Cotyledonoid dissecting leiomyoma in pregnancy: a case report

Samuel Choice,[1] Mildrede Bonglack,[2] Michael Goodheart,[2] Megan McDonald,[2] Audai Alrwashdeh,[3] Holly Bolger[2]

Keywords: cotyledonoid dissecting leiomyoma, leiomyoma, leiomyoma in pregnancy, benign smooth muscle neoplasm

Abstract

Cotyledonoid dissecting leiomyoma (CDL) is a rare variant of uterine leiomyoma characterized by intramural dissection within the uterine corpus and a distinctive gross appearance resembling the cotyledons of the placenta. Despite their alarming, sarcomatous appearance both macroscopically and radiographically, these tumors are typically associated with bland histologic findings. Approximately 70 cases of CDL have been described in the literature. This case represents the second described case of CDL in pregnancy, the first in which a pregnancy was carried to term. A 28-year-old primigravid was incidentally found to have a massive, exophytic growth of the uterus on ultrasound with concomitant 14-week intrauterine pregnancy. The pregnancy was monitored with serial growth ultrasounds. She was delivered at 37 weeks via Cesarean section. After being lost to follow-up, the patient presented 2.5 years later with worsening abdominal fullness and persistent uterine mass.

[1]University of Iowa Carver College of Medicine, Iowa City, Iowa
[2]Department of Obstetrics and Gynecology, University of Iowa, Iowa City, Iowa
[3]Department of Pathology, University of Iowa Carver College of Medicine, Iowa City, Iowa

Introduction

Leiomyomas are extremely common benign smooth muscle neoplasms of the myometrium that occur in nearly 80% of women by 50 years of age.[1] While the typical appearance of well circumscribed, uniformly expansile fibroids are well established, certain leiomyomas may display more unusual growth patterns that can present a diagnostic challenge for clinicians. Cotyledonoid dissecting leiomyomas (CDL) present an exceedingly rare growth pattern for uterine benign smooth muscle neoplasms, and are characterized by an

Please cite this paper as: Choice S, Bonglack M, Goodheart M, McDonald M, Alrwashdeh A, Bolger H. Cotyledonoid dissecting leiomyoma in pregnancy: a case report. Proc Obstet Gynecol. 2023;12(1): Article 6 [10 p.]. Available from: https://pubs.lib.uiowa.edu Free full text article.

Corresponding author: *Holly Bolger, DO, Department of Obstetrics and Gynecology, University of Iowa Hospitals and Clinics, Iowa City, IA, holly-marie-bolger@uiowa.edu*

Financial Disclosure*: The authors report no conflict of interest.*

exophytic, grape-like appearance resembling placental tissue.[2] In total, approximately 70 cases of CDL have been reported,[3] with only one previously described in pregnancy.[4] The large size of the tumor, strong vascularization, heterogeneity, and tendency to invade surrounding structures, can raise suspicion of malignancy, and can lead surgeons to perform more radical surgical resections than needed.[10] This report describes the second case of CDL in pregnancy, and the first in which a pregnancy was carried to term with a cotyledonoid dissecting leiomyoma present within the uterus. The mass and uterus were removed nearly 3 years after the patient's delivery, as she was initially lost to follow-up. Consent from the patient and subsequent IRB approval were obtained for description of this case.

Description of case report

A 28-year-old primigravid female presented for an initial prenatal visit after a positive home pregnancy test. Her previous menstrual history was unremarkable. After initial maternal ultrasound failed to visualize the pregnancy secondary to uterine distortion, an MRI was performed showing both a viable 14-week intrauterine pregnancy, as well as a 10 x 13 x 25 cm cluster of exophytic masses arising from the left-lateral surface of the uterus. The adnexal structures were unremarkable. Figure 1. Physical exam showed a non-tender, non-distended abdomen with a palpable uterine mass above the umbilicus. After referrals to the University of Iowa Gynecologic-Oncology and Maternal Fetal Medicine divisions, the patient was instructed on possible management scenarios. Using the radiologist's read, as well as opinions from MFM and gynecologic oncology physicians present on the case, it was presumed the mass could be a massive sub-serosal uterine fibroid with cystic, degenerative components, but could not rule out uterine sarcoma. Consultations with gynecologic oncology and MFM, team determined based on the vascularity of the tumor and its size, that core needle biopsy, or any surgical intervention, could risk massive hemorrhage and miscarriage. The team believed based on the above factors, the only way to rule out sarcoma would entail termination of the pregnancy and subsequent myomectomy. After explaining the risks of possible delay in treatment of a sarcoma, as well as the possible risks to mother and baby of carrying a pregnancy with such a large uterine mass, the patient elected to continue the pregnancy, and the team arranged for fetal-growth ultrasounds every 4 weeks. Further surgical intervention at 6 months postpartum was planned for the uterine mass.

