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Case report

Extraovarian sex cord tumor with annular tubules discovered arising from a leiomyoma

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ABSTRACT

Background: Sex cord tumors with annular tubules (SCTAT) are a rare (2%) subtype of ovarian sex cord-stromal tumor. SCTATs are usually cured at time of diagnosis by surgical resection with an oophorectomy. SCTATs have a 100%(disease related) five-year survival. One third of SCTAT tumors are associated with Peutz-Jeghers syndrome. Literature review discovered only two published cases of extra-ovarian SCTAT. Due to the rarity there is no standard treatment for extraovarian SCTATs.

Case: A 39-year-old para-1 female with a symptomatic fibroid uterus, heavy menstrual bleeding, and a history of a uterine myomectomy, underwent an elective total abdominal hysterectomy. Intraoperative findings showed a 7.5 cm retroperitoneal mass adhered between the uterus and the right pelvic sidewall that on frozen section was found to be a degenerating leiomyoma. Final pathology demonstrated a 2 mm focus of incidental SCTAT adjacent to the serosal surface of the leiomyoma. The SCTAT was not associated with ectopic ovarian tissue or endometriosis. The patient's ovaries were normal on direct intraoperative examination, preoperative ultrasound and MRI. Six month postoperative surveillance ultrasound also demonstrated normal premenopausal ovaries.

Conclusion: This is the first extraovarian SCTAT in the published literature arising from a leiomyoma. Our patient had no family history and displayed no syndromic features for Peutz-Jeghers Syndrome. Ultimately, she declined genetic testing. The lack of evidence of ovarian involvement on both imaging and on intraoperative examination made localization to either ovary impossible. The patient is currently being managed with surveillance since the morbidity associated with bilateral oophorectomy in the 4th decade of life exceeds the theoretical risk of SCTAT.

1. Case

We present a case of an incidentally identified extraovarian sex cord tumor with annular tubules arising from a leiomyoma following a routine simple hysterectomy. A 39-year-old Gravida-1-para-1 female with a history of a fibroid uterus presented with progressively worsening pelvic pain and heavy menstrual bleeding. The patient underwent a uterine myomectomy two years prior for bleeding and mass symptoms. After one year, her pelvic mass symptoms gradually returned. Examination revealed a 14 week sized, wide based, globular uterus deviated to the patient's left. Preoperative imaging demonstrated an 11.2 × 4.2 × 5 cm fibroid uterus, normal follicular ovaries (Fig. 1), and a 7 cm right lower pelvic mass, suggestive of a pedunculated fibroid within the broad ligament deviating the uterus (Fig. 2). The patient had up-to-date cervical cancer screening. Her menstrual cycle was regular, 7 days long, but heavy and required pad changes every 3 h on the heaviest days. The patient's mass symptoms were not improved by conservative treatment with a levonorgestrel intrauterine device or a gonadotropin-releasing hormone agonist. The patient had satisfied parity and elected for definitive surgical management via a hysterectomy. She had no personal or family history of cancer.

The patient underwent a total abdominal hysterectomy, bilateral salpingectomy, and cystoscopy. Intraoperatively, a large, boggy, 7.5 cm retroperitoneal mass was identified, adhered to the uterus on the right and extending to the lateral pelvic sidewall. The mass was carefully dissected and removed from the uterus intact. Given the size and atypical location of the mass, it was sent for intraoperative frozen section. Frozen intraoperative consultation returned as an indeterminate, but likely benign smooth muscle tumor with edema and degenerative changes. No evidence of mitotic activity or atypia was identified. The

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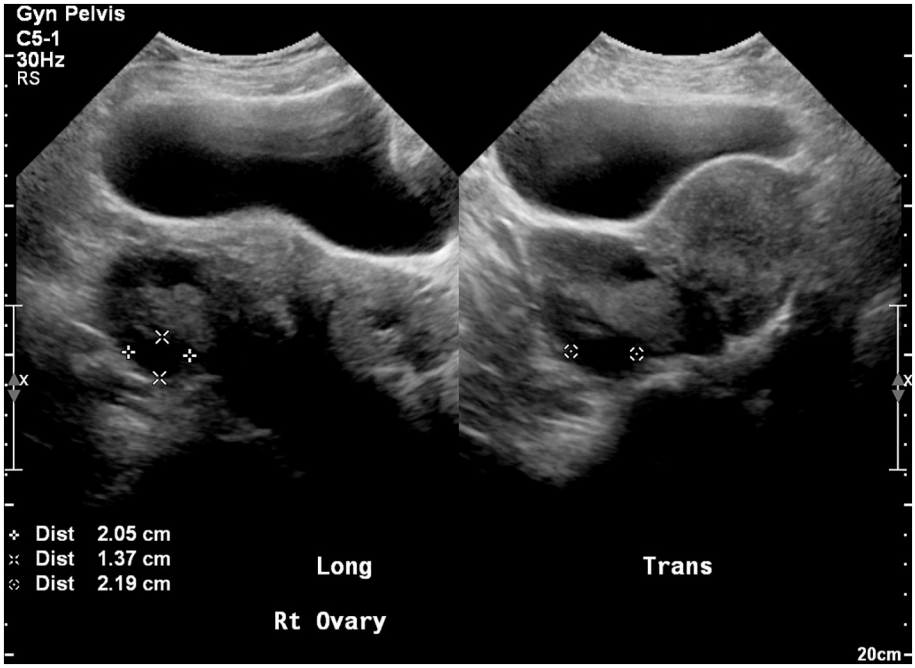

Fig. 1. Preoperative Pelvic Ultrasound Right Ovary.

