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Case Report

Uterine Tumor Resembling Ovarian Sex-Cord Tumor – a Rare Gynecological Neoplasm

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Abstract

Uterine tumor resembling ovarian sex-cord tumor is a rare group of uterine neoplasms with unknown histogenesis and differentiation towards ovarian sex-cord elements. They are benign in nature with low malignancy potential. Diagnosis is based on immunohistochemistry and morphological features, and the distinction from other more malignant differentials is paramount to correctly individualizing treatment.

A 47-year-old patient was admitted to the hospital complaining of abdominal pain and abnormal uterine bleeding. Subsequent detection of multiple uterine fibroids by ultrasound study and CT scans lead to a robot-assisted total hysterectomy. Histological studies revealed that one of the fibroids in the myometrium had a morphology resembling an ovarian sex-cord tumor with low Ki-67 proliferative activity. After 12 months of follow-up, no additional treatment was required, and no signs of recurrence or progression were observed.

Uterine tumor resembling ovarian sex-cord tumor is a rare gynecological entity with no established treatment protocol. Differentiation between benign and malignant behavior is based on structural features and immunohistochemical expression assessment. This highlights the importance of immunohistochemical staining and morphological analysis to determine the degree of surgical radicality. Minimally invasive approach is feasible and safe, but more experience is needed for further conclusions.

Keywords

hysterectomy, immunohistochemistry, robot-assisted, UTROSCT

INTRODUCTION

The resembling ovarian sex-cord tumor is a rare type of uterine neoplasm. It was first described in 1945 by Morehead and Bowman with less than 100 cases reported in the literature.[1,2] They are mainly well-bounded myometrial nodules, with sharp or infiltrative borders, and various histological patterns – trabecular, glandular, solid, diffuse, or mixed. Patients usually present with uterine mass and/or uterine bleeding and their treatment plans includes hysterectomy with bilateral salpingo-oophorectomy or hysteroscopic resection of the tumor.[3] Uterine tumors resembling ovarian sex-cord tumors (UTROSCT) are almost entirely composed of sex-cord-like elements and are found to be benign, despite the possibility of malignancy and relapse. [4] They have polyphenotypic immunohistochemical expression and staining panel for sex cord (WT-1, calretinin, and inhibin), smooth muscle (h-caldesmon, desmin, and

smooth muscle actin), epithelial markers (AE1 and AE3 cytokeratin) and CD10 can be used to confirm and distinguish UTROSCT from other uterine neoplasms.[5]

CASE REPORT

A 47-year-old patient, with a BMI of 36.33 kg/m^2 and five previous pregnancies, was admitted to the hospital with a history of abdominal pain, vaginal bleeding, and secondary anemia. The ultrasound examination revealed a myomatous uterus, and the computer tomography scan showed three fibroids: one on the right with a size of 38×35 mm, another on the left with a size of 47×39 mm, and still another located at the transition between the uterine body and cervix with a diameter of 23 mm ***(Fig. 1).*** *Based on these findings and the patient's statement of completed family planning, a total robot-assisted hysterectomy without the adnexa was performed to overcome the patient's severe obesity and the related perioperative and postoperative complications. The total operative time was 70 min, with an estimated blood loss of 100 ml. There were no complications in the postoperative period and the patient was discharged after four days.*

Macroscopically, the specimen did not present any abnormal features. The endometrium was thin, and no mass was seen in the uterine cavity. On section, the fibroids had a solid structure and pale greyish color without signs of secondary changes.

On microscopic examination, hematoxylin and eosin-stained slides were analyzed. The histopathological report revealed in one of the fibroids findings of a sex-cord-like growth pattern, composed of sheets, nests, cords, and solid tubules. Tumor cells formed cords and trabeculae with small neoplastic cells, round with monotonous nuclei and inconspicuous nucleoli ***(Figs 2, 3)****. The immunohistochemistry was Ki-67 positive with low proliferative activity* ***(Fig. 4)****. After 12 months of follow-up, there are no clinical signs of recurrence or disease progression.*

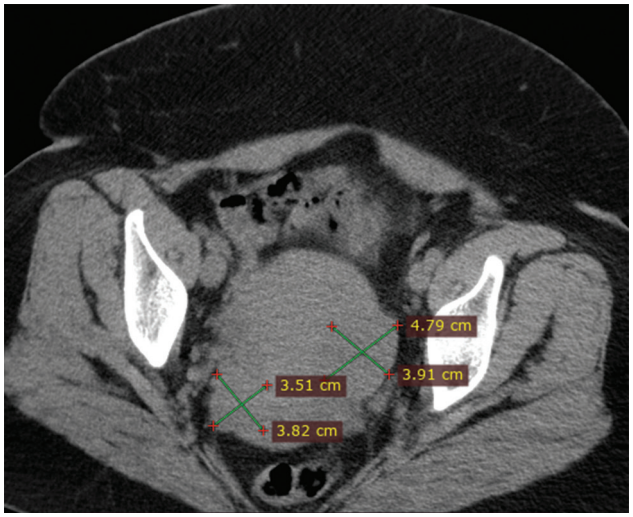

Figure 1. Pelvic computer tomography scan: Uterus myomatosum with two fibroids, located on both sides with sizes 38×35 mm and 47×39 mm.

