CASE REPORT

Inflammatory Myofibroblastic Tumor of the Uterus: An Under-recognized Entity—A Case Report and Literature Review

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Abstract

Aim: We report an uncommon case of uterine inflammatory myofibroblastic tumor (IMT), a unique mesenchymal tumor often misdiagnosed as "fibroid uterus" on radiology.

Background: Inflammatory myofibroblastic tumor is a mesenchymal tumor reported in the lungs, mesentery, omentum, and retroperitoneum. Its occurrence in the uterus is rare and is often misidentified as a smooth muscle or endometrial stromal tumor. Herein, we report a case of IMT of the uterus.

Case description: A 33-year-old lady presented to the OP with menorrhagia. USG abdomen was reported as bulky uterus with an intra-mural (8.7 × 7 cm) fibroid arising from the anterior myometrium. No lymph nodes or free fluid noted. "Myomectomy" was done under spinal anesthesia, and the specimen was sent for HPE. Grossly, it was a circumscribed gray–white soft-tissue mass mimicking a leiomyoma. However, microscopy of the mass was rather unusual to call it a leiomyoma. It had a "tissue-culture-like" or "nodular fasciitis-like" appearance. There were spindle cells in fascicles and prominent myxoid change with lymphoplasmacytic infiltrate. No nuclear atypia, necrosis, or infiltrative margins were noted. So, a morphological diagnosis of IMT was made.

Conclusion: The case is reported for its diagnostic challenge and rarity.

Clinical significance: Inflammatory myofibroblastic tumors are uncommon mesenchymal tumors of low malignant potential with a tendency to recur and rare metastases. Surgical excision is the treatment of choice. Recurrent IMTs can be treated with ALK-based targeted therapy.

Keywords: Inflammatory myofibroblastic tumor, Mesenchymal tumor uterus, Uterus.

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Background

Inflammatory myofibroblastic tumor of the uterus is a rare mesenchymal neoplasm of uncertain malignant potential with a small subset associated with recurrence and metastases. These tumors are often under-recognized in the female genital tract as their morphological features overlap with commoner smooth muscle or stromal tumors.[1–4] We report a case of inflammatory myofibroblastic tumor of uterus, which was clinico-radiologically reported as leiomyoma.

Case Description

A 33-year-old lady (P1L1A3 with history of previous LSCS) presented with the heavy flow during menstruation. P/A – uterus was 16 weeks in size and P/V – cervix ↓ uterus anteverted mobile 16 weeks FF. USG abdomen was reported as bulky uterus with a large intramural fibroid measuring 8.7 × 7 cm probably arising from anterior myometrium; unremarkable adnexa and no lymph nodes or evidence of ascites were noted; both iliac fossae looked normal. The patient was admitted and a myomectomy was performed under spinal anesthesia. Through an anterior incision, the myoma was enucleated and sent for HPE.

Grossly, the "myoma" was a well-circumscribed soft-tissue mass measuring 10 cm in diameter. External surface was smooth and the cut surface was gray-white, slightly whorled with no areas of necrosis or hemorrhage (Fig. 1). Microscopically, there were loosely arranged spindle cells in a myxoid stroma with inflammatory infiltration composed predominantly of lymphocytes and plasma cells; overall the appearance resembled nodular-fascitis. There were also cellular areas consisting of fascicles of bland spindle cells with open vesicular nuclei. No nuclear atypia or mitoses or infiltration was noted in multiple sections studied (Fig. 2). A morphological diagnosis of IMT of the uterus was made.

Discussion

First described in the year 1973 as a reactive "inflammatory pseudotumor" of the lung, IMT is now reconsidered as a unique neoplastic process of low malignant potential based on the different molecular/genetic characteristics and biologic behavior. Inflammatory myofibroblastic tumor is reported most commonly

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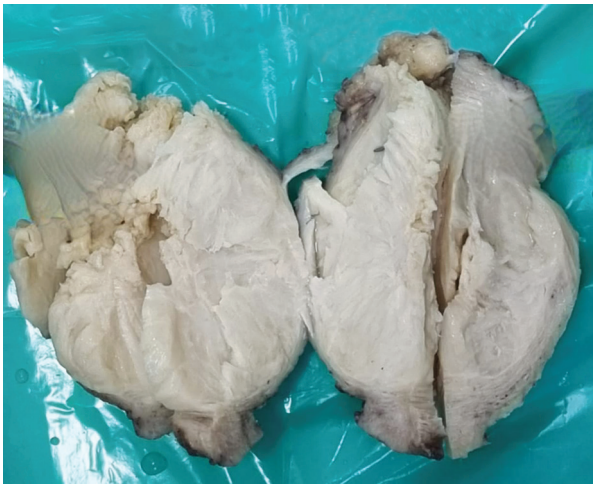

