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Case Report

Uterine Tumors Resembling Ovarian Sex Cord Tumors (UTROSCT): A Case Report on a Young Woman with the Intention of Preserving Fertility

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Abstract

Background: Uterine tumors resembling ovarian sex cord tumors (UTROSCTs) belong to the miscellaneous mesenchymal category of uterine tumors. They typically present in middle-aged individuals, with occurrences in younger patients being rare. In the past, the primary treatment was hysterectomy, but recently, young patients, aiming to preserve fertility, have undergone fertility-preserving treatments such as tumor resection. This article presents a rare case of uterine UTROSCT in a 22-year-old female treated by local resection, under follow-up.

Case Report: A 22-year-old virgin female presented with a complaint of abnormal uterine bleeding (AUB) for the past 18 months. She underwent the initial surgical approach, which involved an incomplete hysteroscopic resection of the mass. Following this procedure, she was referred to our center after the final pathological report. Imaging revealed a uterine mass again. The patient underwent fertility-preserving surgery with laparotomy and resection of the mass, guided by hysteroscopy. Adjuvant therapy was not administered, and she has been under follow-up. Care without recurrence has been maintained for 10 months until now.

Discussion: We have performed searches using the keyword "UTROSCT" in databases including PubMed/MEDLINE, ScienceDirect, and Web of Science with a focus on English articles on under 40 years old patients published in the last decade. Among nine cases reported in scientific texts in patients under the age of 40 years, the median age was 33 years. AUB was the most common initial symptom. The tumor location was predominantly intrauterine in eight cases, including our patient, while in one case, the tumor was located in the cervix. The reported median tumor size in 6 cases was 4 centimeters, with our patient's tumor measuring 3.5 cm. In 5 cases, for whom immunohistochemistry (IHC) was performed, the calretinin marker was positive, consistent with our patient's case. Regarding treatment, hysterectomy with and without bilateral salpingo-oophorectomy (BSO) was performed in cases whose fertility preservation was not a concern. In our patients and cases with a focus on fertility preservation, tumor resection was performed. One reported case received adjuvant therapy after the initial surgical treatment, while our patient did not receive adjuvant treatment.

Conclusion: UTROSCT is a rare tumor, even more uncommon in reproductive age groups. Therefore, the outcomes of fertility-preserving treatments in young patients remain uncertain. Additionally, there is no definitive consensus on adjuvant therapy, and decisions should be made on a case-by-case basis. Further collection of a larger patient cohort and longer follow-up periods could provide valuable insights.

Keywords: UTROSCT, Young woman, Fertility sparing surgery

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Introduction

Uterine cancer takes the top position among gynecologic malignancies in countries with ample resources and ranks as the second most prevalent in those with limited resources. The primary histologic site and type of uterine cancer in such cases is adenocarcinoma of the endometrium, the lining of the uterus. Uterine sarcomas, which originate from the myometrium or the connective tissue elements of the endometrium, make up less than 10 percent of uterine corpus cancers (1,2).

According to the WHO classification, tumors of the uterine corpus are categorized into epithelial tumors (including endometrial carcinoma and all subtypes such as endometrioid, serous, and clear cell carcinomas) and mesenchymal tumors including Smooth muscle tumors, Endometrial stromal and related tumors, and Miscellaneous mesenchymal tumors. Uterine tumor resembling ovarian sex cord tumors (UTROSCT) falls under the subgroup of Miscellaneous mesenchymal tumors.

In 1945, Morehead and Bowman were the first to report uterine tumors containing an ovarian sex-cord component. However, it was not until 1976 that Clement and Scully provided a detailed description and categorized these tumors into two distinct groups (3).

Uterine tumor resembling ovarian sex cord tumors refer to uterine neoplasms that bear a resemblance to ovarian sex cord tumors but lack discernible endometrial stroma. The diagnosis of UTROSCT is established through histological examination, and the precise cell of origin remains unidentified. These tumors typically exhibit features akin to ovarian sex cord tumors and display positive staining for at least two markers commonly found in ovarian sex cord-stromal tumors. These markers include inhibin, calretinin, CD56, melan-A, CD99, FOXL2, or SF-1 (4).

Uterine tumor resembling ovarian sex cord tumor typically manifests in middle-aged women, primarily affecting those in the perimenopausal and postmenopausal age, with rare occurrences in individuals under 25 years of age. The common clinical presentation is AUB and/or abdominal pain, often accompanied by an enlarged uterus or a palpable uterine mass (5,6).

Here, we present a rare case of UTROSCT in a 22-year-old woman. The patient was treated with local resection and is still under surveillance. Additionally, a literature review of similar cases is included.

