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Case Report

# Uterine tumor resembling ovarian sex cord tumor: A case report

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**Keywords:**

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**ABSTRACT**

Uterine tumor resembling ovarian sex cord tumor is a rare uterine neoplasm of uncertain histogenesis, with the presence of ovarian sex cord-like elements. These tumors typically occur in perimenopausal and postmenopausal women and are considered to behave in an indolent fashion in most cases. However, recurrences and metastases have also been reported. We report a case of a 37-year-old female who presented with lower abdominal pain and abnormal uterine bleeding and underwent a hysterectomy following a clinical and radiological suspicion of a submucosal uterine fibroid. Macroscopically the tumor resembled a leiomyoma. However, the diagnosis of uterine tumor resembling ovarian sex cord tumor was made after histopathological examination and immunohistochemical studies. Clinicians and pathologists need to be aware of this diagnosis, given the rarity of this condition and the potential aggressive clinical course.

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## INTRODUCTION

Uterine tumor resembling ovarian sex cord tumor (UTROSCT) is a uterine neoplasm with morphological patterns resembling that of an ovarian sex cord tumor but without a component of recognizable endometrial stroma. UTROSCTs are rare and constitute <1% of uterine mesenchymal neoplasms. Although most UTROSCTs are benign, recurrences have been documented. No definite morphological features have been identified to be predictive of recurrence and metastasis. These are considered to have low malignant potential.[1-3] UTROSCTs are mostly surgically

removed with a preoperative clinical suspicion of uterine fibroid. Morphologically, however, these may be easily overlooked as cellular leiomyoma and also endometrial stromal sarcomas with sex cord-like elements.

Here, we present a case of a 37-years female who underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy with a preoperative diagnosis of uterine fibroid, and upon complete histopathological evaluation was diagnosed with UTROSCT.

## # CASE REPORT

A 37-year female presented with a chief complaint of lower abdominal pain for the past 4 years. The pain was gradual in onset, dull in nature, intermittent, non-radiating, and relieved by taking over-the-counter analgesics. However, for the past 4 months, she had been experiencing acute episodes of left lower abdominal pain of cramping nature. In addition to the aggravated lower abdominal pain over the past 4 months, she also experienced urinary symptoms such as dysuria, burning micturition, and increased frequency of micturition. Other systemic examinations were unremarkable. Her menstrual history was remarkable for menorrhagia and metrorrhagia for the last 4 years. She had been married for the past 19 years and had two children. Other personal history was unremarkable. Other physical examinations were within normal limits. Ultrasonography of the abdomen and pelvis revealed a well-defined cystic lesion in the anterior myometrium measuring 2.5x3.2 cm, with possible differential diagnoses of focal adenomyosis and cystic uterine fibroid. With a provisional clinical diagnosis of uterine fibroid, the patient underwent a total abdominal hysterectomy with bilateral salphingo-oophorectomy. The post-operative period was uneventful.

Macroscopically, the uterus showed a submucosal mass within the myometrium measuring 3.5x3.5x3cm, which was well-circumscribed, non-encapsulated, with solid, white, firm cut surface exhibiting a whorled pattern, reminiscent of leiomyoma. No hemorrhage or necrosis was identified.

Microscopic examination of the submucosal myometrial mass revealed proliferation of smooth muscle cells in fascicles, interspersed by irregular aggregates of round to ovoid cells in nests, trabecular arrangement, at few foci imparting tongue-like protrusions, tubular formations, cords and few areas exhibiting retiform pattern. Tumor cells exhibited only a mild degree of pleomorphism with vesicular chromatin, inconspicuous nucleoli, and scant cytoplasm. Mitotic figures amounted to 1/10 HPFs. No overt atypia, necrosis, or hemorrhage were seen. Intervening stroma showed a smooth muscle component. Apart from the tumor, sections from other parts of the specimen were unremarkable. Patchy cellular nests of round to ovoid cells, tongue-like protrusions and at places exhibiting retiform and tubular patterns raised two differentials, viz. Endometrial stromal sarcoma with sex cord-like elements and Uterine tumour resembling ovarian sex cord stromal tumour (UTROSCT) (fig. 1).

