Neuroendocrine tumour of the cervix: A case report and review of literature

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Abstract

Cervical neuroendocrine tumours are rare accounting for only 2% of cervical cancers. They pose a management challenge. We present a case of a 26-year old who presented with bleeding *pervaginum* three months post partum and a cervical mass on speculum examinations. Further examination and histology revealed an advanced Stage 3b, poorly differentiated tumour which was confirmed to be neuroendocrine in origin on immunohistochemistry. Unfortunately the patient succumbed to the illness one month after diagnosis.

Key words: Neuroendocrine, Cervix, Tumour, Small cell carcinoma

Introduction

Neuroendocrine tumours of the genital tract are rare and pose a real management challenge to clinicians. Indeed, small cell carcinomas of the cervix account for only 2% of cervical cancers identified on histology (1). With better and advanced diagnostics including immunohistochemistry we may see an increase in the incidence of these tumours. However, diagnosis still remains a challenge in low resource settings, as it requires special staining that may not be readily available locally. Once diagnosis is made the treatment modalities include surgical excision, platinum based chemotherapy and radiation therapy (2). There have been few cases of these tumours reported in literature, with the youngest patient successfully treated being an 18-year old woman (3). Treatment is based on the consensus developed by the Society of Gynaecology Oncology (4). We present our experience with this tumour in a young lady presenting postpartum with the aim of contributing to the body of knowledge on the subject.

Case presentation

A 26 year-old woman was referred to us from a district hospital with bleeding *per vaginum* and a feeling of heaviness in the perineum three months after a normal vaginal delivery. On speculum examination she was found to have a cervical mass. She underwent a further staging examination and a biopsy was taken for histopathology. She was initially categorized as cervical cancer stage III B and counseled appropriately. As part of her workup, she was noted to have deranged renal functions with elevated serum creatinine. She was consequently commenced on renal haemodialysis. Final histology revealed a malignant poorly differentiated neoplasm. Immunohistochemistry results were as follows: tumour cells negative for pankretin (SCC/ Adeno), CD10 (endometrial), CD45 (Leucocyte common antigen-Lymphomas) and S100 (neural/ melanoma) and diffusely positive for synaptophysin (neural marker that gives granular staining pattern). Figures 1-5 are sample slides of the tumour.

Histology slides:

