The Primary Squamous Cell Carcinoma of The Endometrium: A Case Report and Literature Review

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ABSTRACT

Introduction: The primary squamous cell carcinoma of the endometrium is a rare tumor. The incidence of these tumors is extremely low. Case Presentation: We report the case of an old patient of 67 years old, married with 5 children, 9 years since menopause. She consults for spontaneous postmenopausal bleeding with alteration of general state. The patient underwent hysteroscopy with biopsy endometrial curettage. Histology showed an adenocarcinoma endometroide grade 2. Then a total hysterectomy with bilateral adnexectomy and pelvic node dissection and multiple biopsies were realized. The anatomopathological examination showed a moderately differentiated non keratinizing squamous cell carcinoma without cervical location. The decision of the multidisciplinary consultation meetings was to complete the surgery by adjuvant radiotherapy. The patient presented a large retroperitoneal later aortic mass of 105mm and deceased one year later. Conclusion: The primary endometrial carcinoma, squamous cell (PESCC) is an extremely rare tumor and its pathogenesis remains unclear. There are no guidelines for how it should be treated: a first surgery with or without adjuvant therapy are offered.

Keywords: The Primary squamous cell carcinoma; the endometrium; surgery; prognosis

1. INTRODUCTION

The primary squamous cell carcinoma is an uncommon tumor of the endometrium which diagnosis is based on pathological examination. Its etiopathogenesis remains unclear. Its therapeutic care stays controversy with poor prognostic.

We report the case of a patient of 67 years old, and we discuss diagnostic, therapeutic and prognostic features.
2. CASE PRESENTATION

Mrs. BF, 67 years old, married and has 5 children, 9 years since menopause, with no notion of taking oral contraception or Hormone replacement therapy. She reported no personal or family history of endometrial, breast, colon (Lynch syndrome) or ovarian cancer.

The patient consults for spontaneous postmenopausal bleeding associated with hydrorrhea evolving for two years in context of deterioration of general condition with no other signs gynecological or extra gynecological including no associated urinary or gastrointestinal symptoms.

The general examination is a patient with World Health Organization (WHO) score is 1.

Gynecological examination finds atrophic vulva (given the age of the patient). The examination with speculum find reddish bleeding originating from endocervix and a macroscopically normal appearance cervix. On vaginal examination: uterus of normal size, no mass or latero-uterine tenderness.

Abdominal examination found a soft abdomen with no collateral venous circulation and no hepatosplenomegaly.

The examination of breasts, lymph nodes and the rest of examination are within limits.

Pelvic ultra sound: uterus with regular contours (103/70 millimeter), the presence of an intracavitary image of 56x70mm not vascularized with doppler and absence of intraperitoneal effusion (Figure 1).

Hysteroscopy diagnosis: Taking all the uterine cavity whitish evoking endometrial neoplasia, an endometrial biopsy and curettage were done. Histology showed an adenocarcinoma endometroide grade 2.

Resonance Imaging: Mass intra cavity (75x80x81mm) with heterogeneous necrotic tissue appearance andendometrial implantation. The mass is responsible of a total myometrial invasion without crossing the serosa. It skims the endocervix down without invading it and stays away from the vagina. (Figure 2 & 3)

Figure 2: MRI Mass intra cavity with heterogeneous necrotic tissue appearance and endometrial implantation.

Figure 3: MRI shows total myometrial invasion without crossing the serosa
The preoperative evaluation was unremarkable. The patient underwent a midline laparotomy exploring a uterus of normal size, the two appendages of the womb were unremarkable and noninfraventricular effusion. Performing a total hysterectomy with adnexectomy and pelvic node dissection and multiple biopsies and peritoneal cytology samples. Histological study showed macroscopically burgeoning whitish tumor occupying the entire uterine cavity and infiltrating less than 50% of the thickness of the myometrium. Under the microscope, the sample showed a tumor proliferation made of masses of large cells with net cytoplasmic boundaries and atypical nuclei with high numbers of mitoses. Dyskeratotic maturing signs are noted. No glandular tumor proliferation has been seen in all samples examined: It is an epidermal carcinoma of the endometrium (Figure 4: HES*20 shows a tumor proliferation made up of large cell masses of sharp cytoplasmic boundaries and atypical nuclei with numerous mitosis figures and abundant eosinophilic cytoplasm).

