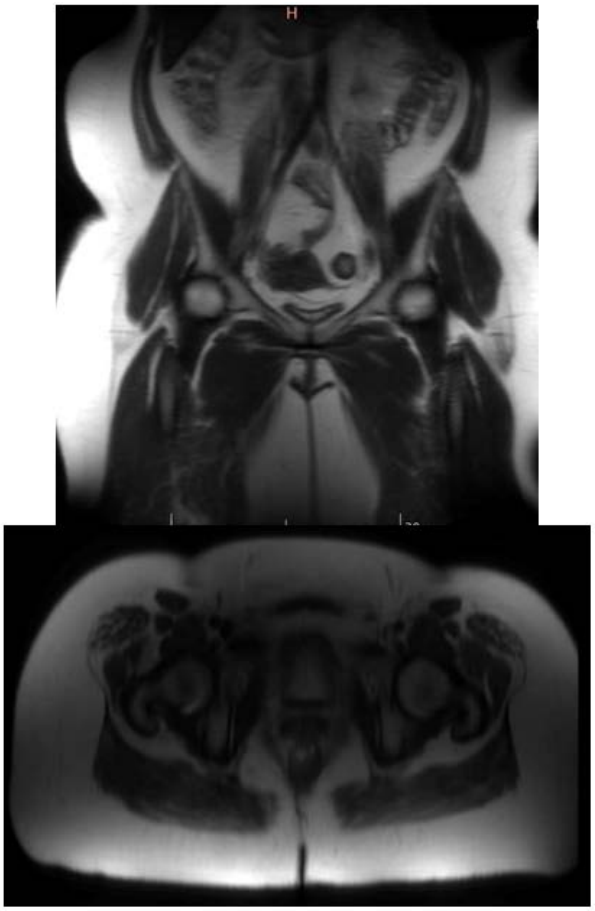


Figure 4. (A) 3/6/2023 MRI of pelvis- no mass in cervix or vagina. (B) Decreased and resolved pelvic nodes

3. Discussion

Historically, gynecologic small cell neuroendocrine carcinoma is a rare form of cancer with poor survival rates of approximately 15% at 1 year after diagnosis, with median time to relapse of 8 months from initial treatment in older studies [1]. In a 2004 review of 21 cases, no patients with lymph node metastases survived [2]. Vaginal cases remain one of the least common forms of this disease [3]. Although additional isolated cases have been studied over the subsequent decades, treatments continue to vary widely with poor survival rates [4,5]. Most evidence comes from treatment of the more prevalent small cell cancer of the lung.

This case initially presented as a typical, uncomplicated ASC-H for which the algorithm for SIL (Squamous Intraepethelial Lesion) was followed. As it turns out, the ASC-H was in reality a result of the evolution of small cell neuroendocrine tumor as patient's colposcopy, LEEP, and endocervical curettage were negative for SIL/CIN.

This case highlights the importance of a thorough gynecologic exam. A gynecologic exams should be a full evaluation for any and all abnormal findings, not merely a pap smear. All abnormal findings should be appropriately evaluated and biopsied with proper consideration for surgical planning and referred to the appropriate tertiary/quartenary care experts upon diagnosis.

Certain considerations should be taken for surgical planning in unique cases such as these. Firstly, surgery

should be done under ideal conditions with adequate anesthesia to facilitate a complete exam and resection. This case was done under MAC sedatation at an ambulatory surgery center with access to a tertiary care hospital if needed.

Futhermore, when unusual findings are noted such as change in the character of the lesion initially diagnosed (Gartner's duct cyst) to a more ominous pre-op appearance, there should be higher index for suspicion for unusual pathology and surgical planning should change accordingly. Although this lesion was excised at its base, there was a subcentimeter margin. Perhaps consideration should have been taken for a wider margin approaching two centimeters. Certainly we would not advocate perfomring a PET scan for everyone with ASC-H pap, albeit for a patient with a rare finding perhaps pre-op workup with imaging may be considered.

In order for the patient to receive comprehensive care, it is important for referalls to be given to tertiary or quarternary care centers in rare cases such as these due to lack of experience of even the most experienced gynecologist oncologist.

Our patient is still relatively early in her treatment course after initial local resection, but has thus far been successfully treated with cisplatin, etoposide, external beam radiation, and interstitial brachytherapy. These results are consistent with existing evidence that small cell gynecologic tumors should be treated with etoposide combined with a platinum-based agent, regardless of disease stage [6].

In the setting of negative evidence of disease within 5 months of diagnosis, patient will continue to require close follow-up to evaluate for recurrence. We hope that aggressive early management and close follow-up with PET-CT with FDG avidity will allow for longer progression-free survival.

Fertility preservation is an important consideration for young oncology patients and should be discussed. Our patient desired to preserve fertility. She chose to undergo oocyte retrieval prior to initiating chemoradiation therapy but did not have adequate retrieval. As a sequale of these events, patient was subsequently diagnosed with depression. She was referred to psychotherapy which was successful.

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