Massive cotyledenoid leiomyoma treated with uterine-conserving surgery

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Objective: To describe and illustrate a massive cotyledenoid leiomyoma treated with uterine-conserving surgery.
Design: Case report.
Setting: Medical center.
Patient(s): A 39-year-old woman with a large abdominal mass and a magnetic resonance imaging scan showing a 28-cm multi-lobulated mass.
Intervention(s): Laparotomy and myomectomy.
Main Outcome Measure(s): Recurrence and need for repeat surgery.
Result(s): No recurrence at 8 years of follow-up.
Conclusion(s): Cotyledonoid leiomyomas are rare. These benign tumors may be suspected preoperatively based on MRI appearance. Frozen section suggests a benign process and uterine-conserving surgery may be successfully accomplished. (Fertil Steril Rep® 2020;1:314–6. ©2020 by American Society for Reproductive Medicine.)
Key Words: Cotyledonoid leiomyomas, leiomyoma variants, benign fibroids, uterine-conserving surgery

Benign fibroids are the most common tumor of the genital tract in women and the diagnosis on both imaging and pathologic assessment is usually straightforward (1). However, some benign smooth muscle tumors have unusual growth patterns that suggest malignancy.

Cotyledonoid leiomyoma is a rare benign smooth muscle tumor variant that has been described in women of all ages (2). These tumors have an exophytic component with red, spongy, irregular bulbous protuberances that resemble cotyledons of the placenta. They may have an intramural component dissecting into the fascicles of the myometrium in an irregular fashion. The appearance is disconcerting and suggests malignancy.

We describe a 39-year-old woman with a 28-cm abdominal mass that appeared to resemble a malignancy on both magnetic resonance imaging (MRI) and at surgery. Frozen section suggested a benign fibroid, which was confirmed on final pathology with immunohistochemical analysis. Uterine-conserving surgery was performed. Eight-year follow-up shows no regrowth of the remaining broad ligament cotyledonoid fibroid. Written consent was obtained from the patient for publication of her case.

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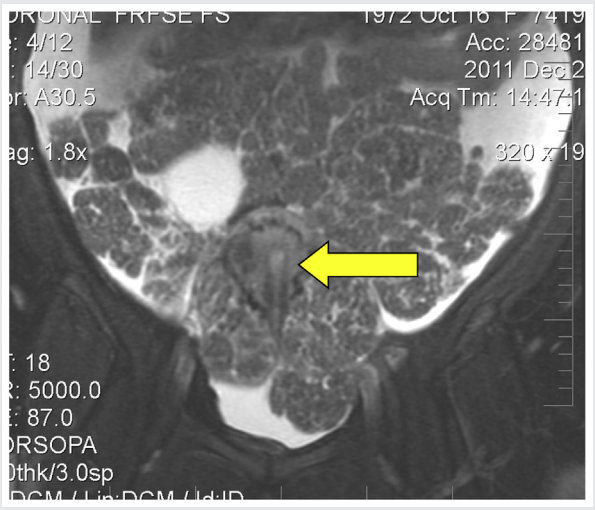
CASE

A 39-year-old patient who was G0P0 was diagnosed with uterine fibroids 4 years prior to being seen in our office. Ultrasonographic examination had described three fibroids, the largest being 13 cm. She had been recommended to have a hysterectomy and, wishing to preserve her fertility, sought an alternative-medicine provider. Her periods were regular and she wished to preserve fertility. When alternative medical treatment failed to provide symptom relief, she presented for evaluation and treatment.

The patient described symptoms of abdominal pressure and increasing abdominal girth, but denied weight loss, change in appetite, or other gastrointestinal symptoms. Menstrual periods were regular, lasting 4 days with normal flow. Abdominal examination revealed a 26-week-size mass, which did not extend into the pelvis on bimanual examination.

