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Small cell carcinoma of the endometrium: A clinicopathological study and management of three cases

Fatoumata Matokoma Sidibe [1,2], Zakaria Traore [2], Aspasia Georgala [1], Rajae Kanab [2], Denis Larsimont [1], Ahmad Awada [1], Martine Piccart-Gebhart [1]

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1. Université libre de Bruxelles, institut Jules-Bordet, Brussels, Belgium
2. Centre hospitalier universitaire Hassan II, Department of medical oncology, Fez, Morocco

Correspondence:
Fatoumata Matokoma Sidibe, Centre hospitalier universitaire Hassan II, Department of medical oncology, Fez, Morocco.
fatsi_2@hotmail.com

Carcinome à petite cellule de l'endomètre : étude clinicopathologique et prise en charge de trois cas

Keywords
Endometrial small cell carcinoma
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Literature review

Summary

Endometrial small cell carcinoma (ESCC) is an extremely rare and aggressive tumor with poor prognosis. It is characterized by early regional and systemic spread leading to rapid development of lymph nodes, pelvic and extrapelvic metastasis and compromising the outcome. In this paper, we reported three cases of ESCC confirmed by pathological and immunohistochemistry studies. In one case, ESCC was associated with endometrioid carcinoma and carcinosarcoma, while the other two cases were pure ESCC. Two cases were diagnosed at early stage IA of the International Federation of Gynecology and Obstetrics (FIGO) cancer staging system. They were treated by surgery followed by pelvic external radiation and brachytherapy with favorable outcome (no recurrence was confirmed and a survival was 1 and 5 years, respectively). The third case was diagnosed with visceral metastasis and was treated with 6 cycles of cisplatin plus etoposide. She died 8 months after diagnosis. Due to its rarity, there is no standard guideline for the management of ESCC. Its treatment is extrapolated from that of both, the conventional endometrial carcinoma and the small cell carcinoma of the lungs, which share similarities with ESCC. Thus, multimodal therapeutic including surgery, radiation therapy and chemotherapy, seems to be the best therapeutic approach. Randomized clinical trials with multiples cases of ESCC are encouraged to clearly define the optimal therapeutic approach to this rare tumor.

Introduction

Small cell carcinoma (SCC) is a type of highly neuroendocrine tumor (NET) which arises from the diffuse neuroendocrine cell system [1]. Its diagnosis requires at least one positive neuroendocrine marker among chromogranin A, Neurone Specific Enolase (NSE), synaptophysin and neural cell adhesion molecule (NCAM, CD56).

There are three histological categories of NETs which consider the cell differentiation and its biologic aggressiveness (grade of the tumor): poorly-differentiated (high-grade tumor),

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moderate-differentiated (intermediate-grade) and well-differentiated (low-grade).

The SCC is a poorly-differentiated and high-grade tumor [2]. It is highly aggressive, with a propensity for systemic spread and a poor prognosis. Although NETs occur mainly in the lung, they can be found in a variety of organs such as the gynecologic tract. In the female reproductive system, SCC constitutes only 2% of all gynecological malignancies [3]. SCC occurs in order of decreasing frequency in the cervix, ovary, endometrium, vaginal and vulva [1–3].

Due to its rarity and lack of prospective data to guide decisions, there are no standard guidelines for management of endometrial SCC (ESCC). In this paper we report three cases of ESCC and we review its clinicopathological and prognosis features and its management.