Further immunohistochemical evaluation was performed, which showed tumor cells to be positive for Cytokeratin, EMA, Inhibin, Calretinin, and ER, while they were negative for CD10 and PAX8. SMA highlighted intervening smooth muscles of myometrium (fig. 2). Overall immunohistochemical findings were supportive of UTROSCT.

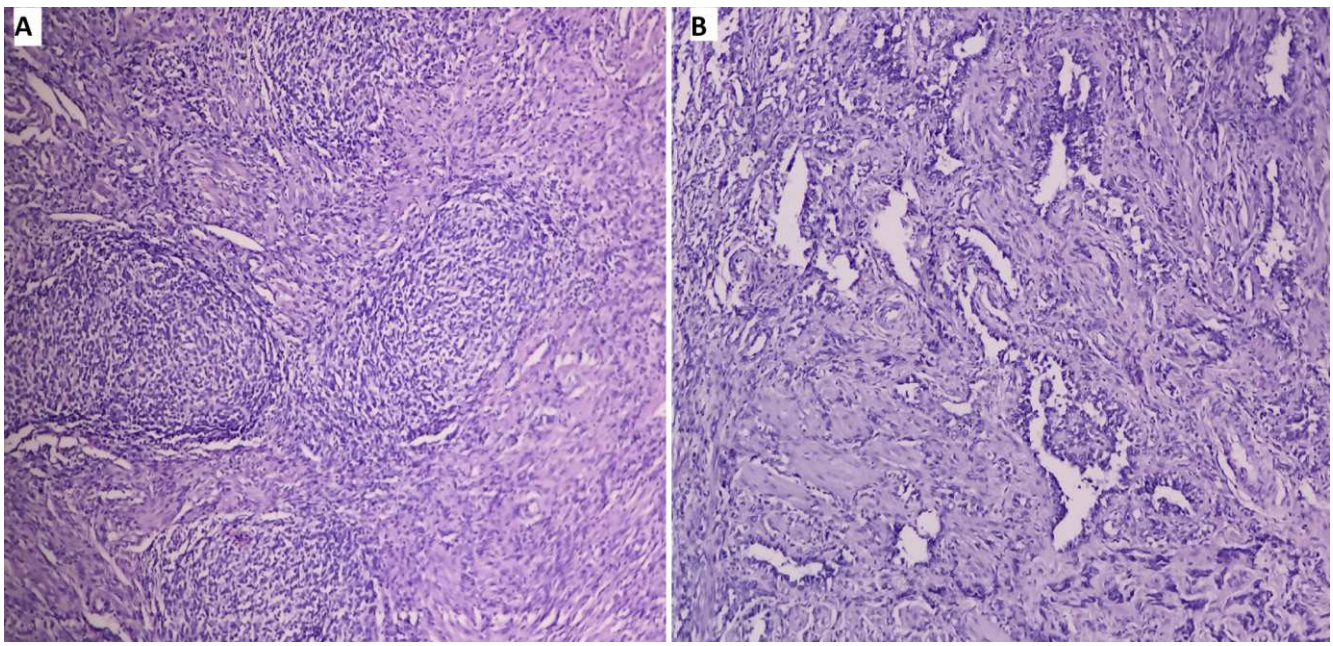


***Figure 1:*** *Representative photomicrograph of UTROSCT (A&B). Tumor cells seen in nests, cords and retiform pattern with intervening smooth muscle cells of myometrium. (HE stain, X10)*

