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Case Report

A Rare Case of UTROSCT Tumor in A Young Woman: Successful Pregnancy Followed by Definitive Surgical Management

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Abstract

Background: Uterine tumor resembling ovarian sex-cord tumor (UTROSCT) is an exceedingly rare uterine neoplasm. These tumors exhibit histological and immunohistochemical similarities to ovarian sex-cord tumors but lack endometrial stromal components. Given their rarity, diagnosis, treatment, and follow-up strategies remain challenging and are often tailored to individual needs.

Case Presentation: A case of a 36-year-old woman was reported who initially presented with menorrhagia and was diagnosed with UTROSCT following hysteroscopic myomectomy. Despite residual disease, the patient deferred definitive surgery to preserve fertility. She later conceived and delivered successfully but presented again with recurrent menorrhagia three years after initial diagnosis. Imaging confirmed recurrence, and the patient underwent total laparoscopic hysterectomy with bilateral salpingo-oophorectomy. Histopathology confirmed recurrent UTROSCT without lymphovascular invasion. No adjuvant therapy was required, and the patient remains disease-free on follow-up.

Conclusion: This case highlights the importance of accurate histopathological diagnosis, multidisciplinary planning, and individualized care in UTROSCT, particularly for women who desire fertility. Although UTROSCTs exhibit generally indolent behavior, they carry a risk of recurrence, warranting long-term follow-up even after completing surgical excision.

Keywords: bilateral salpingo-oophorectomy, case report, pregnancy, surgery, uterine tumor resembling ovarian sex-cord tumor.

Introduction

Uterine tumor resembling ovarian sex-cord tumor (UTROSCT) is an uncommon type of mesenchymal tumor in the uterus, making up less than 0.5% of all uterine malignancies and approximately 10–15% of mesenchymal uterine cancers [1]. Histologically, these tumors mimic ovarian sex-cord tumors but lack any identifiable endometrial stromal component. The first case resembling this tumor type was described in 1945 by Morehead and Bowman, who reported a uterine tumor, like an ovarian granulosa cell tumor [2]. In 1976, Clement and Scully further characterized this entity, identifying it as a uterine tumor with sex-cord differentiation, and categorized it into two subtypes based on morphology and clinical behavior [3]. The first subtype (Group I) includes endometrial stromal tumors with less than 50% sex-cord differentiation (ETSCLEs), often linked to recurrence and metastasis. The second subtype (Group II) is composed mostly or entirely of sex-cord-like elements and is defined as UTROSCT. In the current World Health Organization (WHO) classification, UTROSCT is listed under “Miscellaneous mesenchymal tumors.” These tumors are recognized for their resemblance to ovarian sex-cord tumors, but notably lack any distinguishable endometrial stromal tissue [4].

The exact origin of UTROSCTs remains unclear, though several hypotheses have been proposed. Some researchers suggest they

may develop from embryonically displaced ovarian sex-cord cells. Others propose a mesenchymal stem cell origin or consider the tumors to arise from an overgrowth of sex-cord-like elements within endometrial stromal tumors, adenosarcomas, or even areas affected by adenomyosis or endometriosis [5]. However, molecular analyses have shown that UTROSCTs do not exhibit typical genetic changes found in endometrial stromal tumors, such as JAZF1–JJAZ1 or PHF1 gene fusions [6]. Additionally, despite their morphological similarity to ovarian granulosa cell tumors and other sex-cord stromal tumors, UTROSCTs lack common mutations seen in those neoplasms, such as DICER1 and FOXL2 mutations [7, 8]. Interestingly, some cases of UTROSCT have been reported in patients using tamoxifen for breast cancer treatment, suggesting a possible link between the drug and tumor development [9, 10] Due to their rarity and unique histological appearance, UTROSCTs often present diagnostic challenges for pathologists. Here we report a rare case of UTROSCT, in a 36-year-old woman who was first treated conservatively and, after the relapse, with total laparoscopic hysterectomy with bilateral salpingo-oophorectomy.

Case Presentation

A 36-year-old unmarried woman presented to the gynaecology department in March 2021 with a chief complaint of menorrhagia. She had no significant past medical history. On physical examination, she was found to be clinically anaemic. Her vital signs were stable with a pulse rate of 82 beats per minute and blood pressure of 110/80 mmHg. Abdominal examination revealed a firm, mobile uterine mass, corresponding to approximately 14 weeks' gestational size. Transvaginal ultrasound demonstrated a large submucosal fibroid measuring 6 × 3.4 cm. Magnetic Resonance Imaging (MRI) further characterized the lesion as a submucosal fibroid measuring 6 × 5.8 × 5.3 cm.

