Small Cell Neuroendocrine Carcinoma Of Endometrium: A Case Report


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ABSTRACT

Small cell gynecological neuroendocrine carcinomas are unusual, and they account for only <1% of endometrial tumors. Given their rarity and the absence of randomized trials, the diagnostic and therapeutic management of these tumors is difficult and is essentially modeled on that of pulmonary neuroendocrine tumors. We report a case of a patient with a small cell neuroendocrine carcinoma of endometrium and through the data of the literature we point out the different aspects of this rare entity.

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INTRODUCTION:
Small cell carcinoma is a type of neuroendocrine cancer that starts in cells of the neuroendocrine system. It tends to be aggressive and is associated with a less encouraging prognosis, even if diagnosed at an early stage (1,2).

Clinical Case
We report the case of a 56-year-old postmenopausal patient with no specific history admitted for low abundance bleeding with no other associated signs and in whom clinical examination revealed no abnormalities. The patient underwent pelvic ultrasound showing an endometrial thickening of 9 cm long axis requiring biopsy curettage, the histological result of which was in favor of carcinomatous proliferation with neuroendocrine differentiation. The extension assessment was supplemented by a pelvic MRI which showed the presence of a large intra-cavity circumferential isthmo-corporeal lesion with presence of deep peri-aortic lymphadenopathy, primary internal and external iliac and images in favor of peritoneal carcinosis, the procedure to be followed was to perform a hysterectomy without adnexal preservation and omentectomy. On gross examination, the uterine wall was the seat of a 9x6x3 cm ulcerative-bluish whitish-colored, firm consistency occupying the entire uterine cavity and infiltrating the wall (Figure 1).

![Figure 1: Macroscopic image of an ulcerative-budding whitish neoplasm, occupying the entire uterine cavity, and infiltrating the wall](image-url)
The histological examination at low magnification found an invasive carcinomatous proliferation arranged in layers, in acini, in clusters and in isolated cells (Figures 2 and 3). With a richly vascularized stroma. The tumor cells were small to medium in size, with anisokaryotic nuclei, hyperchromes and seat of...
mitosis. The cytoplasm was eosinophilic and reduced. The stroma reaction was fibro-inflammatory with the presence of vascular embolism. This proliferation infiltrated the uterine wall over 50%, the isthmus, the right and left parameters and the right and left horns. An immunohistochemical complement was carried out showing: a positivity of the tumor cells to the anti chromogranin and anti synaptophysin antibodies, an absence of expression of the tumor cells to the hormonal receptors and to the anti CK7 (Figure 4) and anti CK20 anticoprs. The tumor proliferation index (Ki67) was 70%. The diagnosis was grade 3 small cell endometrial neuroendocrine carcinoma. The indication retained after multidisciplinary consultation was to start first line chemotherapy with cisplatin etopside.

**DISCUSSION**

Neuroendocrine carcinoma is a rare and aggressive malignant tumor, developing mainly at the expense of the lung and digestive tract. It represents only <1% of endometrial tumors which are predominantly adenocarcinomas. WHO 2020 defined these entities as a group of neoplasms of the genital tract having a neuroendocrine phenotype with 3 subtypes (large cell, small cell carcinoma and carcinoïd tumor) (1.2).

It is usually seen after menopause with an average age of 60 years for small cell neuroendocrine carcinomas, and 55 years for large cell neuroendocrine carcinomas (3).

The clinical symptomatology is not very specific which can manifest itself as post menopausal metrorrhagia, a pelvic mass or rarely a hormonal secretion (cushing syndrome, carcinoïd syndrome, hypoglycemia, syndrome of inappropriate secretion of antidiuretic hormone, hypocalcemia). paraneoplastic syndrome. This lack of specificity makes the diagnosis purely histological and immunohistochemical (4,5).

The abdomino-pelvic or endovaginal ultrasound is the first-line examination for endometrial thickening. The extension assessment is mainly based on abdominopelvic MRI to assess pelvic extension and also CT for distant metastases. In our patient, the extension assessment was negative. Histologically, these tumors are organized in cords and nests composed of a population of weakly cohesive ovoid cells of small or intermediate size, hyperchromatic nuclei condensed with an abundant cytoplasm, and a high mitotic rate. Diffuse or single-cell necrosis and vascular invasion are generally present. The positivity of an endocrine antibody is mandatory for a positive diagnosis. The differential diagnosis is made with mixed Mullerian tumors, lymphomas, sarcomas, endometrial hyperplasia and metastatic neuroendocrine carcinomas. Endometrial small cell carcinoma is aggressive. An advanced stage is generally observed at the time of diagnosis (1.6,7).

In most small published case series, the 5-year survival rate ranged from 17% to 64%, with a median survival between 12 months and 21 months. Surgery is the cornerstone of treatment. In cases of advanced disease, adjuvant radiation and chemotherapy can be used alone or in combination, which is similar to the therapies used for endometrial carcinoma (1,10).

**CONCLUSION**

Neuroendocrine carcinomas of the endometrium are rare and aggressive tumors. They do not represent any clinical or radiological specificity making the diagnosis essentially anatomopathological and immunohistochemical. The rarity of these tumors and the limited number of series did not allow their proper analysis in order to determine effective treatments and improve patient survival.

**AUTHORS' RESPONSIBILITIES**

Imane.Boujguenna, Salma Belmaachi, Fatima.Boukis and Houda Jouihri: drafting of the manuscript

Anass Fakhri and Hanane Rais: correction of the manuscript

Abderraouf Soummani, Rhizlane Belbaraka and Houda Jouihri: clinical and surgical management of the patient

Chihab bouyaali and Najat Cherif Idrissi Gannouni: radiological follow-up of the patient

All authors contributed to the conduct of this work

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