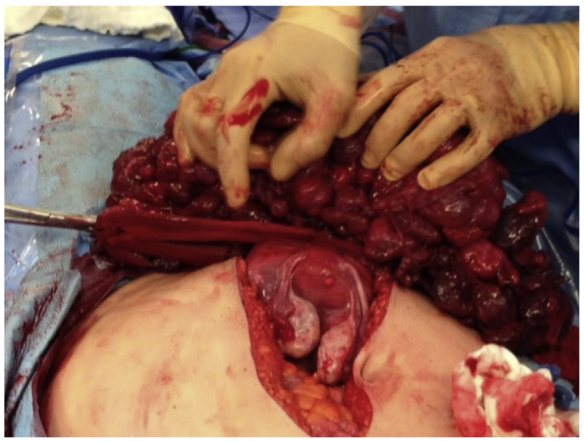
FIGURE 1



Magnetic resonance image, coronal T2 showing a multi-lobular mass with ascites. *Yellow arrow* points to normal uterine cavity.

Parker. Cotyledenoid leiomyoma. Fertil Steril Rep 2020.

FIGURE 2



Mass attached to uterine fundus with normal adnexa.

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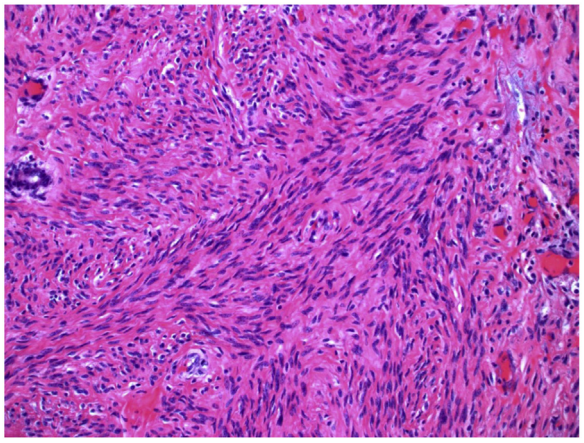
FIGURE 3



Spindle cell tumor with mild-moderate cellularity and bland uniform elongated nuclei.

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Pelvic and abdominal MRI showed a 28 x 24 cm multi-lobular pelvic/abdominal mass arising out of the anterior uterine myometrium. The uterus otherwise appeared normal and there was no evidence of invasion of the mass into surrounding structures. A moderate amount of pelvic fluid was present (Fig. 1).

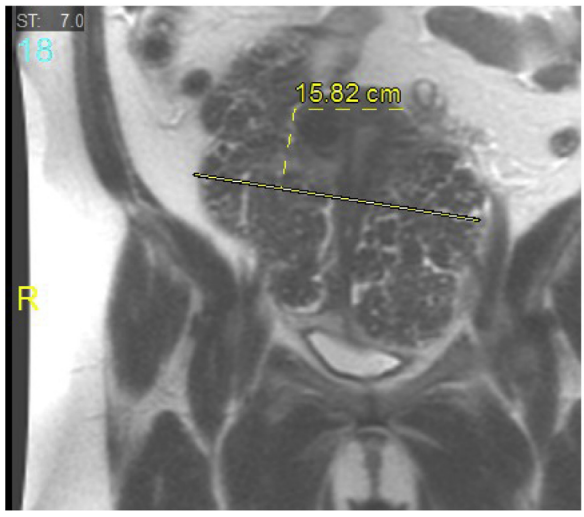
The possibility of uterine sarcoma or soft tissue sarcoma was considered and the patient was referred to a surgical oncologist. Laboratory data showed hemoglobin level of 17, CA-125 elevated to 80 (normal <35), and CA-27.29, CEA, and CA-19-9 were normal.

The patient consented to laparotomy and wished to preserve her uterus if possible. In February 2012, the patient had surgery through a midline incision. A large amount of ascites was suctioned, an irregular, multi-lobed tumor was exposed, and parasitic vascular attachments to the omentum were tied off and severed. Three separate segments of the tumor originating from the uterine fundus were dissected carefully and the vascular pedicles were clamped and severed (Fig. 2). The tumor was exophytic (International Federation of Gynecology and Obstetrics [FIGO] type 6) and dissection did not breach the myometrium, which remained intact. Running sutures and bipolar desiccation were used to achieve hemostasis. Tumor in the right and left broad ligaments was not resected in an effort to preserve the uterus.