The patient underwent hysteroscopic myomectomy on April 28, 2021. The excised specimen was sent for histopathological evaluation, which revealed a rare diagnosis, uterine tumor resembling ovarian sex-cord tumor (UTROSCT). Immunohistochemistry confirmed the diagnosis. A positron emission tomography-computed tomography (PET-CT) scan performed in June 2021 showed evidence of residual disease. Given the rarity and potential for recurrence, the case was presented at a multidisciplinary Tumor Board meeting. The board recommended total hysterectomy with bilateral salpingo-oophorectomy (TLH with BSO). However, as the patient was unmarried and had not completed her family, she declined the proposed surgical management.

The patient subsequently married and conceived in 2022. She delivered a healthy baby girl via lower segment caesarean section (LSCS) in September 2023. In April 2024, the patient re-presented with complaints of recurrent menorrhagia. Repeat imaging (ultrasound and MRI) revealed a recurrent submucosal fibroid measuring 6 × 5 cm. The case was revisited in the Tumor Board, and given recurrence, consensus was again to counsel the patient for definitive surgical management with TLH and BSO.

After thorough counselling, the patient consented to surgery and underwent a total laparoscopic hysterectomy with bilateral salpingo-oophorectomy on May 21, 2024. Postoperative histopathology confirmed recurrent UTROSCT, with no evidence of lymphovascular space invasion. Her postoperative recovery was uneventful. No adjuvant chemoradiation was deemed necessary. The patient continues to be under regular follow-up, and as of the latest evaluation, there is no evidence of metastasis or disease recurrence.

Discussion

This case highlights the importance of individualized, multidisciplinary management, especially in patients desiring fertility. Definitive surgery with total hysterectomy and bilateral salpingo-oophorectomy remains the treatment of choice for recurrent disease. Close surveillance is recommended given the unpredictable behavior of UTROSCT.

UTROSCT are rare uterine neoplasms characterized by a unique histological and immunohistochemical profile that is essential for diagnosis. Histologically, these tumors mimic ovarian sex-cord tumors, exhibiting corded, trabecular, and nested patterns of bland, uniform cells with varying cytoplasm and minimal atypia [11]. Immunohistochemically, UTROSCTs coexpress multiple lineage markers, with consistent positivity for sex cord markers such as calretinin, inhibin, CD99, and WT-1 [12]. They also express epithelial markers (cytokeratins), smooth muscle markers (desmin, h-caldesmon), and hormone receptors (estrogen and progesterone receptors). Molecular diagnosis shows recurrent gene fusions involving NCOA1-3 (e.g., ESR1-NCOA3, GREB1-NCOA2) provide further diagnostic confirmation and help distinguish UTROSCT from other uterine tumors, especially endometrial stromal tumors with sex cord-like differentiation (Dickson et al., 2019). Imaging findings, including MRI, are nonspecific and often show a well-defined uterine mass with signal characteristics overlapping leiomyomas, limiting their diagnostic utility [13]. Definitive diagnosis relies primarily on histopathology and immunohistochemical profiling supported by molecular testing when available

Clinically, UTROSCT typically presents with abnormal uterine bleeding or pelvic pain, though it may be asymptomatic in some cases. In our case, the patient reported menorrhagia for one year, which aligns with the most reported symptom across all age groups and menopausal statuses. In certain instances, the tumor is discovered incidentally during routine gynecological exams or infertility assessments [14]. Less commonly, hormonal disturbances may manifest as the initial presentation. Cases of

galactorrhea and hyperprolactinemia have been reported [15,16] as well as hypercalcemia attributed to ectopic prolactin or parathyroid hormone-related peptide (PTHrP) production [17]. Rarely, UTROSCT may present as a gynecologic emergency, with intra-abdominal bleeding due to tumor rupture leading to its diagnosis.