Her pregnancy was uncomplicated aside from breech presentation and enlargement of the exophytic mass from a first trimester volume of 25 x 12 x 15 cm (18,850 cm ³) to a third trimester volume of 30 x 15 x 16 cm (30,159 cm ³) Figure 2. Delivery, done by a high-risk obstetrician, was performed via classical cesarean section thru a vertical midline incision. At the time of cesarean section, intra-operative consultation with gynecologic oncology occurred and a 2x2 cm biopsy of the mass was obtained, via clamping, cutting, and tying a small protrusion of the mass. The mass was described as subserosal, violaceous, and hypervascular, arising from the lateral fundus and extending anteriorly into the right pelvis. Pathologic analysis

of biopsies demonstrated leiomyoma versus adenomyoma with unsampled glands. The patient's postpartum course was uneventful; however, the patient was lost to follow-up.

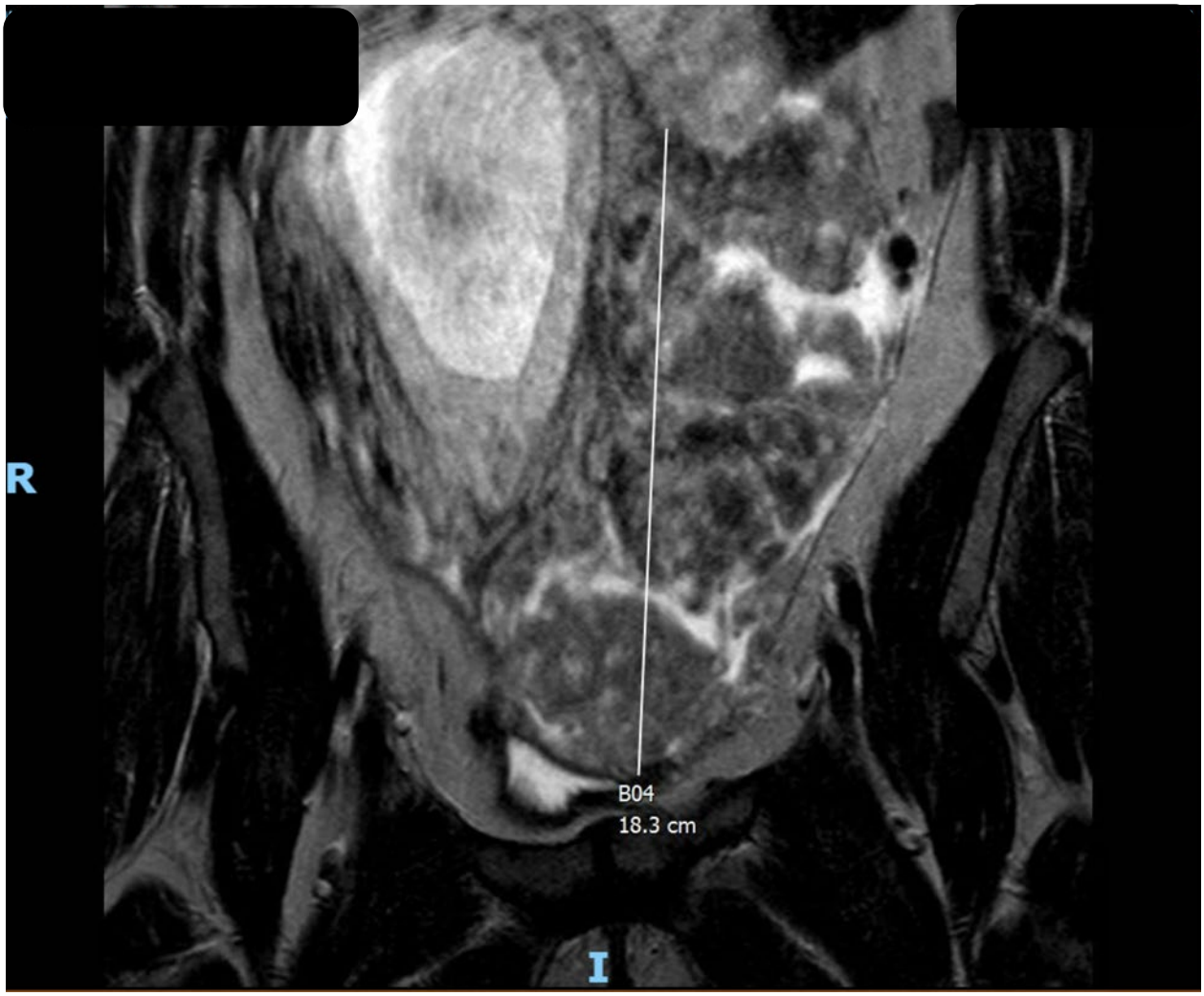

Figure 1: MRI. Initial T2 Coronal MRI of patient's abdomen at 14 weeks, showing gravid uterus displaced to the right, with anterior placenta. A lobulated extrauterine mass, 12x25x15 predominantly on the left, was isointense on T1, and hyperintense on T2. Noted on read to have vascular stalk tracing to L uterine wall. Areas of hyperintense fluid on T2 can be noted throughout.

Upon presentation 2.5 years later for worsening abdominal fullness, a bimanual exam revealed the cervix to be barely palpable, with the uterus palpable at the level of the umbilicus. An MRI showed a T1/T2 hyperintense, 27 x 11 x 26 cm (32,346 cm^3), exophytic mass arising from the fundus and body of the

uterus, extending into the upper abdomen, right pelvis, and herniating into the umbilicus. Additional cystic and hemorrhagic components up to 5 cm in size were also noted within the mass. Adnexal structures were unable to be visualized. The patient had completed childbearing and elected for definitive surgical management via an exploratory laparotomy with total abdominal hysterectomy and bilateral salpingectomies.