Fig. 1: Gross appearance of the tumor

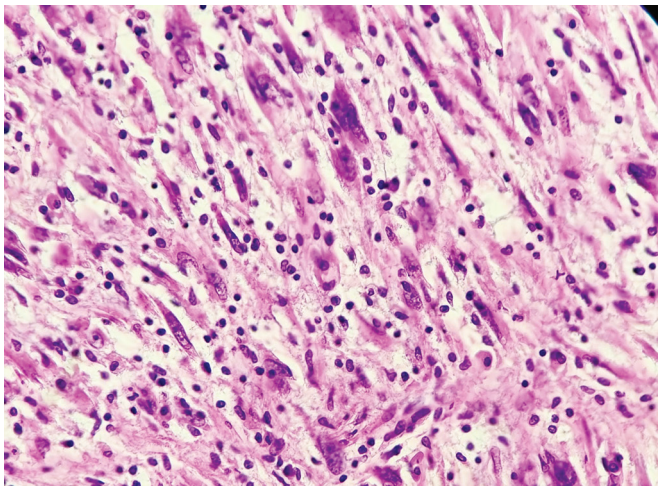

Fig. 2: Microscopy of the tumor showing "nodular fasciitis-like" appearance. (H&E, HP ×400)

in lungs, mesentery, omentum, and retroperitoneum. Rarely, IMT is reported in the uterus. Gynecological IMTs are often underrecognized as their morphological features overlap with other mesenchymal tumors common in the female genital tract like smooth muscle or endometrial stromal tumors.[2–4]

In a meta-analysis of uterine IMT by Mandato et al., the age at presentation of this tumor ranged from 6 to 78 years [mean, 40.6 ± 14.9 years; symptoms were reported by 48.6% patients – abdominal/pelvic pain (34.3%), vaginal bleeding (37.1%), fever/weight loss (14.3%), abdominal distension (8.6%), urinary disorders (5.7%), fatigue (5.7%), uterine prolapse (2.9%), and mass (17.1%)]. In 8.6%, a pregnant woman discovered the IMT during prenatal routine visit, and 2.9% discovered the IMT during surgery for endometriosis.[3] Our case was a 33-year-old lady who presented clinically with menorrhagia and anemia. Whereas about half of the uterine IMTs present as a submucosal polypoid mass, others present as discrete intramural or subserosal mass. In the present case, the 8.7 × 7 cm intramural mass was localized within the anterior myometrium that was interpreted as leiomyoma both radiologically and grossly. In the published literature, the size of IMT varied from 1.5 to 20 cm. Because of a presumptive diagnosis of leiomyoma, myomectomy was performed and the specimen was sent for histopathology. Three histologic growth patterns are reported: myxoid, fascicular/compact, and hyalinized patterns with variable degrees of lymphoplasmacytic inflammation.[2–4] Mitotic index is low and necrosis is uncommon. Histology of our case had predominantly a myxoid pattern with lymphoplasmacytic infiltration giving a nodular-fascitis-like appearance. Although smooth muscle tumors, endometrial stromal tumors, and extra-gastrointestinal tumors (E-GIST) were considered differential because myxoid change is reported in these entities (rather uncommonly), the typical nodular-fascitis-like is diagnostic of IMT. ALK immunohistochemistry can be helpful in doubtful cases.

Clinical Significance

Uterine IMT is a rare mesenchymal neoplasm often diagnosed as "fibroid" clinically, radiologically, and grossly. Inflammatory myofibroblastic tumors in general are indolent tumors with 25% recurrence rate and distant metastasis in approximately 2% of cases. Complete surgical resection seems to represent the best treatment for IMT. Both hysteroscopy and laparoscopy are effective and safe for diagnosis and management of uterine IMTs. However, IMT morcellation should be avoided because of the risk of abdominal/pelvic recurrence. Large size (>7 cm), moderate-to-severe atypia, high mitotic activity (>10 per 10 high-power fields), tumor cell necrosis, and infiltrative border have been hypothesized to be associated with aggressive behavior.[1] Patients with recurrent or metastatic IMT can benefit from ALK inhibitor-based targeted therapy, therefore, accurate diagnosis of these lesions is important.

References

1. Kertowidjojo EC, Bennett JA. Update on uterine mesenchymal neoplasms. Surg Pathol Clin 2022;15(2):315–340. DOI: 10.1016/j.path.2022.02.008.
2. Shukla PS, Mittal K. Inflammatory myofibroblastic tumor in female genital tract. Arch Pathol Lab Med 2019;143(1):122–129. DOI: 10.5858/arpa.2017-0575-RA.
3. Mandato VD, Valli R, Mastrofilippo V, et al. Uterine inflammatory myofibroblastic tumor: More common than expected: Case report and review. Medicine (Baltimore) 2017;96(48):e8974. DOI: 10.1097/MD.0000000000008974.
4. Pickett JL, Chou A, Andrici JA, et al. Inflammatory myofibroblastic tumors of the female genital tract are under-recognized. Am J Surg Pathol 2017;41(10):1433–1442. DOI: 10.1097/PAS.0000000000000909.