Our case is unique in demonstrating successful conception and full-term pregnancy following an initial diagnosis of UTROSCT, reinforcing the value of individualized care. Fertility-preserving approaches with delayed definitive surgery have been successfully employed in other cases [18]. A previous case was reported of a patient who underwent two organ-preserving surgeries before ultimately delivering a healthy child, followed by a hysterectomy [18]. Given the potential for late local recurrence, hysterectomy post-childbearing is considered prudent [18, 19]. In our case, recurrence prompted total abdominal hysterectomy with BSO, aligning with recommendations emphasizing complete tumor removal. Surgical excision remains the cornerstone of treatment, even for recurrent cases, with follow-ups extending beyond 30 years [20]. The recurrence rate of UTROSCT is estimated at approximately 6.3% [21]. Unfortunately, chemotherapy and hormonal therapy offer limited benefit. Regimens including ifosfamide, carboplatin, bleomycin + cisplatin+ etoposide, and CYVADIC have shown poor response rates [22, 23]. Hormonal agents such as letrozole, tamoxifen, and megestrol acetate also demonstrated minimal efficacy [21, 23].

Follow-up protocols vary significantly. While transvaginal ultrasound and MRI are effective in detecting recurrences, they cannot reliably differentiate UTROSCT from benign conditions. Some reports note elevated CA-125 [22, 24] or prolactin [15, 16] preceding recurrence. Repeat hysteroscopy has been proposed post-hystero scopic resection to ensure no residual tumor [14, 25]. Suggested follow-up schedules include biannual clinical and ultrasound evaluations, with annual hysteroscopy for 3–5 years [14, 25]. Due to the unpredictable recurrence timeline and reports of progression-free survival (PFS) ranging from 7 to 32 years [20, 22], long-term surveillance remains essential. This case reinforces the importance of comprehensive immune histo-chemical and molecular evaluation, individualized treatment planning, and prolonged follow-up in managing UTROSCT.

Conclusion

UTROSCT is a rare uterine neoplasm with distinct histopathological and immune histochemical features that pose diagnostic and therapeutic challenges. This case illustrates that fertility-sparing management may be feasible in selected patients, but definitive surgery remains the mainstay for recurrent disease. Given its unpredictable clinical course and recurrence potential, long-term surveillance is essential. Accurate diagnosis, multidisciplinary care, and individualized treatment planning are critical for optimizing outcomes in patients with UTROSCT.

Author Contributions

Conceptualization, methodology, validation, formal analysis, writing—review and editing. All authors have read and agreed to the published version of the manuscript.

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References

1. Giordano G, Guareschi D, Thai E (2024) Uterine Tumor Resembling Ovarian Sex-Cord Tumor (UTROSCT): A Rare Polyphenotypic Neoplasm. Diagnostics 14: 1271.
2. Morehead R P, Bowman M C (1945) Heterologous Mesodermal Tumors of the Uterus: Report of a Neoplasm Resembling a Granulosa Cell Tumor. Am J Pathol 21: 53-61.
3. Clement P B, Scully R E (1976) Uterine tumors resembling ovarian sex-cord tumors. A clinicopathologic analysis of fourteen cases. Am J Clin Pathol 66: 512-25.
4. LEE C, CHANG S (2020) WHO Classification of Tumours Editorial Board. Female Genital Tumours.
5. Staats P N, Garcia J J, Dias-Santagata D C, Kuhlmann G, Stubbs H, et al. (2009) Uterine tumors resembling ovarian sex cord tumors (UTROSCT) lack the JAZF1-JJAZ1 translocation frequently seen in endometrial stromal tumors. Am J Surg Pathol, 33: 1206-12.
6. Croce S, De Kock L, Boshari T, Hostein I, Velasco V, Foulkes W D, at al. (2016) Uterine Tumor Resembling Ovarian Sex Cord Tumor (UTROSCT) Commonly Exhibits Positivity With Sex Cord Markers FOXL2 and SF-1 but Lacks FOXL2 and DICER1 Mutations. Int J Gynecol Pathol, 35: 301-8.

7. chiang S, Staats P N, Senz J, Kommoss F, De Nictolis M, et al. (2015) FOXL2 mutation is absent in uterine tumors resembling ovarian sex cord tumors. Am J Surg Pathol 39: 618-23.

8. Segala D, Gobbo S, Pesci A, Martignoni G, Santoro A, et al. (2019) Tamoxifen related Uterine Tumor Resembling Ovarian Sex Cord Tumor (UTROSCT): A case report and literature review of this possible association. Pathol Res Pract 215: 1089-1092.

