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Case report

Uterine Tumors Resembling Ovarian Sex Cord Tumors – Treatment, recurrence, pregnancy and brief review

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ABSTRACT

Background: Uterine Tumors Resembling Ovarian Sex Cord Tumors (UTROSCT) are rare tumors of low malignancy. In the past, these tumors were mainly treated by hysterectomy. More recently, some authors have proposed conservative surgical management for women wishing to preserve fertility. This article is the first to report on organ-preserving treatment in the case of recurrence or disease persistence.
Cases: We report on three patients with UTROSCT, two of them young, not having completed family planning. One even gave birth to a healthy child after fertility-preserving treatment of a persistent UTROSCT. To our knowledge, this is the first pregnancy reported after surgical treatment of a persistent UTROSCT so far.
Conclusion: A fertility-sparing approach should always be considered in young women with UTROSCT who wish to preserve their fertility, also in cases of recurrence or disease persistence.

1. Introduction

The occurrence of Uterine Tumors Resembling Ovarian Sex Cord Tumors (UTROSCT) was first described in 1945 (Morehead and Bowman, 1945). In 1976, a series of 14 cases was added (Clement and Scully, 1976). To date, less than 100 cases of UTROSCT have been published (Morehead and Bowman, 1945; Clement and Scully, 1976; O'Meara et al., 2009; Blake et al., 2014; Jeong et al., 2015; De Franciscis et al., 2016; Berretta et al., 2009; Giordano et al., 2010; Anastasakis et al., 2008; Hillard et al., 2004; Garuti et al., 2009; De Leval et al., 2010; Biermann et al., 2008; Gomes et al., 2015; Lantta et al., 1984). According to the WHO, UTROSCT are classified in the group of endometrial stromal and related tumors. The entity is defined as a "neoplasm resembling ovarian sex cord tumors without a component of recognizable endometrial stroma" (I.A.R.C. 2014, 4th Ed). The classification of UTROSCT found in literature is sometimes unspecified and a distinction between the more aggressive ESTSCLE (endometrial stromal tumors with sex cord-like elements) and UTROSCT is not made. However, this distinction is highly important because of the different behavior of these tumors.

UTROSCT are tumors of low malignant potential. They usually behave in a benign fashion; however, some may recur. The patients typically present with a bleeding disorder and/or a uterine mass.

These tumors are usually well-demarcated myometrial nodules with sharp or infiltrating borders. Some grow as polyps. They are smoother, fleshier and yellow to tan compared to leiomyoma. They may present different histological patterns such as trabecular, glandular, solid, diffuse or mixed. The cytoplasm can be scant or more abundant, often rich in lipid. The nuclei are small, inconspicuous and mitoses are very rare.

The immunohistochemical profile is variable. Using a marker panel (De Leval et al., 2010) is useful with sex cord markers (inhibin, calretinin, WT-1), one or more smooth muscle markers (desmin, h-caldesmon, smooth muscle actin), CD 10 and an epithelial marker (AE1/AE3 cytokeratin).

Due to potential recurrence and limited experience, in the past, UTROSCT were mainly treated by hysterectomy. More recently, conservative surgical management for women wishing to preserve fertility has been proposed (O'Meara et al., 2009; Blake et al., 2014; Jeong et al., 2015; De Franciscis et al., 2016; Berretta et al., 2009; Giordano et al., 2010; Anastasakis et al., 2008; Hillard et al., 2004; Garuti et al., 2009).

In this article, we report on three patients with UTROSCT, two of them young, not having completed family planning. One of them even gave birth to a healthy child after a second extensive fertility-sparing surgical treatment. To our knowledge, this is the first pregnancy reported under these conditions so far (Blake et al., 2014; Jeong et al., 2015; De Franciscis et al., 2016).

2. Case reports

The first patient, a 24-year-old woman, suffered from abnormal uterine bleeding (hypermenorrhea) and secondary dysmenorrhea.

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Ultrasound examination revealed a persistent submucosal mass resembling a leiomyoma in the fundal anterior wall of the uterine corpus. Over a two-year period, the size remained stable. Due to increasing symptoms a hysteroscopy (Fig. 1) with resection of the submucosal tumor was performed. The histological diagnosis was UTROSCT with expression of calretinin, one of the smooth muscle markers, WT1 and AE1/AE3 cytokeratin. It did not express inhibin. Three months later a fundal lesion of 8 mm was visible in a pelvic MRI. The patient strongly desired fertility-preserving treatment, so a repeat hysteroscopy with biopsies was performed, showing no histological evidence of UTROSCT. We agreed on regular clinical surveillance visits with imaging by ultrasound or additionally MRI. Six months later, a pelvic MRI suggested disease recurrence with a uterine mass of 15 mm. The patient strongly desired another fertility-preserving surgery. The diagnostic hysteroscopy showed no abnormalities. In the concurrent open abdominal surgery, the intramural tumor was located by palpation and completely resected. Pathological results were consistent with UTROSCT. The margins were free of tumor and the peritoneal lavage did not exhibit any tumor cells. The patient is under clinical as well as radiological surveillance since the last surgery and has now been disease free for 56 months.