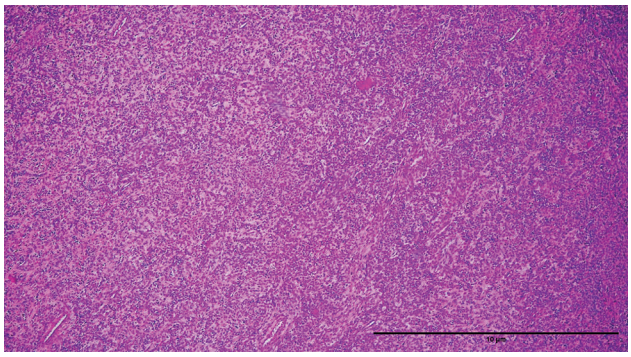

Figure 2. H&E stain ×4: UTROSCT – tumor composed of sheets, nests, cords and solid tubules.

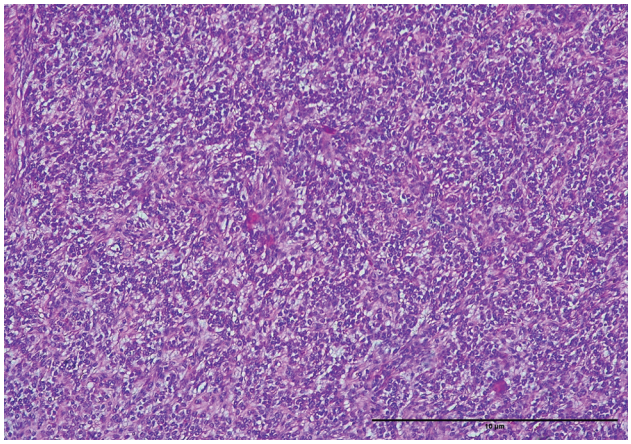

Figure 3. H&E stain ×10: UTROSCT – tumor cells form cords and trabeculae. Neoplastic cells are round and small, with monotonous nuclei and inconspicuous nucleoli.

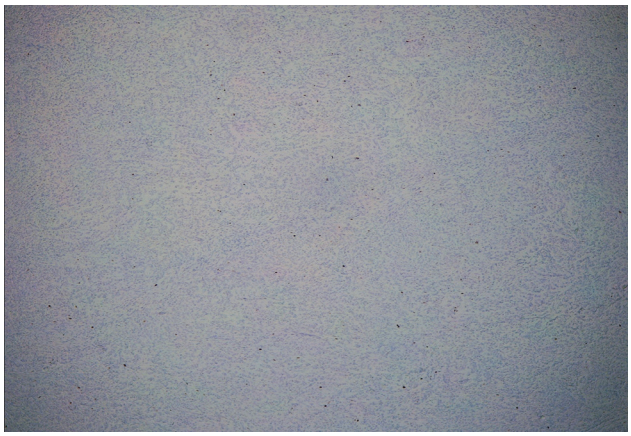

Figure 4. Immunohistochemistry: Ki-67 positive with low proliferative activity.

DISCUSSION

Uterine tumor resembling ovarian sex-cord tumors are rare, indolent neoplasms. The etiology is still unknown de-

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spite the emersion of different theories – derivation from uncommitted mesenchymal stem cells or from displaced ovarian sex-cord cells during embryogenesis.[6] The average age at diagnosis is 52 years as the tumor is seen in both premenopausal and postmenopausal women.[7] The clinical symptoms presented are typically abdominal pain and uterine bleeding with the tumor being submucosal, intramural, or subserous.[8]

In 1976, Clemente and Scully, based on histopathological and clinical features of 14 cases of uterine tumors with sex cord differentiation, classified them into two types.[9] The first, endometrial stromal tumors with sex-cord-like elements (ESTSCLE), are traditional endometrial stromal sarcomas with focal epithelioid formations that resemble sex-cord-like elements of an ovarian tumor, comprising less than 50% of the overall tumor mass. In 15% of the cases, they show a malignant character and are associated with an increased risk of recurrence.[8] The second type, an uterine tumor resembling ovarian sex-cord tumors (UTROSCT), is composed almost entirely of elements simulating ovarian sex-cord neoplasms. They are well demarcated with occasional infiltration in between muscle fibers, causing them to appear in different microscopic architectures like sheets, tight nests, trabeculae, thin cords, hollow or solid tubules with varying amounts of hyalinized or fibrous stroma. Cells have low mitotic indexes and contain little amounts of foam or clear eosinophilic cytoplasm.[8] They are generally benign with rare local recurrence, but their metastatic character has been reported in a few cases with dissemination to the appendix and the pelvic lymph nodes.[10,11]