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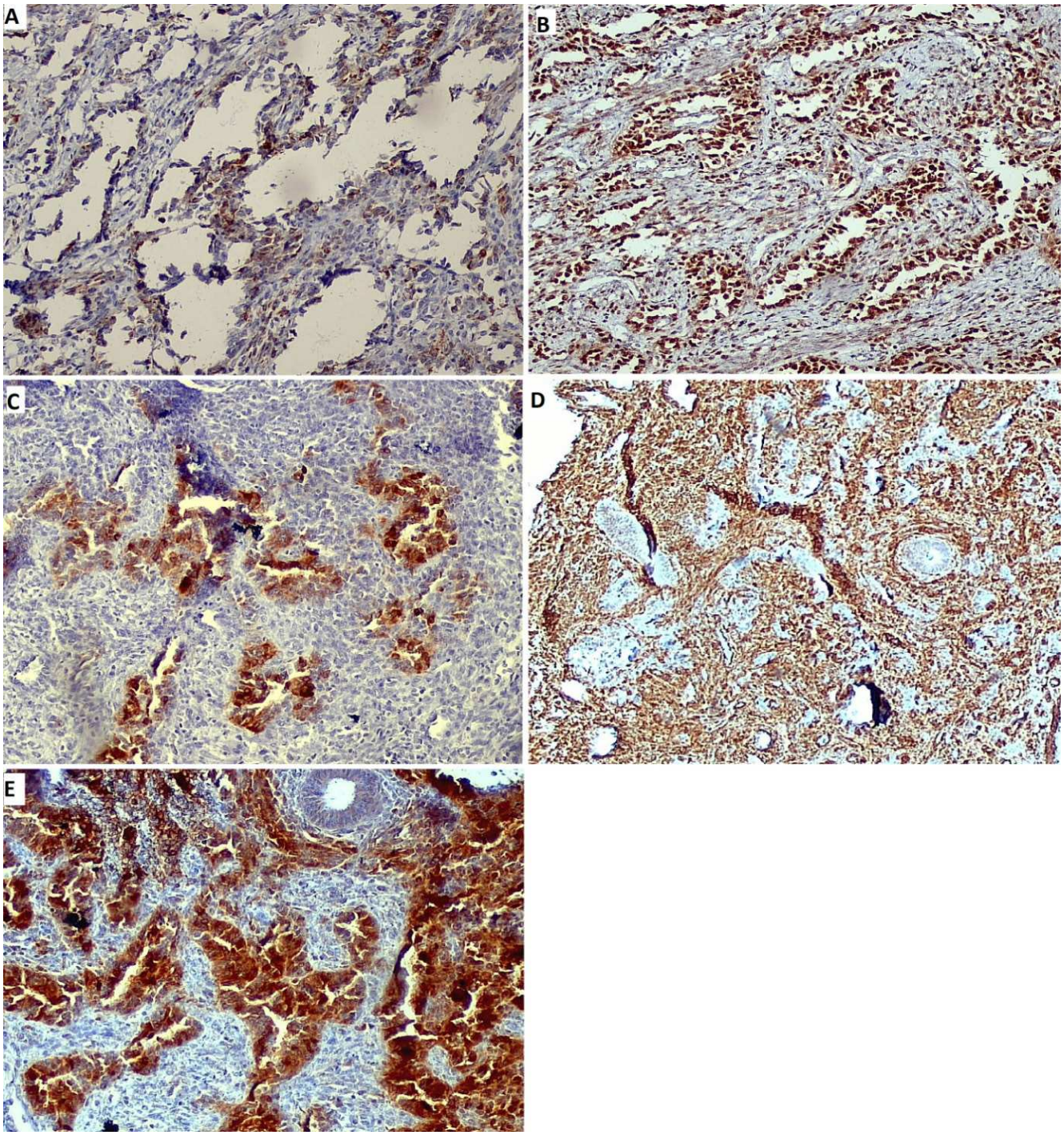


***Figure 2:*** *Immunohistochemistry. Tumor cells were positive for Cytokeratin (A), ER (B), Inhibin (C), Calretinin (E), and negative for SMA (D). SMA highlighted intervening smooth muscle cells (D).*