Case report 1

A 50-year-old perimenopausal and woman smoker with personal history of inflammatory bowel disease presented with vaginal bleeding. Pelvic Magnetic resonance imaging (MRI) showed a 5 × 6.8 cm sized mass in the uterine cavity (*figure 1*) and Positron Emission Tomography scan (PET scan) showed only uterine hypermetabolism. Her Eastern Cooperative Oncology Group Performance Status (ECOG PS) was 0. She underwent surgery with radical hysterectomy plus bilateral salpingo-oophorectomy, pelvic and para-aortic lymphadenectomy and peritoneal cytology. The pathology study revealed a 6 mm sized poorly-differentiated endometrioid carcinoma associated with small round neuroendocrine cell and carcinosarcoma components, originated in the endometrium. The tumor invaded less than 50% of the myometrial surface. The small cell component was the most important of three components (which occupied more than half of the tissue). No tumor cells were identified at the junction of uterus and cervix, in the ovaries, ascitic fluid or in the nine removed nodes. Tumor was surgically classified as pT1aN0M0 and IA of the FIGO staging system. Immunohistochemical staining demonstrated that the small round cells were positive for synaptophysin (Syn), chromogranine A (Chr A) and neuron specific enolase (NSE). Endometrioid carcinoma was positive for estrogen receptor (ER), antikeratin (AK) AE1/AE3, cytokeratin 5/6, while carcinosarcoma was positive for PS100. There was no evidence of CA125, CEA, desmine and CD99. Patient was given a postoperative pelvic external radiation with 45 grays (Gy) in 25 fractions (Fr) and brachytherapy with unique Fr of 6 Gy. One year after surgery, the patient died of unknown cause but no recurrence was diagnosed before the death.

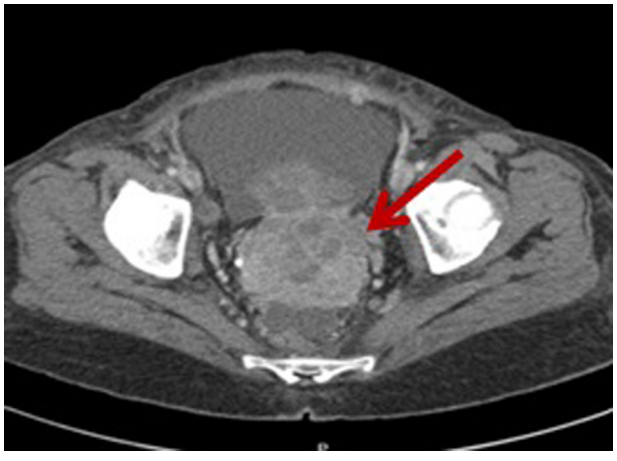

FIGURE 1

Case no. 1: pelvic MRI showing an intrauterine mass of 5 × 6.8 cm in size

Case report 2

An 80-year-old female with good ECOG PS 1, diabetes and deafness history suffering postmenopausal bleeding presented in our institution for physical examination. Pelvic MRI showed a centimetric intrauterine mass (*figure 2*). There were no distant metastases and patient underwent radical hysterectomy and salpingo-oophorectomy. The pathology study confirmed the presence of poorly-differentiated small round cells in the endometrium, which invaded least 50% of the muscle wall of the uterus. No tumor cells were identified at the junction of uterus and cervix or in the ovaries. Tumor was surgically classified as IA of the FIGO stage. Tumor cells were positive for Syn in immunohistochemical analysis, suggesting a SCC diagnosis. Patient was given a postoperative pelvic external radiation with 45 Gy in 25 Fr and brachytherapy with unique Fr of 6 Gy. She is currently alive 5 years after surgery.

Case report 3

A 47-year-old peri-menopausal and female smoker with ECOG PS 1 visited our institution because of vaginal bleeding, abdominal distension and abdominal pain. The abdominal and pelvic MRI showed a voluminous uterine mass extended to the left rectal, splenic mass, hepatic metastases, peritoneal effusion and

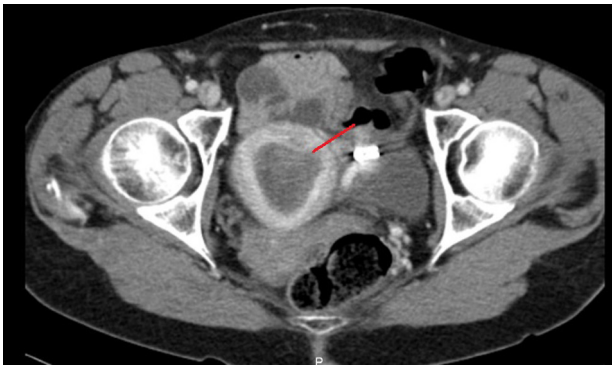

FIGURE 2

Case no. 2: pelvic MRI with uterine mass