UTROSCT is usually detected incidentally after surgery for other benign lesions.[7] Diagnosis is based on morphologic features and immunophenotype marker expression, as there are no specific histopathological characteristics. UTROSCT are not detected by endometrial biopsy due to their frequent localization within the uterine wall.[11] Because of their uncertain behavior, it is paramount to distinguish the benign and indolent UTROSCT from endometrial stromal sarcoma, endometroid carcinoma, and soft tissue sarcoma, all of which require aggressive treatment. The immunohistochemical panel includes expression of sex cord (Wilms tumor-1, calretinin, inhibin, CD56, CD99, SF1, FOXL2, MelanA), smooth muscle (h-caldesmon, desmin, histone deacetylase-8, smooth muscle actin, smooth muscle myosin heavy chain) and epithelial markers (AE1 and AE3 cytokeratin, KL1, EMA, CAM5.2), progesterone or estrogen receptors.[5]

Endometrial stromal sarcoma has an infiltrative growth pattern. It shows a diffuse expression with CD 10 immunostaining and the lack of JAZF1/SUZ12 fusion (JAZF1-JJAZ1) and PFH1 gene rearrangements distinguish these neoplasms from UTROSCT.[5] Endometroid carcinoma can present with sex-cord differentiation, but often areas with the tumor's morphology are present and there is a diffuse expression with EMA and pancytokeratin and negativity with WT-1. Soft tissue sarcomas can appear in the uterine wall as extra skeletal myxoid chondrosarcoma and synovial sarcoma. The former can have similar histology to UTROSCT, but they will not present with sex cord markers and the latter typically shows spindle cell appearance and epithelioid differentiation. Synovial sarcomas also have a negative expression of sex cord markers and a positive expression of EMA, CK7, and Bcl2, which will help its differentiation from UTROSCT.[4]

There is no established treatment protocol or guidelines for this diverse group of neoplasms. The main approach is total hysterectomy with 96% disease-free survival rate for a five-year period.[12] Despite that UTROSCT's unpredictable nature is associated with recurrence and metastases to lymph nodes, peritoneum, ovaries, lung, liver, and bones.[13] A total of 11 cases of fertility-sparing surgery have been reported with a mean age of the patients of 28 years.[14] All women were nulliparous – four of them conceived spontaneously and one with IVF after minimally invasive tumor resection. Sato et al.[11] reviewed 10 cases of malignant UTROSCTs and found that patients who underwent hysterectomy alone had a higher rate of recurrence. Therefore, extended radical surgery, including bilateral adnexectomy, lymphadenectomy, and omentectomy, may reduce the rate of recurrence in the more aggressive cases.[11] Although no effective chemotherapy regimen has been defined, according to the literature, bleomycin, etoposide and cisplatin seem to be a choice for adjuvant treatment.[15] High-dose progesterone therapy may be effective for UTROSCTs which often exhibit diffuse positivity for hormone receptor expression.[10]

CONCLUSIONS

UTROSCT is a group of uterine neoplasms with unknown histogenesis, which can present with a wide array of architectural patterns and immunohistochemical expressions. The distinction between the two subtypes is fundamental for optimal treatment. Therapeutic options are based on the scarce available data in literature. Total hysterectomy remains the recommended treatment procedure, with fertility-sparing approach showing promising results in the absence of extrauterine spread of the disease. Although they have a benign clinical course, there are reported cases of recurrence, local and distant metastasis, suggesting the need for more radical surgical approach and long-term follow-up.

Minimally invasive surgery is feasible, but additional case reports and experience are required to establish treatment guidelines.

Authors' statement

The data underpinning the analysis reported in this paper are deposited at Harvard Dataverse, V1 at https://doi.org/10.7910/DVN/AKFENP

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Опухоль матки, подобная опухоли полового тяжа яичника – редкое гинекологическое новообразование

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Резюме

Опухоль матки, подобная опухоли полового тяжа яичника, представляет собой редкую группу новообразований матки с неизвестным гистогенезом и дифференцировкой в сторону элементов полового тяжа яичника. Они доброкачественные по своей природе с низким потенциалом злокачественности. Диагноз основывается на иммуногистохимии и морфологических особенностях, а отличие от других, более злокачественных различий имеет первостепенное значение для правильной индивидуализации лечения.

Больная 47 лет поступила в больницу с жалобами на боли в животе и аномальные маточные кровотечения. Последующее обнаружение множественной миомы матки с помощью ультразвукового исследования и компьютерной томографии приводит к роботизированной тотальной гистерэктомии. Гистологические исследования показали, что одна из мином миометрия имела морфологию, подобную опухоли полового тяжа яичника, с низкой пролиферативной активностью Ki-67. Через 12 месяцев наблюдения дополнительного лечения не потребовалось, признаков рецидива или прогрессирования не наблюдалось.

Опухоль матки, подобная опухоли полового тяжа яичника, является редкой гинекологической опухолью, для которой не существует установленного протокола лечения. Дифференциация доброкачественного и злокачественного поведения основана на структурных особенностях и оценке иммуногистохимической экспрессии. Это подчеркивает важность иммуногистохимического окрашивания и морфологического анализа для определения степени радикальности хирургического вмешательства. Минимально инвазивный подход осуществим и безопасен, но для дальнейших выводов необходимо больше опыта.

Ключевые слова

гистерэктомия, иммуногистохимия, робот-ассистированная, UTROSCT