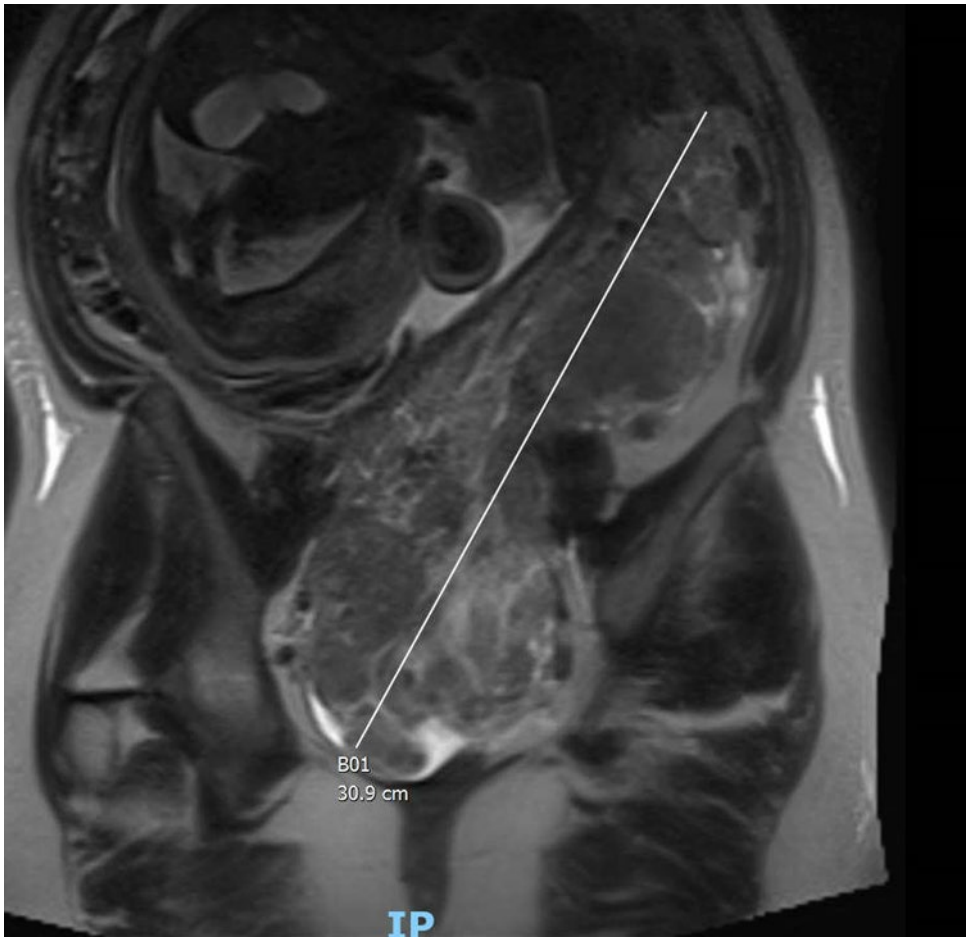

Figure 2: Coronal MRI Pelvis, T2, of patient at 36 weeks pregnancy, showing large, broad-based, lobulated exophytic mass arising from the left lower uterine segment. Interval enlargement to 30 x 15 x 16cm. The mass is hypointense on T1 and heterogeneously hypointense on T2.

Upon entry into the peritoneal cavity, a villous, partially cystic, finger-like group of masses arose from the uterus anteriorly and posteriorly, with a small

anterior projection herniating into the abdominal wall in the supra-umbilical region Figure 3, Figure 4. The posterior mass demonstrated retroperitoneal and pelvic side-wall invasion, with further fingerlike extension into the sigmoid mesentery, adherence to the rectum, and extension into the left ovary and left infundibulo-pelvic ligament. Bilateral fallopian tubes and right ovary were normal. Intra-operative pathologic frozen section revealed a smooth muscle neoplasm.