The second patient, a 28-year-old woman, was referred to us after the operation of a symptomatic, slowly growing, cystic-solid tumor of 10 cm in the uterine anterior wall (Fig. 2) performed at another hospital. The tumor had been completely resected macroscopically through a lower abdominal incision. However, the surgery was complicated by strong bleeding and unintended opening of the tumor. The histological diagnosis was UTROSCT. Because of the possibility of recurrence, the hospital, where the initial surgery was performed, recommended a subsequent hysterectomy. As the patient did not want to undergo another surgery, regular clinical surveillance visits with additional imaging by MRI were performed. In the surveillance visit two months after surgery, the MRI indicated disease persistence with a tumor mass of $3 \times 4 \times 5$ cm in the anterior uterine wall (Fig. 3). As family planning was not completed, the patient did not wish to undergo a hysterectomy. She was referred to us for a second opinion and requested fertility-preserving surgery. There was no evidence of a macroscopic spread of the disease during open abdominal surgery. A round, yellowish mass the size of 3×3 cm (Fig. 4) was found on the anterior wall of the uterus. In contrast to the first case, the palpated texture was identical to the rest of the myometrium. The whole tumor was macro- and microscopically removed. The immunohistochemistry showed positivity for calretinin, one of the smooth muscle markers, WT1 and AE1/AE3 cytokeratin. It was negative for inhibin.

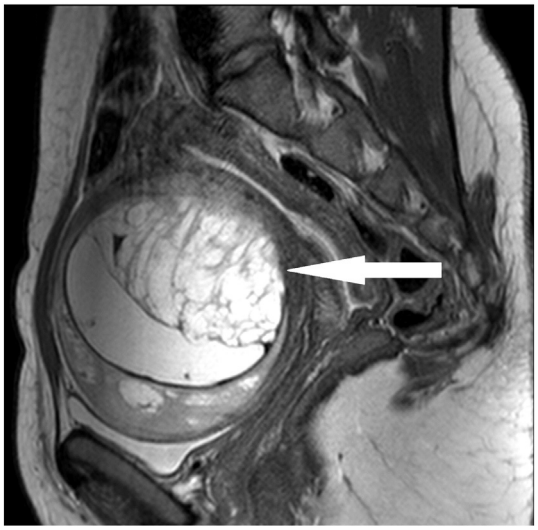

Fig. 2. Original tumor of the second patient in MRI.

The patient conceived without any problems and gave birth to a healthy child by cesarean section 19 months after the last surgery. Due to completed family planning, an abdominal hysterectomy with simultaneous removal of the distal part of the fallopian tubes on both sides was performed directly after the cesarean section. The entire tissue was free of tumor.

We agreed on the same procedure for oncological surveillance visits as in the first case. In one of the following surveillance visits, 20 months later, sonography revealed a 7 cm-large, polycystic tumor in the small pelvis. MRI indicated the same finding as well as strong activity of the contrast agent. Because of strong suspicion of recurrence, a third laparotomy was performed. The lower abdomen showed peritoneal carcinomatosis. The polycystic tumor originated from the right adnexa, infiltrating a part of the vaginal wall. The tumor, both adnexa, part of the vaginal wall as well as the affected peritoneum were removed.

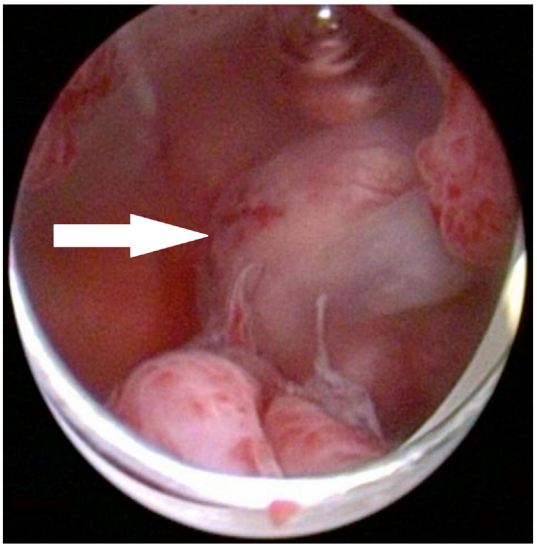

Fig. 1. UTROSCT of the first patient in hysteroscopy.

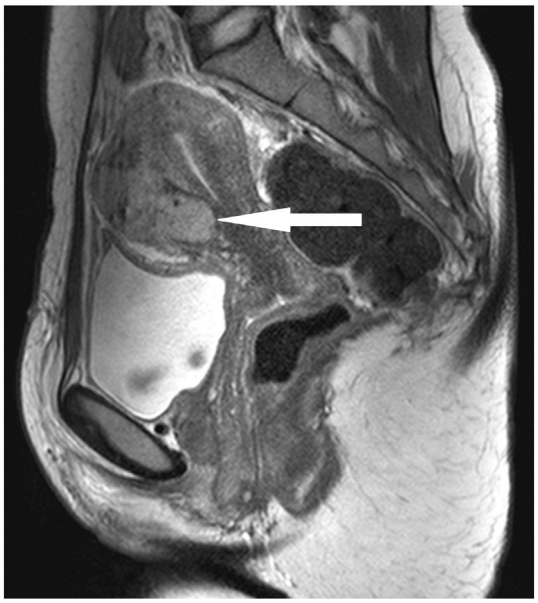

Fig. 3. Recurrence of UTROSCT in the second patient (MRI).