![Figure 4: HES*20 shows Large cell masses of sharp cytoplasmic boundaries](image)

The exocervix is covered by a regular squamous free of disruption or stigma of infection with Human papillomavirus infection (HPV). The endocervical glands includes regular devoid of atypia. The parameters and the isthmus are not invaded. The two ovaries and right and left fallopian tubes are not invaded. There is no neoplastic emboli. Pelvic lymph node (N) dissection bringing is negative (8N- / 8N right and 9N- / 9N left). Peritoneal cytology and biopsies show no tumor invasion. The multidisciplinary consultation meetings decision was completed by adjuvant radiotherapy. Patient received a total dose of 45Gy due to 1.8Gy / Fr in 25 sessions completed by 2 sessions of brachytherapy at the rate of 5Gy per sessions.

An MRI check performed 4 months after the end of treatment showed a voluminous retroperitoneal and latero aortic mass measuring 105x100 mm. Patient was lost to follow up then deceased one year later.

**3. DISCUSSION**

The primary squamous cell carcinoma of the endometrium is an extremely rare tumor, it has been reported less than 100 times in medical literature since its characterization in 1892. Fluhmann et al [1] highlighted in 1928, 3 diagnostic criteria of primary squamous cell carcinoma of the endometrium:
- Must not coexist with endometrial adenocarcinoma
- No connection between the endometrial tumor and squamous epithelium of the cervix

• Absence of any primitive epidermoid carcinoma of the cervix.

2 criteria have been added by the WHO in 1975: the presence of keratinization and intercellular bridges. Thus, for a positive diagnosis, careful scrutiny of the tumor is to exclude adenocarcinoma with squamous differentiation. Also, the entire cervix must be fully investigated to exclude an extension to the endometrium of a cervical carcinoma. The etiopathogenesis of early squamous cell carcinoma of the endometrium remains a matter of debate. Several assumptions were made:

  • Complete malignant squamous differentiation of endometrioid adenocarcinoma. [2]
  • Development from ectopic cervical tissue in the endometrium [3]
  • Involvement of HPV in pathogenesis, particularly type 16 [4]

Clinical and radiological aspects are not specific. In a literature review, a study led by Goodman et al. [5], reported 8 cases of patients suffering from a The primary endometrial carcinoma, squamous cell (PESCC) in a series of 1182 patients treated for endometrial cancer at the Hospital (Massachusetts), he also reported in his study 56 cases previously described in the literature. Based on this study several characteristics PESCC were highlighted.

The average age of women was 67 years (60 for endometrioidadenocarcinomas). 84% of patients were postmenopausal women.

Many predisposing factors to the development of PESCC have been proposed: chronic inflammation, radiotherapy, the deficit or excess estrogen [6].

In 64 cases, obesity and the use of estrogen, that are well-established risk factors for endometrial carcinoma in general, have not been identified as factors predisposing to PESCC. However, nulliparity, with 36% of frequency, may be associated.

The other preexisting condition in common with endometrial carcinoma was pyorrhea found in 31% of cases. The main clinical manifestations were post menopausal bleeding found in 44 patients (68%), vaginal discharge (28%), pelvic pain (17%), weight loss and pelvic mass sensation (6%). Distant metastases (1.5%) were observed in the urethral meatus, the vaginal orifice, the peritoneal space, the lung, liver, and brain [7].

Preoperative diagnosis of PESCC is often difficult, explaining in part lateness of patient cares by an average of 11.5 months. Only 50% of patients who underwent biopsy curettage before receiving definitive treatment had a preoperative diagnosis.

Although surgery is the main therapy for the PESCC optimal, currently available data are insufficient to assess the effectiveness of the type of surgery (hysterectomy with pelvic lymphadenectomy alone or associated with lumbo-aortic node dissection) and the association of radiotherapy and chemotherapy. Our patient underwent a total hysterectomy with bilateral adnexectomy and pelvic node dissection followed by external radiotherapy and brachytherapy with no chemotherapy. The prognostic factors are the same as other types of endometrial carcinoma.

All of stage I patients survived in addition to tumors limited to the uterus regardless of depth of myometrial invasion remains of good prognosis. However, the five patients with stage IV tumor, and most patients with stage III died despite adjuvant therapy. The importance of the grade has not been discussed in the literature.

The presence of vascular emboli is an important risk factor for recurrence in endometrial carcinomas [8], in the series of Goodman et al [5] all patients with vascular invasion died.In our case vascular emboli were absent.

4. CONCLUSION

The primary endometrial carcinoma, squamous cell (PESCC) is an extremely rare tumor and its pathogenesis remains unclear.

There are no guidelines for how it should be treated: a first surgery with or without adjuvant therapy are offered.

The prognosis seems depending on the stage of the tumor, the type of surgery and adjuvant treatment as well as the presence or absence of neoplastic emboli.

CONFLICT OF INTEREST STATEMENT

There is no conflict of interest between the authors.

COMPLIANCE WITH ETHICAL STANDARDS

Informed Consent was obtained from participant included in the study.
REFERENCES