Uterine tumor resembling ovarian sex cord tumor: we can diagnose it if we consider it. Case report and literature review

Călina Maier[1,2], Anca Potecă[2], Maria Olinca[1,2], Radu Vlădăreanu[1,3], Elvira Brătilă[1,2]

1. Department of Obstetrics-Gynecology and Neonatology, "Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania

2. "Prof. Dr. Panait Sirbu" Clinical Hospital of Obstetrics and Gynecology, Bucharest, Romania

3. Department of Obstetrics-Gynecology and Neonatology, "Elias" University Emergency Hospital, Bucharest, Romania

Corresponding author:
Călina Maier
E-mail: calinamaier@yahoo.com

Abstract

Uterine tumor resembling ovarian sex cord tumor (UTROSCT) represents a rare uterine neoplasm with uncertain malignant potential. Several reports have described recurrences, metastases and even deaths associated with these generally considered benign tumors, thus making them a controversial disease entity. UTROSCT has an unclear histogenesis and it possesses a polyphenotypic immunophenotype. We present a case of UTROSCT in a patient accusing abnormal genital bleeding and symptoms associated with anemia. After performing a total abdominal hysterectomy with bilateral salpingectomy, the final pathology report revealed UTROSCT with no sarcomatous features. The patient is still disease-free after 30 months of follow-up. We also present the data available in literature regarding this relative new type of uterine tumor. We are of the opinion that a high index of suspicion is of paramount importance when examining a patient with an uterine mass in order to promptly identify the risk factors that may predict agressive tumor behaviour.

Keywords: *UTROSCT, abnormal uterine bleeding, ultrasound, case report*

Rezumat

Tumora uterină asemănătoare tumorilor de cordoane sexuale ovariene (UTROSCT) reprezintă un tip rar de neoplasm uterin, având un potențial malign incert. Mai multe studii au raportat recurențe, metastaze și chiar decese asociate cu aceste tumori generic considerate benigne, ceea ce le încadrează totuși într-o tipologie controversată. UTROSCT prezintă un model de histogeneză încă neclar și un poliimunofenotip. În această lucrare, aducem în discuție cazul unei paciente care s-a prezentat în clinica noastră acuzând modificări ale ciclului menstrual și simptome asociate anemiei. În urma investigațiilor clinice și paraclinice, s-a decis practicarea histerectomiei totale cu salpingectomie bilaterală pentru o formațiune tumorală intracavitară, diagnosticul anatomopatologic fiind UTROSCT fără modificări sarcomatoase. Pacienta prezintă în continuare o evoluție favorabilă la 30 de luni postoperatoriu. De asemenea, am efectuat un review al literaturii de specialitate referitoare la acest tip relativ nou de tumoră. Opinăm că este deosebit de important un indice ridicat de suspiciune în cazul pacientelor diagnosticate cu tumori uterine pentru a putea identifica factorii de risc potențial predictivi pentru evoluția agresivă a acestora.

Cuvinte-cheie: *UTROSCT, tulburări de ciclu menstrual, ecografie, case report*

Submission date: 28.02.2022
Acceptance date: 10.03.2022

Tumoră uterină asemănătoare tumorilor de cordoane sexuale ovariene: o diagnosticăm dacă o cunoaștem. Prezentare de caz și review al literaturii

Suggested citation for this article: Maier C, Potecă A, Olinca M, Vlădăreanu R, Brătilă E. Uterine tumor resembling ovarian sex cord tumor: we can diagnose it if we consider it. Case report and literature review. Ginecologia.ro. 2022;35(1):26-28.

Introduction

Uterine tumor resembling ovarian sex cord tumor (UTROSCT) represents a rare distinct neoplasm included in the current World Health Organization (WHO) classification of endometrial stromat tumors[1] and its clinical characteristics are not fully understood. It was first described by Clement and Scully in 1976[2].

These neoplasms resemble morphologically sex cord tumors of the ovary and have a polyphenotypic immunophenotype. Their molecular pathogenesis has not been clearly described yet; however, tumors lack alterations found in other uterine tumors bearing sex cord-like differentiation, such as endometrial stromal sarcoma[3].

UTROSCTs usually occur in middle-aged women and behave as tumors of low malignant potential. The clinical characteristics are abnormal uterine bleeding, abdominal pain associated with an enlarged uterus or a palpable uterine mass without specific imaging aspects[4]. Most of the times, the diagnosis is incidental, following immunohistochemical and ultrastructural studies on the surgical specimen[5].

The treatment options include hysterectomy with or without bilateral salpingo-oophorectomy or even the hysteroscopic resection of the tumor[6]; yet, the management, prognosis, morbidity and mortality of this rare pathology are still under current debate.

In this paper, we present the case of a 32-year-old patient with abnormal uterine bleeding who was finally diagnosed with UTROSCT. Her treatment and follow-up after diagnosis are discussed and the data published regarding this entity are reviewed.

Case report

A 32-year-old gesta 3 para 1 Caucasian woman attended a local hospital accusing a six-month history of abnormal genital bleeding and symptoms associated with anemia. She had no prior history of allergy, comorbidities, use of medication or surgery, and there was no family history of gynecological cancer. The clinical gynecologic examination showed a large, bleeding mass protruding through the cervical os, and the initial pelvic ultrasound examination revealed an uterine hypoechogenic mass with vascularization at power Doppler (RI 0.67) with a diameter of 6/2.5 cm (Figure 1, bottom right). Interestingly, the endometrial sampling revealed a normal microscopic appearance. The patient was referred to our hospital for further investigation; after imaging and diagnostic workup and taking into account that the patient had completed the family planning, we decided to perform a total abdominal hysterectomy with bilateral salpingectomy. At gross pathologic examination, the intracavitary lesion appeared yellow, polycyclic, with a soft consistency and being poorly delineated with respect to the surrounding myometrium (Figures 1 and 2).