Frozen section revealed a bland spindle cell tumor consistent with a benign leiomyoma. The specimen was an aggregate of red hemorrhagic nodular masses connected by fat and fibrous bands, weighing 3,797 g. Sectioning revealed a pink nodular, bulging, solid cut surface with a few areas of cystic degeneration. Final histological sections showed a uniform lobulated growth pattern of a spindle cell tumor with mild-moderate cellularity and bland uniform elongated nuclei (Fig. 3). No cytologic atypia, increased mitotic activity, coagulative necrosis, or intravascular presence of tumor was present. Immunocytochemistry was positive for actin and desmin and positive for estrogen receptors (1–2+) and progesterone receptors (3+). Ki-67, a cellular marker for proliferation, was low (2%–3%). The final diagnosis was leiomyoma, cotyledonoid type.

Three months following surgery, hemoglobin and CA-125 levels reverted to normal. Eight years following surgery, the patient is symptom free and doing well. Her pelvic examination is unchanged, and a recent MRI shows no further growth of the cotyledonoid leiomyoma within the broad

FIGURE 4



Recent magnetic resonance image shows no further growth of the cotyledonoid leiomyoma within the broad ligaments.

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ligaments (Fig. 4). Follow-up continues with pelvic examinations every 6 months and yearly ultrasonography to monitor growth or further extension into the broad ligament.

DISCUSSION

Cotyledonoid leiomyomas are rare benign uterine tumors, although they have a worrisome appearance with a grossly irregular exophytic mass that may raise concern for malignancy (3). Women found to have cotyledonoid leiomyomas range from 23 to 73 years, with distribution evenly throughout the decades. Common presenting symptoms include pelvic pain, pelvic mass, and vaginal bleeding (4). Tumor markers are usually normal, although CA-125 was elevated in our patient.

Magnetic resonance imaging of a cotyledonoid leiomyoma shows heterogeneous intensity on T2-weighted sequences due to partially confluent patchy nodules isointense to muscle (5). Hyperintense nodules on T2-weighted images without a signal drop after fat suppression indicate hydropic degeneration and congested connective tissue and the presence of flow voids within the mass indicates high vascularity of the lesion. Cotyledonoid leiomyomas may dissect into the myometrium and can involve the broad ligament and retroperitoneum, but they do not invade surrounding structures. Magnetic resonance imaging findings are not specific and tissue sampling is required to establish the diagnosis.

Hysterectomy, including bilateral salpingo-oophorectomy, is often performed due to the malignant appearance of the tumor. Frozen section was performed intraoperatively in our case and in four other reported cases and bland, mitotically inactive nuclei were present, consistent with a benign tumor (6). For women who wish uterine conservation, these features allow uterine-conserving surgery pending the final pathologic diagnosis, which can be confirmed with immunohistochemical analysis.

Of 44 reported cases of cotyledonoid leiomyomas, five cases of uterine-conserving surgery (with incomplete resection of the tumor) have been described (2, 7–10). Only one recurrence of tumor growth has been reported within 6–60 months of follow-up (11). Our patient has had no regrowth in 8 years (96 months) of follow-up. Although the mechanism for regulation of leiomyoma cell growth is unknown and likely complex, leiomyoma cells are known to be sensitive to mechano-transduction. By enclosing the small remaining broad ligament tumor with sutures and electrosurgery, cell proliferation may have been altered (12). Our patient had an elevated hemoglobin level that reverted to normal 3 months after surgery, suggesting erythropoietin production by the tumor, as has been previously reported for benign fibroids (13).

In summary, cotyledonoid leiomyoma of the uterus is a very rare benign tumor that is suggested by the noninvasive appearance on MRI and at surgery. Frozen section suggests a benign process and can help the patient avoid unnecessary hysterectomy.

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