Figure 1: Nuclear moulding on H&E staining

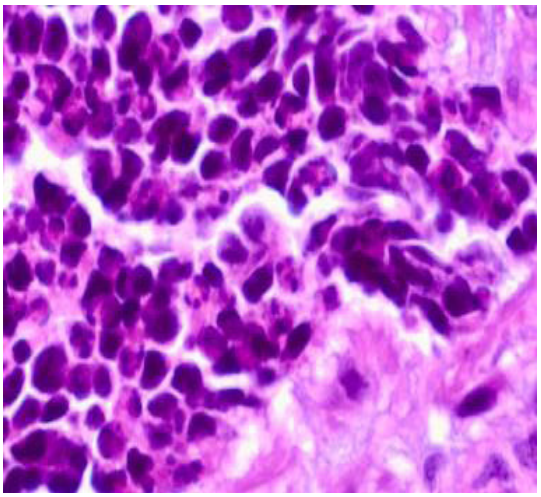


Figure 2: Small crowded cells

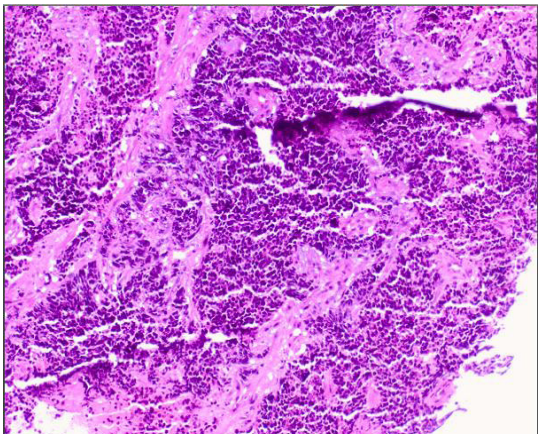


Figure 3: diffuse infiltration of cervical stroma by tumour cells

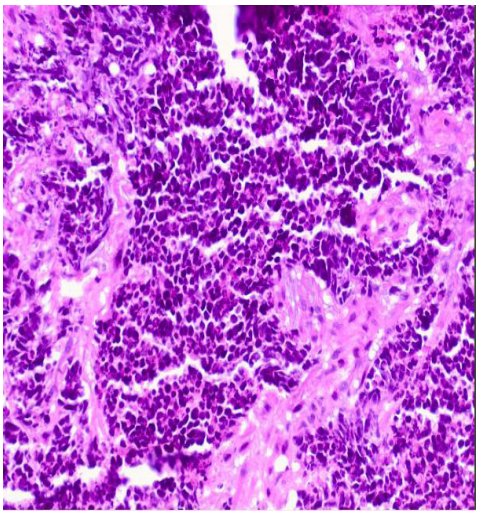


Figure 4: Synaptophysin uptake by tumour

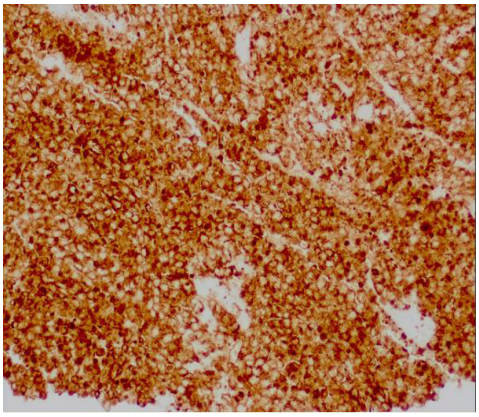
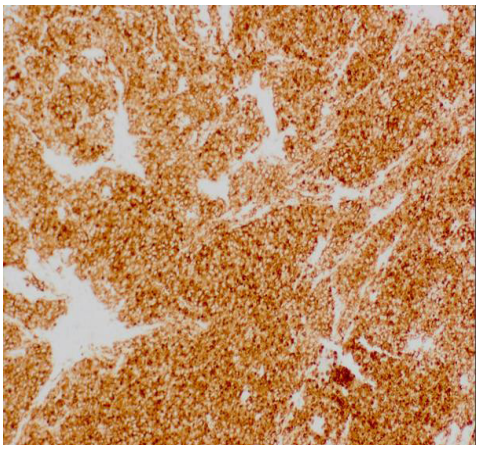


Figure 5: No uptake of synaptophysin by normal tissue



Discussion

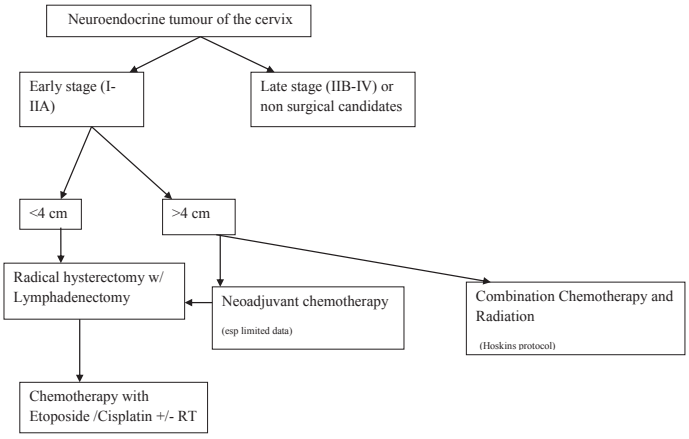
Neuroendocrine tumours of the cervix are rare; consequently their treatment as for all small cell carcinomas of the cervix is mainly hypothetical, based on consensus as opposed to strong clinical trial-based evidence (3). Furthermore most cancers of the cervix tend to have a worse prognosis if diagnosed post partum (5). Based on reported cases and reviews most of the patients with small cell carcinomas who have survived beyond 5 years were in early stages and managed aggressively. According to the Society of Gynaecologic Oncology, these tumours are given histologic classification similar to that of pulmonary nomenclature (4).

The median age of presentation for most small cell carcinomas is the 5th decade. Our patient presented at age 26, which is quiet young. The usual presenting symptoms are bleeding per vaginum with a cervical mass. Very rarely these patients will have features of ectopic hormone production (4).

Staging of the tumours follows the traditional cervical cancer staging (4). Treatment options are drawn from data on pulmonary small cell carcinomas includes the use of platinum base compounds +/- etoposide. Radiation is used for limited stage disease. Surgery appears to have better outcomes than the above modalities. Vincristine/doxorubicin/ cyclophosphamide and topotecan are considered as alternate or second-line therapies extrapolating from small cell lung cancer (4). For limited stage small cell lung cancer, 20% of cases are long-term survivors. In comparison, small cell cervical cancers have a reported 5-year survival of 36%. Approximately half of cases are an early clinical stage, 41% stage 20% and I stage II, and approximately 50% have positive nodes at diagnosis. For early stage disease treated with multi-modality regimens, recent reports have achieved an 80% 3-year disease free survival.

Recommendation of treatment

Figure 6: With permission from the SGO paper (4).



Conclusions

Neuroendocrine tumours of the cervix are rare and hold a poor prognosis due to their aggressive nature. Systems are needed to make a prompt diagnosis and allow speedy initiation of treatment. The case adds to the body of knowledge on these tumours and to lay emphasis on the aggressiveness of this tumour.

References

1. Liao L-M, Zhang X, Ren Y-F, *et al.* Chromogranin A (CgA) as poor prognostic factor in patients with small cell carcinoma of the cervix: Results of a retrospective study of 293 patients. Rota R, ed. *PLoS ONE*. 2012; **7**(4):e33674. doi:10.1371/journal.pone.0033674.
2. D'Antonio A, Addesso M, Caleo A, Guida M, and Zeppa P. Small cell neuroendocrine carcinoma of the endometrium with pulmonary metastasis: A clinicopathologic study of a case and a brief review of the literature. *Ann Med Surg (Lond*). 2016; **5**: 114–117.
3. Wang Q, Liu Y-H, Xie L, Hu W-J and Liu B-R. Small cell carcinoma of the uterine cervix in pregnancy: A case report and review of the literature. *Oncol Lett*. 2015; **9**(1): 91–95.
4. Gardner GJ , Reidy-Lagunes D and Gehrig PA. Neuroendocrine tumors of the gynecologic tract: A Society of Gynecologic Oncology (SGO) clinical document. *Gynecol Oncol.* 2011; **112:** 190–198.
5. Sood AK, Sorosky JI, Mayr N, Anderson B, Buller RE and Niebyl J. Cervical cancer diagnosed shortly after pregnancy: prognostic variables and delivery routes. *Obstet Gynecol.* 2000; **95**(6 Pt 1):832-838.