On microscopic examination, the tumor was composed of elements similar to the sex cord cells with a trabecular and alveolar arhitectural pattern and tubular elements. The cells showed an epithelioid appearance with irregular nuclei and abundant cytoplasm, fine chromatin and a high nucleoplasm ratio. These findings were consistent with UTROSCT. Immunohistochemical staining showed that the tumor cells were positive for Ki67, WT1, ER CD99, caldesmon and calretinin (Figure 1, above right). Both of the tubes were histologically normal.

At the regular checkups, the patient remained asymptomatic with normal pelvic examination and ultrasound findings up until the present time, at 30 months after surgery.

The patient has consented to the submission of the case report to the journal.

Discussion

To date, less than 100 cases of UTROSCT have been reported in literature[7]. The average age at diagnosis is 50.6 years old and the median age is 51 years old, while the tumor size is, on average, 47.6 mm[8], the latter being consistent with the one in our case report.

The clinical diagnosis of UTROSCT is challenging as the symptoms vary among patients and are not typical in

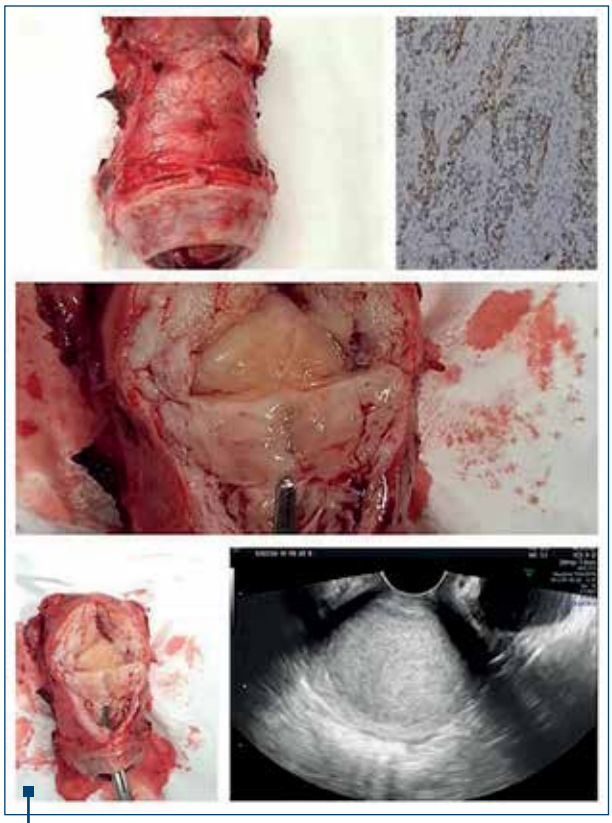

Figure 1. *Macroscopic intraoperative appearance of the uterus showing a yellowish intracavitary mass with soft consistency. Right above: immunohistochemical staining for caldesmon. Right below: Ultrasound appearance of the intrauterine mass*

Figure 2. *Intraoperative aspect of the tumor protruding through the distended cervical os*

most of the cases, therefore being misleading. The common symptoms consist in abnormal uterine bleeding, including postmenopausal bleeding (33.9%)[8] and pelvic pain. Dimitriadis et al. reported ectopic hyperprolactinaemia due to a malignant UTROSCT in a patient with a normal cranial MRI and in whom the prolactin rose further with dopamine agonist treatment[9].

The diagnosis can only be confirmed by histology after tumor resection, in most cases incidentally following hysterectomy performed due to an uterine mass (endometrial polyp or fibroid)[10]. Until now, there have not become available any noninvasive diagnostic tests such as specific serum markers or imaging findings. As a type of rare uterine tumor, UTROSCT can be diagnosed based on morphological and immunohistochemical conditions. Recently, UTROSCTs were found to be defined by recurrent fusions involving NCOA2 (nuclear receptor coactivator 2) or NCOA3, a finding that is directly amenable to diagnostic evaluation[3].

The standardized treatment for UTROSCT is lacking, due to its rarity, at present surgery being the preferred therapeutic method. It includes total hysterectomy with or without bilateral salpingo-oophorectomy or mass resection alone. Fertility sparing surgery should be offered to patients who wish to preserve their fertility; however, radical surgery must be considered after childbirth[1]. The youngest patient with UTROSCT reported so far is a 22-year-old nulligravida who underwent a hysteroscopic resection of an intrauterine mass presumed as grade I submucous myoma and was free of disease after 20 months, without any adjuvant therapy[6]. In our case, the patient opted for radical surgery as she completed childbearing.

Having an uncertain malignant potential and relapse, the patients diagnosed with UTROSCT should be followed-up closely. Tumor metastasis can occur to the ovary, omentum[11], lymph nodes and epiploic appendix[12], lungs[13] and vaginal vault[5]. In 2016, Endo et al. reported UTROSCT pelvic lymph node recurrence in a 62-year-old woman, 23 years after hysterectomy[14].

Moreover, there has been described the death of a 49-year-old woman from a metastatic UTROSCT initially considered as metastatic epithelial ovarian cancer[15].

Conclusions

UTROSCT are generally considered benign tumors with a favorable progosis, but they can easily relapse through incomplete resection. Surgery remains the primary management therapy and fertility preservation in patients of childbearing age is a vital consideration. It is crucial for pathologists to recognize this rare entity, as the clinical findings are nonspecific. Close follow-up is required for all patients due to the lack of prognostic biomarkers. ■

Conflict of interests: The authors declare no conflict of interests.

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