9. Gutierrez-Pecharroman A, Tirado-Zambrana P, Pascual A, Rubio-Marin D, García-Cosío M, (2014) A uterine tumor resembling ovarian sex cord tumor associated with tamoxifen treatment: a case report and literature review. Int J Gynecol Pathol 33: 151-5.

10. Dickson B C, Childs T J, Colgan T J, Sung Y S, Swanson D, et al. (2019) Uterine Tumor Resembling Ovarian Sex Cord Tumor: A Distinct Entity Characterized by Recurrent NCOA2/3 Gene Fusions. Am J Surg Pathol 43: 178-186.

11. Ye S, Wu J, Yao L, He J 2022. Clinicopathological characteristics and genetic variations of uterine tumours resembling ovarian sex cord tumours. Journal of Clinical Pathology 75: 776-781.

12. Cho Sh K H, Han Hy, Hwang It, Kim Jh, Lee S (2017) MRI Findings of Uterine Tumor Resembling Ovarian Sex-Cord Tumor: A Case Report. J Korean Soc Radiol 76: 298-302.

13. De Franciscis P, Grauso F, Ambrosio D, Torella M, Messalli E M (2016) Conservative Resectoscopic Surgery, Successful Delivery, and 60 Months of Follow-Up in a Patient with Endometrial Stromal Tumor with Sex-Cord-Like Differentiation. Case Rep Obstet Gynecol 57: 36865.

14. Dimitriadis G K, Wajman D S, Bidmead J, Diaz-Cano S J, Arshad S, et al. (2020) Ectopic hyperprolactinaemia due to a malignant uterine tumor resembling ovarian sex cord tumors (UTROCST). Pituitary 23: 641-647.

15. O'meara A C, Giger O T, Kurrer M, Schaer G (2009) Case report: Recurrence of a uterine tumor resembling ovarian sex-cord tumor. Gynecol Oncol 114: 140-2.

16. Suzuki C, Matsumoto T, Fukunaga M, Itoga T, Furugen Y, et al. (2002) Uterine tumors resembling ovarian sex-cord tumors producing parathyroid hormone-related protein of the uterine cervix. Pathol Int 52: 164-8.

17. Schraag S M, Caduff R, Dedes K J, Fink D, Schmidt A M (2017) Uterine Tumors Resembling Ovarian Sex Cord Tumors - Treatment, recurrence, pregnancy and brief review. Gynecol Oncol Rep 19: 53-56.

18. Hillard J B, Malpica A, Ramirez P T (2004) Conservative management of a uterine tumor resembling an ovarian sex cord-stromal tumor. Gynecol Oncol 92: 347-52.

19. Bennett J A, Lastra R R, Barroeta J E, Parilla M, Galbo F, et al. (2020) Uterine Tumor Resembling Ovarian Sex Cord Stromal Tumor (UTROSCT): A Series of 3 Cases With Extensive Rhabdoid Differentiation, Malignant Behavior, and ESR1-NCOA2 Fusions. Am J Surg Pathol 44: 1563-1572.

20. CÖMERT G K, KILIÇ Ç, ÇAVUŞOĞLU D, TÜRKMEN O, KARALOK A, et al. (2018) Recurrence in Uterine Tumors with Ovarian Sex-Cord Tumor Resemblance: A Case Report and Systematic Review. Turk Patoloji Derg 34: 225-233.

21. Blinman P, Tattersall M H (2009) A case of uterine tumour resembling ovarian sex cord tumour responding to second-line, single agent anastrazole. Intern Med J 39: 617-9.

22. Endo D, Todo Y, Okamoto K, Suzuki H (2016) A case of recurrent group II uterine tumor resembling ovarian sex-cord tumors, against which two hormonal agents were ineffective. Taiwan J Obstet Gynecol 55: 751-753.

23. Chen Z, Lan J, Chen Q, Lin D, Hong Y (2021) A novel case of uterine tumor resembling ovarian sex-cord tumor (UTROSCT) recurrent with GREB1-NCOA2 fusion. Int J Gynaecol Obstet 152: 266-268.

24. Kuznicki M L, Robertson S E, Hakam A, Shahzad M M (2017) Metastatic uterine tumor resembling ovarian sex cord tumor: A case report and review of the literature. Gynecol Oncol Rep 22: 64-68.

25. Garuti G, Gonfiantini C, Mirra M, Galli C, Luerti M (2009) Uterine tumor resembling ovarian sex cord tumors treated by resectoscopic surgery. J Minim Invasive Gynecol 16: 236-40.