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Case Report

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NEUROENDOCRIN CARCINOMA OF ENDOMETRIUM: REPORT CASE

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ABSTRACT

Endometrium cancer is the fourth gynecological cancer. Endometrioïd adenocarcinoma is the most frequent histological type but neuroendocrin carcinoma is very rare. We report the case of menopaused woman with neuroendocrin carcinoma revealed by acute abdominal pain in relation with hydrometria. The aim of our work is to focus on specificities of this exceptional pathological entity in diagnosis, prognosis and care.

KEYWORDS: Neuroendocrin carcinoma – endometrium - surgery –immunohistochemistry.

INTRODUCTION

Neuroendocrin tumors with small cells are commonly localised at lungs and digestive tract, endometrial localisation is exceptional. It's frequency is less than 0.8% of endometrial cancers. Because of it's rarety, only few cases were reported in literature, and no management consensus has been formally established until now. The target of our study is to report a new case of neuroendocrin carcinoma of endometrium, through which we will identify particularities of these tumors whose prognosis remains pejorative.

CASE REPORT

We report the case of 60 years old woman, mother of three children, menopaused since 10 years without HRT of menopause, without pathological history. She had isolated chronic pelvic pain without vaginal bleeding and without urinary or digestive signs. Physical examination found retroversed uterus, increased in size. Gynecological examination found normal cervix. Pelvic ultra-sound showed hydrometria and endometrial process, echogenic, irregular, heterogenous, localised in isthmus region and measuring 45/25mm.

A biopsy was done, histopathological examination found undifferentiated carcinoma process. Immunohistochemical study showed CD56 and cytokeratin compatible with the diagnosis of small cells neuroendocrin carcinoma.

Abdomino-pelvic MRI was done as part of the extension balance sheet and showed endometrial process hypointense T1, isosignal T2 with intense grip of Gadolinium, invading the myometer without signs of locoregional extension (figure 1).

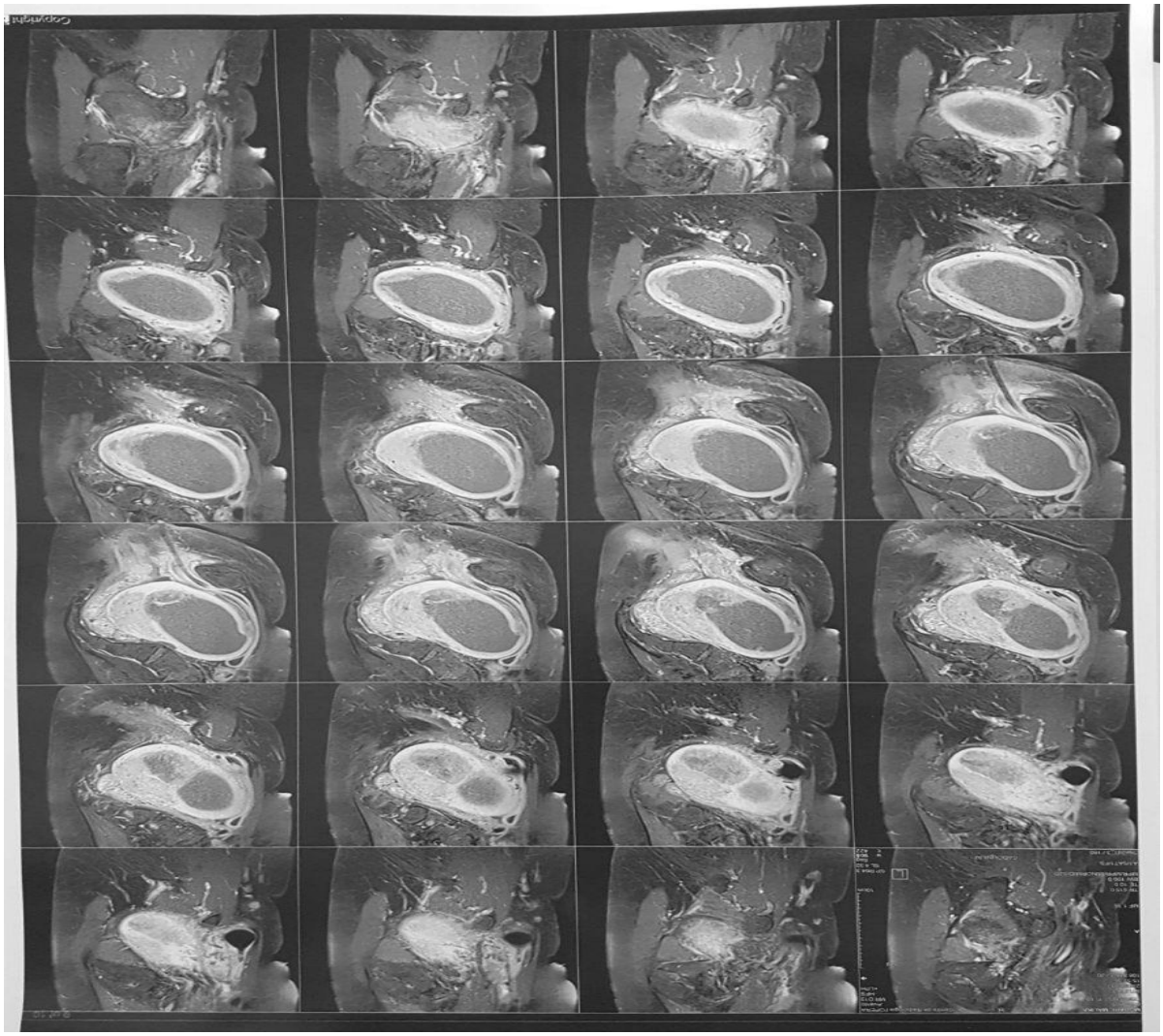

Figure 1: Abdomino-pelvic MRI in T1 ponderation showing an endometrial process with important hydrometria.