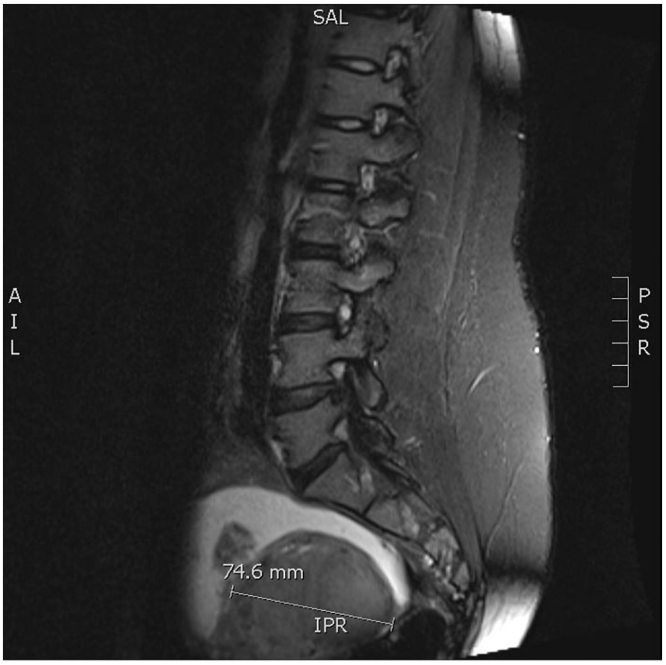

Fig. 2. Preoperative MRI Pelvis, Right Fibroid.

ovaries were normal appearing bilaterally, and no other pelvic pathology was noted on abdominal survey.

The final pathology for the sidewall mass returned as a large hydropic leiomyoma with a small focus of incidental sex cord tumor with annular tubules. The tumor measured at least 2 mm and was located adjacent to the serosal surface, on the stalk of the leiomyoma. The tumor displayed simple and complex tubules with palisading of cells around the basement membrane, with dense central hyaline material (Fig. 3). The immunohistochemical staining pattern was characteristic of SCTATs, including diffuse positivity for inhibin (Fig. 4). There was no ovarian or endometrial tissue associated with the tumor. The remainder of the uterine, cervical, and tubal pathology was benign. The specimens

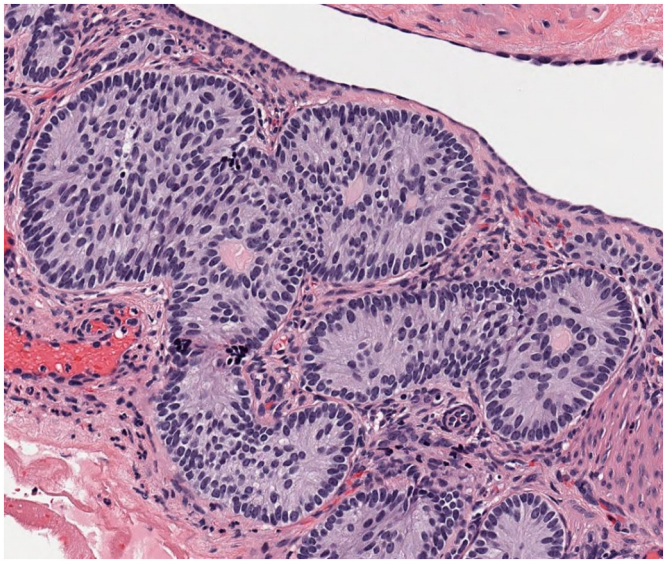

Fig. 3. SCTAT H&E Stain 200×.

were sent for expert review at a regional referral center. Their pathologists concurred with the rare finding of extraovarian SCTAT arising from a leiomyoma.

Tumor presence on the ovaries and adnexa was excluded by intraoperative inspection and review of preoperative imaging. Given the patient's young age and remoteness from menopause, our recommendation was for repeat imaging in six months, rather than proceeding with a bilateral oophorectomy to rule out further disease. Due to the association of SCTAT with Peutz-Jeghers syndrome(PJS), the patient was also referred to a genetic counselor. She did not display any syndromic features and ultimately declined genetic testing. On her ultrasound six months later, her adnexa remained normal in appearance and there was no evidence of tumor.