## DISCUSSION

UTROSCT are rare neoplasms of the female genital tract, accounting for <1% of uterine mesenchymal tumors. These typically occur in middle-aged women with a mean age of 50 years. Patients present with symptoms of abnormal vaginal bleeding, and pelvic pain, which was also observed in the present case.[1] Macroscopically, UTROSCTs are well-circumscribed, and may present as a polypoid mass located submucosally or intramurally with a yellow to tan cut surface.[1] In the present case, the tumor was located submucosally with a solid white homogenous cut surface resembling leiomyoma.

Clement et al. in 1976 described two variants of uterine mesenchymal tumors exhibiting sex cord-like elements, based on the percentage of ovarian sex cord-stromal tumor-like areas. First (type I) has 10-40% sex cord-like areas, which are commonly referred to as Endometrial stromal tumors with sex cord-like elements (ESTSCLE), while

UTROSCTs have exclusive/ predominance of sex cord-like areas.[4]

These sex cord-like elements can be identified by its characteristic tubule formations, plexiform cords, and retiform pattern with intervening smooth muscle of myometrium. The tumor cells in UTROSCTs tend to be uniform with small, bland nuclei and have rare mitotic figures. The tumor cells may be present in nests with apparent tongue-like extensions within the myometrium (Fig. 1A) and only sparse areas of tubule formations. This architecture and bland nuclear morphology may lead to an erroneous diagnosis of a cellular leiomyoma, epithelioid leiomyoma, or even low-grade endometrial stromal sarcoma (ESS) due to tongue like extensions. Immunohistochemistry is essential in morphologically ambiguous cases because of significant prognostic differences between these tumors. In the present case, the presence of tubule formations, and retiform areas clinched towards differentials of UTROSCTs and Low-grade ESS with sex cord-like differentiation. On immunohistochemistry, sex cord-like structures were immunoreactive for cytokeratin, inhibin, calretinin, and Estrogen receptor (ER), supporting their sex cord differentiation, which constituted approximately 70% of the tumor. CD10 and PAX-8 were negative, ruling out ESS and Mullerian adenocarcinoma respectively (fig. 2).

Various authors have reported the utility of Melan-A, CD56, WT-1, SF-1, and FOXL2 for confirming the sex cord differentiation with variable sensitivity and specificity.[5] Stewart et al. have noted high specificity and sensitivity of SF-1 and calretinin respectively for UTROSCT. They found SF-1 to be particularly helpful in differentiating UTROSCT from other histological mimics, as calretinin may be positive, albeit weak or focally, in other tumors as well.[6] Croce et al. observed 53% (10/19) and 58% (11/19) positivity rate for FOXL1 and SF1 respectively.[7]

Although UTROSCT is mostly benign, WHO recommends considering them as having a low malignant potential as recurrences and metastases have been reported. In a series of 34 cases published by Moore et al. in 2017, as high as 23.5% developed extrauterine metastasis and 8.8% died of the disease. In their study, older age, larger size, necrosis, lymphovascular involvement, cervical involvement, significant nuclear atypia, and mitotic activity were significant for increased malignant behavior of UTROSCT.[8] However, objective prognostic morphological parameters are yet to be determined.[1]

## CONCLUSION

The present case adds to the growing repository of uterine tumors with sex cord-like differentiations. UTROSCTs are characterized histologically by the presence of ovarian sex cord-like elements in varying amount, and express immunohistochemical markers for epithelial, stromal and sex cord elements. Because of varying morphological features, these may be miscontrued for endometrial stromal sarcoma, cellular leiomyoma, and even adenocarcinoma. The tumor follows a benign course in most of the cases, but few cases with recurrences and metastases have been reported. Hence, its identification is necessary and pathologists need to be well aware of this entity.

**Conflict of Interest:** None

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