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Large Cell Neuroendocrine Carcinoma of the Uterine Cervix: A Case Report and Review of the Literature

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Abstract **Case Report**

Large cell neuroendocrine carcinoma (LCNEC) of the uterine cervix is a rare and aggressive malignancy with poor prognosis even in its early stage, despite multimodality treatment strategy. Here, we report a case of a woman with LCNEC of the cervix, which was detected by pelvic pain. A literature review was also conducted to evaluate current therapeutic approaches and potential new strategies.

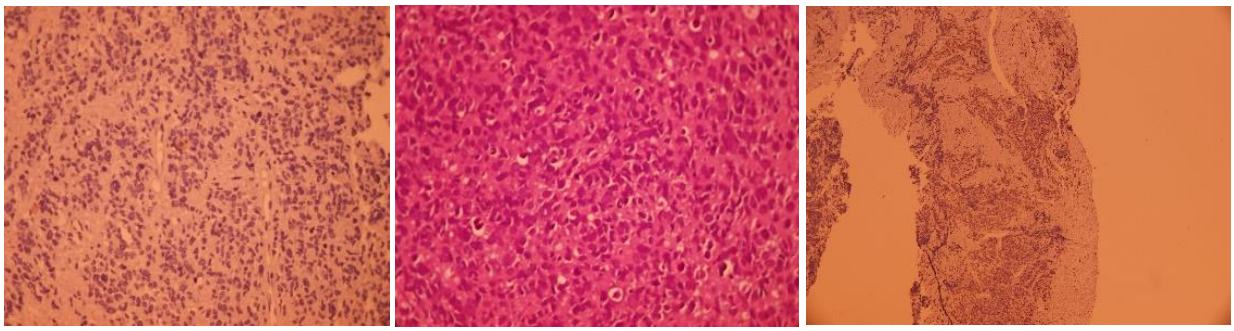
Keywords: Large cell neuroendocrine carcinoma, carcinoma of the uterine cervix, Brain metastases in (LCNEC), chemotherapy, etoposide and cisplatin.

INTRODUCTION

Neuroendocrine tumors of the uterine cervix represent 0.9 to 1.5% of cervical tumors [1]. They are rare, aggressive tumors with a bad prognosis and their therapeutic management remains a dilemma for several medical teams. We report a case of a neuroendocrine tumor of the uterine cervix at the center of onco-radiotherapy CHU MOHAMMED VI MARRAKECH.

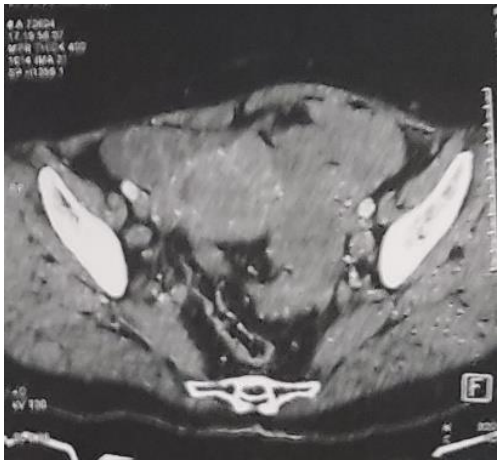
PATIENT AND OBSERVATION

The patient was 55 years old, single, menopausal, with no other personal or family history. The patient consulted for pelvic pain evolving for 7 months before her admission, without digestive or urinary signs or other associated gynecological signs. The whole evolving in a context of alteration of the general state. On examination: A firm mobile pelvic mass was found reaching the level of the left iliac fossa. On pelvic touch, a 10 cm process was noted at the level of the uterine cervix, infiltrating the vaginal pouches which reached the junction of the middle 1/3 of the vagina, the parameters were infiltrated. Biopsies carried out at this level, came back in favor of a malignant tumor proliferation of little differentiated carcinomatous appearance, seat of undifferentiated tumor proliferation made of large cells sometimes round and sometimes elongated. A complementary immuno-histochemical study of the different fragments showed that the carcinomatous cells were marked with antibodies: anti-synaptophysin, anti epithelial membrane antigen, anti chromogranin, anti CD99 lanti pancytokeratine. This makes this tumor compatible with a differentiated large cell neuroendocrine carcinoma of the uterine cervix.