Case Report

A 22-year-old virgin patient presented with a complaint of AUB for the past 18 months. Initially, she had been under treatment with combined estrogen and progesterone pills, but due to the lack of improvement, she sought care at another center five months after discontinuing the medication and she underwent hysteroscopic resection of submucosal myoma. During the surgery, a 35×30 mm myoma with a thick and broad base in the posterior wall of the uterine body was observed, extending into the cavity, due to prolonged surgery, it had been impossible to complete the surgery. Pathology results reported UTROSCT, and the review confirmed endometrial mesenchyme neoplasm, likely UTROSCT.

The patient had no history of underlying diseases. Two months post-initial hysteroscopic surgery, an MRI revealed a uterus measuring approximately 69×22 mm. In the left corner of the uterus, at the site of the previous hysteroscopic resection, a lesion with dimensions of 23×22 mm was identified, demonstrating hyperdensity and relative restriction on DWI images (Figure 1).

According to the team decision including experienced radiologist and gynecologic oncologist, the uterine lesion was deemed suitable for resection.

Adjacent to this lesion, a myoma with a diameter of 8.5 mm was visualized. No pelvic adenopathy or other pathology was observed. The ovaries appeared normal on imaging. A normal chest CT scan was reported. Subsequently, the patient underwent laparotomy and tumor resection under hysteroscopic guidance. In the final pathology, the tumor's margin was free, and the immunohistochemical results were as follows: Calretinin: focally positive. CD99, Inhibin, WT1, AFP, CD10, HMB45: Negative. PR, Beta-catenin, H-Cal Desmon, SMA: Positive. PanCK: Dot-like positivity (Figure 2).

After the surgery with the diagnosis of UTROSCT, the patient was placed under follow-up until now without recurrence for 10 months.

Discussion

We have performed searches using the keyword "UTROSCT" in databases including PubMed/MEDLINE, ScienceDirect, and Web of Science. We focused on English language case reports published in the last decade involving individuals under 40 years of age, resulting in the identification of 9 relevant cases (Table 1). In the realm of medical terminology, UTROSCT represents a rare group of uterine neoplasms that are defined by their distinct morphological and immunohistochemical features. The differential diagnosis for UTROSCT is crucial, as it

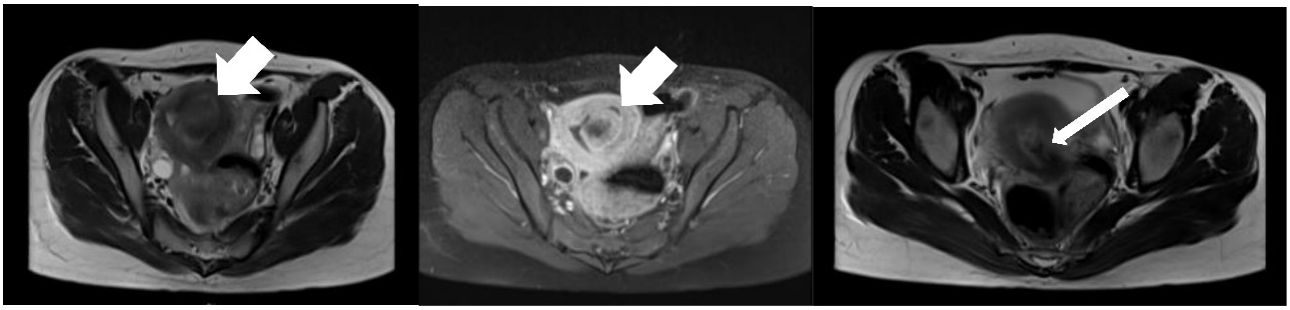

Figure 1. Axial T2 weighted (A&C) and T1 weighted post-contrast (B) MRI images: heterogeneous enhancing mass is bulging in the left side of the uterine cavity (thick arrow), suspicious vascular pedicle is shown in the distal wall of the uterus (thin arrow). Normal ovaries are seen (not shown). (from left to right: A, B, C)

Table 1. Clinical characteristics of nine young (below 40 years old) UTROSCT cases and comparison with the present case