This tumor is classified stage IB corresponding to FIGO 2009 classification. Indication of enlarged colpohysterectomy with bilateral annexectomy and lymphadenectomy. Surgical exploration found an enlarged uterus with flooding of the serous (figure 2).

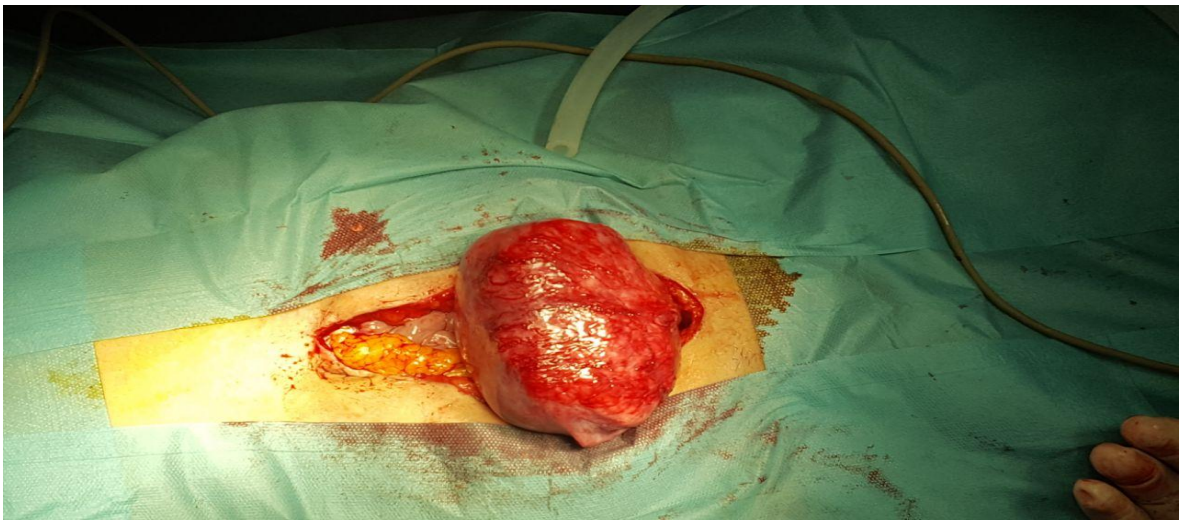

Figure 2: Macroscopical aspect of uterus showing flooding of the serous by endometrial process.

Postoperative period was normal and chemotherapy based on cisplatine-etoposide was started. The prognosis is bad with frequent recurrence due to the aggressiveness of these tumors.

DISCUSSION

neuroendocrin tumors represent 2% of all gynecological cancers with preferential localisation in cervix and ovaries.[1,2] Endometrial localisation is exceptional with bad prognosis. Contrary to what has been reported in our case, postmenopausal metrorrhagia is the main sign. Diagnosis can be established in theses cases: when we have pelvic mass, signs of compression of the pelvic organs or pelvic pain.

Pelvic ultra-sound found thickened endometrium, heterogenous, with irregularities between endometer and myometer. Endometer biopsy with histological and immunohistochimical study allows us to confirm diagnosis throw tumoral cells with neuroendocrin markers such as CD 56, chromogranin A and synaptophysine in endometrium.[1,3] The first question to ask is about localization if it's primary or secondary. Absence of pulmonary localization at radiological exploration, allows us to retain primary endometrial origin.

The extension balance sheet is based on pelvic MRI. Neuroendocrin tumors are typically heterogeneous, isosignal on T1-weighted sequences and in hypersignal on T2-weighted sequences. There are hyposignal bands in T2-weighted sequences at myometer corresponding to non-invasive myometer beams. PET scanner is interesting for detection of distant metastasis and for the progressive follow-up of the disease.

For treatment of these tumors, TNM classification is very useful, but actually it's necessary to add the grade established following to Ki67 or mitotic index. The more Ki67 is high, the more metastasis and risk of re-offending increases. 2010 classification of WHO distinguishes neuroendocrin tumors grade 1 (Ki67<2%), neuroendocrin tumors grade 2 (Ki67 between 3 and 20%) and neuroendocrin tumors grade 3 (Ki67>20%).[1,4]

The management of the cases described in the literature is based on surgery : colpohysterectomy enlarged with bilateral salpingo-ovariectomy, iliac and lombo-aortic lymph node dissection without omentectomy completed by chemotherapy based on etoposid and cisplatine or carboplatine.[1]

The prognosis remains pejorative with frequent recurrences.

Conflict of interest: None.

CONCLUSION

Nenroendocrin carcinoma of endometrium is an exceptional entity, only few cases have been reported in literature. There is no formal consensus on ownership of these tumors whose prognosis remains gloomy with frequent recurrences.

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