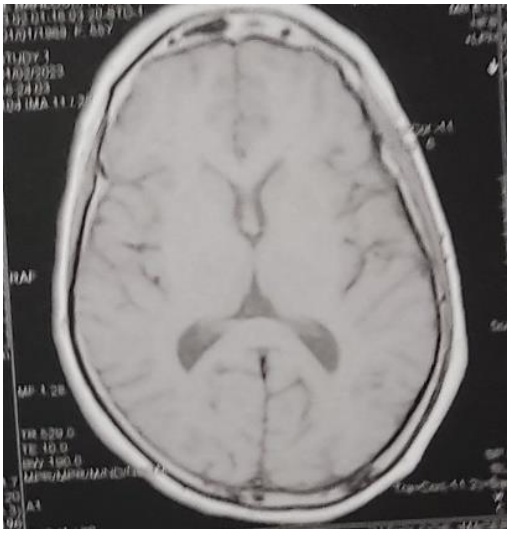
Histological appearance of Large cell neuroendocrine carcinoma

PELVIC MRI reveals a uterine tissue mass infiltrating the cervico-isthmic region 83*84*50mm, the endometrium and myometrium with multiple mallimited pelvic tissue masses encompassing the sigmoid, we note a diffuse infiltration of pelvic fat and the presence of internal and external iliac adenopathy, inguinal adenopathy measuring for the most voluminous 25*17mm external left iliac then the patient presented a brutal installation of consciousness disorders with GCS 10/15.



PELVIC MRI Axial sections T1 sequences showing locally infiltrating cervico-isthmic tumor process stage IVb

The cerebral CT scan showed nodular lesions in the right frontal parenchymal area measuring 12.2*9.5 mm, suggesting a secondary origin.



Axial slice injected brain CT showing right frontal intra parenchymal lesions

The decision taken at the multidisciplinary consultation meeting was to perform etoposide and cisplatin chemotherapy for 3 courses, followed by evaluation. The patient evolved unfavorably during the 5 months following the start of chemotherapy with HTIC syndrome with the onset of engagement cerebral which caused her death in the emergency room.

DISCUSSION

Neuroendocrine tumors are mainly found in the digestive tract and the lungs; in the cervix, they represent only 0.9 to 1.5% of cervical tumors, which are generally predominated by squamous cell carcinoma [1].

They are distinguished from squamous cell carcinomas by their higher recurrence rate and the delay in their diagnosis due to the inefficiency of screening for this type of tumor by the cervico-uterine smear. On the other hand, the association with the "human papillomavirus (HPV) 16 and 18" constitutes a common risk factor between the two carcinomas. Moreover, according to a study carried out in 2018 concerning the contribution of HPV on the formation of neuroendocrine tumors of the cervix in a series of 10575 cases of invasive tumors of the cervix; HPV DNA was detected in 85.7% of the cases of neuroendocrine tumor (HPV16 54.8% and HPV18 40.5%) [2].

Due to the rarity of cervical LCNEC, no consensus has been reached on an optimal treatment plan, and current multimodal strategies that combine radical hysterectomy (with or without bilateral salpingo-oophorectomy), chemotherapy and radiation are mainly adapted from treatments used for neuroendocrine carcinomas of the lung [3].

While the role of surgical intervention remains controversial with no sign of improving long-term survival so far, chemotherapy has become the mainstream of management, especially considering LCNEC's aggressive, early metastatic behavior [4].

Among various chemotherapy options, the combination of cisplatin and etoposide is most common and was used for our patient as well. Three other commonly used combinations include vincristine, doxorubicin and cyclophosphomide, carboplatin plus paclitaxel and occasionally epirubicin, topotecan plus thalidomide. A recent systematic review of published cases has also indicated that, while the addition of chemotherapy at any point of initial treatment will offer survival benefits, platinum with (P=0.0027) or without etoposide (P=0.0034) in particular is associated with statistically significant improvement in survival compared to chemo-regimens without these agents. However, regardless of great efforts invested, the majority of LCNEC patients do not survive more than two years after being diagnosed. Accordingly, novel treatment strategies have been proposed that require further evaluation [5].

Tangjitgamol *et al*., measured the prevalence of estrogen receptor and/or progesterone receptor among neuroendocrine tumors to evaluate the feasibility of applying hormonal treatment to cervical LCNEC patients, but unfortunately only a very small portion of recruited patients (3 out of 24) expressed these hormonal receptors [6].

Another strategy was proposed by Kajiwara *et al*., using the somatostatin type 2A (SSTR2A) analog, octreotide, to treat neuroendocrine tumors, given that 3 out of 7 cases (2 out of 5 SCC and 1 out of 2 LCNEC) expressed SSTR2A receptors; however, this strategy has not been tested yet in a larger study [7].

In conclusion, chemotherapy is associated with improved survival and should be considered in resected cases.

CONCLUSION

Neuroendocrine tumors of the uterine cervix are very aggressive and rare tumors, which explains the lack of randomized trials and makes their management increasingly difficult.

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