Study	Age	Diameter (cm)	Presentation	Duration of symptoms (month)	Location of mass in the uterus	IHC[a]	Initial Surgical approach	Adjuvant Therapy[b]	Disease free F/U (months)
Amiri et al (Present case)	22	3.5	AUB	18	Submucosal	Calretinin	Laparotomy and hysteroscopic resection	No	6
Wang et al, 2022 (6)	19	3	AUB+pain	2	Cervical	Calretinin	Cervical lumpectomy	No	24
Schraag et al, 2017 (8)	28	10	-	-	Uterine	Calretinin	Local resection	No	2 Recurrence
Schraag et al, 2017 (8)	24	-	AUB+pain	-	Submucosal mass	Calretinin	Hysteroscopic mass resection	No	Recurrence
Watrowski et al, 2015 (9)	22	2	AUB	12	Submucosal	Calretinin, CD99, CD56, Melan-A	Hysteroscopic resection	No	28
Umeda et al, 2014 (10)	38	4.5	AUB	48	Sub mucosal	Unknown	TAH+BSO	Yes	11
Zhang et al, 2019 (11)	33	3.5	AUB	-	Uterine	CD99, Calretinin	TAH	No	12 years
Hermsen et al, 2015 (12)	36	-	AUB	-	Submucosal	Unknown	Local resection	No	24
Yang et al, 2016 (13)	35	10	AUB	3	Fundus	Unknown	Local resection followed by TAH+BSO	No	20
Yang et al, 2016 (13)	36	-	-	-	Polyp	Unknown	TAH+BSO	No	32

AUB, Abnormal uterine bleeding; IHC, immunohistochemistry.
[a] Positive immunohistochemical markers associated with uterine tumors resembling ovarian sex cord tumors (UTROSCTs) are mentioned here.
[b] Adjuvant therapy after initial treatment

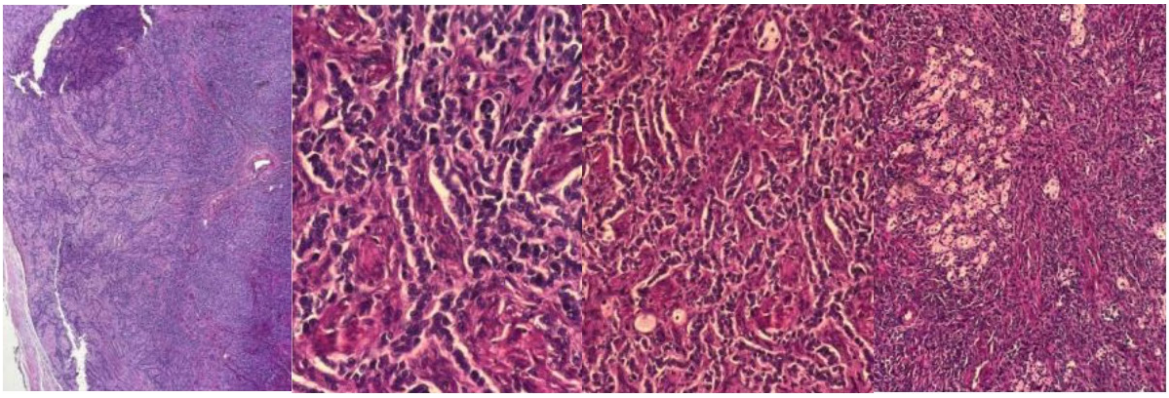

Figure 2. Pathology of UTROSCT: Hematoxylin & eosin stain, ×40(A), ×400 (B, C), ×100 (D): Cords and sheets of neoplastic cells in a fibrotic stroma, which are reminiscent of an adult granulosa cell tumor (from left to right: A, B, C, D)

involves distinguishing these tumors from other uterine pathologies. When considering such neoplasms, the primary differential diagnoses often include leiomyomas, endometrial polyps, and malignant uterine neoplasms including endometrial carcinoma and sarcoma (5). UTROSCT typically affects women in their mid-50s, often presenting with abnormal bleeding or pelvic pain (7). In some cases, the tumors are found by chance. It is more common in perimenopausal and postmenopausal women and is uncommon in individuals under 25 years old (6).

So, based on the assessment of 9 reported cases under 40 years old, the median age of the patients was 33 years, whereas our patient was 22 years old. The most common presenting symptom among the patients was AUB, which was also the case for our patient who presented with the same symptom. In all cases, the location of the mass was within the uterus, except for one case in which the mass presented in the cervix.

These tumors are considered to have a benign course, meaning they have a low likelihood of spreading or existing as a nodule or polyp. In terms of appearance, they tend to be smoother, fleshier, and have a yellow to tan color compared to leiomyomas (benign uterine tumors) (14,15).

Notably, in 6 out of the 9 cases under review, in which the size of the mass had been reported, the median size of the largest diameter was 4 centimeters. Our patient's mass size was 3.5 cm, aligning closely with the observed median size in these cases.

UTROSCTs can be presented with various histological patterns, such as trabecular, glandular, solid, diffuse, or mixed. The cytoplasm may be scant or more abundant, often containing a lot of lipids. The nuclei are small and mitoses are very rare. UTROSCTs typically resemble ovarian sex cord tumors and often show positive staining with markers commonly found in ovarian sex cord-stromal tumors, such as inhibin, calretinin, CD56, melan-A, CD99, FOXL2, or SF-1 (4).

It is remarkable that in 5 out of the 9 cases where immunohistochemical (IHC) staining was performed, the calretinin marker was found to be positive. Similarly, in the present case, calretinin was also positive upon testing (Table 1). In the present case, the immunohistochemical results were as follows: Calretinin showed focal positivity; CD99, Inhibin, WT1, AFP, CD10, and HMB45 were negative; PR, Beta-catenin, h-caldesmon, and SMA were positive; and PanCK exhibited dot-like positivity.

The management of UTROSCTs in young patients who wish to preserve fertility is challenging due to the rarity of the tumor and limited experience in this area. While conservative surgical management has been proposed as an option, there are limited guidelines for preserving the uterine corpus in these cases. Consensus guidelines for gamete or gonadal tissue conservation and trachelectomy exist, but recommendations specifically for the preservation of the uterine corpus in the context of UTROSCTs are lacking. The experiences with uterus preservation in young women with atypical hyperplasia or early low-grade endometrial cancer may not be directly applicable to uncommon tumors of non-endometrial origin like UTROSCTs. Overall, the rarity of UTROSCTs and the desire for fertility preservation present unique challenges in managing these tumors in young patients. More research and experience are needed to develop effective treatment strategies that balance the need for oncological control with the preservation of fertility and organ function (8,9).

Regarding the treatment, hysterectomy with and without BSO was performed in individuals among whom preserving fertility was not a concern. In cases with fertility concerns, similar to our patient, tumor resection was carried out.

The decision to pursue adjuvant therapy should be carefully considered and individualized based on the patient's specific circumstances and preferences. Close monitoring and counseling regarding potential long-term effects on fertility and pregnancy outcomes are crucial in the management of UTROSCTs in young patients. Further research is needed to guide the optimal management of these rare and complex tumors to achieve the best possible outcomes for patients (14,15).

One out of 10 presented cases received high-dose progesterone therapy for the treatment of an adjuvant (10). Instead, regular clinical follow-up with pelvic examinations and imaging as needed may be a more appropriate approach to monitor for recurrence. Additionally, ongoing research into potential biomarkers or genetic markers for UTROSCT may provide more targeted and effective surveillance strategies in the future.

Conclusion

In conclusion, the management of UTROSCTs requires a multidisciplinary approach that takes into account the unique characteristics of these rare tumors. Close collaboration between gynecologic oncologists, pathologists, radiologists, and other specialists is essential to ensure the best possible outcomes for patients. With ongoing research and advancements in treatment strategies, we hope to improve the prognosis and quality of life for individuals affected by UTROSCT.

Two out of the nine patients under 40 years old investigated experienced a relapse (Table 1). Our patient is still under observation after a 5-month follow-up. Two cases of relapse have been reported by Schraag et al (Table 1). In the first case, relapse was reported in a 28-year-old patient. The patient underwent laparotomy and tumor resection while preserving the uterus due to fertility desire. Two months later, based on the MRI follow-up, a suspicious mass measuring $3 \times 4 \times 5$ cm was detected in the anterior part of the uterus, leading to a second laparotomy and tumor resection. Nineteen months later, following a term pregnancy, the patient underwent cesarean hysterectomy and salpingectomy while preserving the ovaries. In the 20-month follow-up after the cesarean, ultrasound, and MRI detected a 7-centimeter mass suspicious for relapse. The third laparotomy revealed a carcinomatous peritoneum, suggesting that the tumor originated from the right adnexa. At this stage, bilateral BSO and peritoneal debulking were performed. Pathology confirmed UTROSCT in all three laparotomies. In the final surgery, both ovaries, vaginal wall, and peritoneum were involved. Chemotherapy with the bleomycin, etoposide, and cisplatin (BEP) regimen was recommended for three cycles, but the patient refused and opted for treatment with anastrozole. After 34 months of follow-up, the patient is alive and without relapse. The second case of relapse involved a 24-year-old patient who initially underwent local tumor resection. Nine months later, due to relapse, another tumor debulking was performed, and the patient was placed under surveillance (8). UTROSCT is a rare tumor, even more rare in the reproductive age group. Therefore, the outcomes of fertility preservation treatments in young patients are unclear. Additionally, there is no definite consensus on adjuvant treatment, and decisions need to be made on a case-by-case basis. Gathering more patients and longer follow-ups can be beneficial in gaining insights into the management of this rare condition.

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Authors' Contribution

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Formal analysis: Fatemeh Amiri.
Investigation: Fatemeh Amiri.

Methodology: Fatemeh Amiri.
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Resources: Fatemeh Amiri.
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Validation: Fatemeh Amiri.
Visualization: Fatemeh Amiri.
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Competing Interests
The authors declare that they do not have any conflict of interest.

Ethical Approval
Not applicable.

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