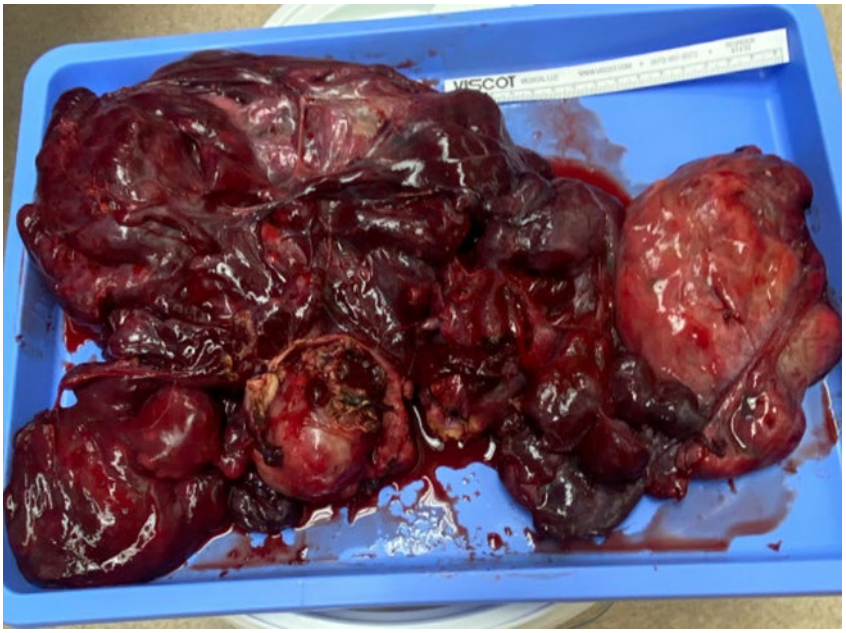

FIGURE 3: Gross image of the resected posterior uterine mass showing cystic, hemorrhagic components.

A resection of the uterine mass, extensions into pelvic sidewall, posterior-cul-de-sac, left adnexa, and mesenteric implants was performed without complication although the procedure was quite challenging given the cotyledonoid, infiltrative nature of the mass. With careful dissection of the mass, the left ovary was preserved and a left oophoropexy was performed to secure the ovarian pedicle to the left pelvic sidewall. There was concern for subtotal resection of microscopic implants near the rectum, due to risk of iatrogenic injury. Pathologic analysis indicated a benign smooth muscle neoplasm, consistent with cotyledonoid dissecting leiomyoma Figure 4.

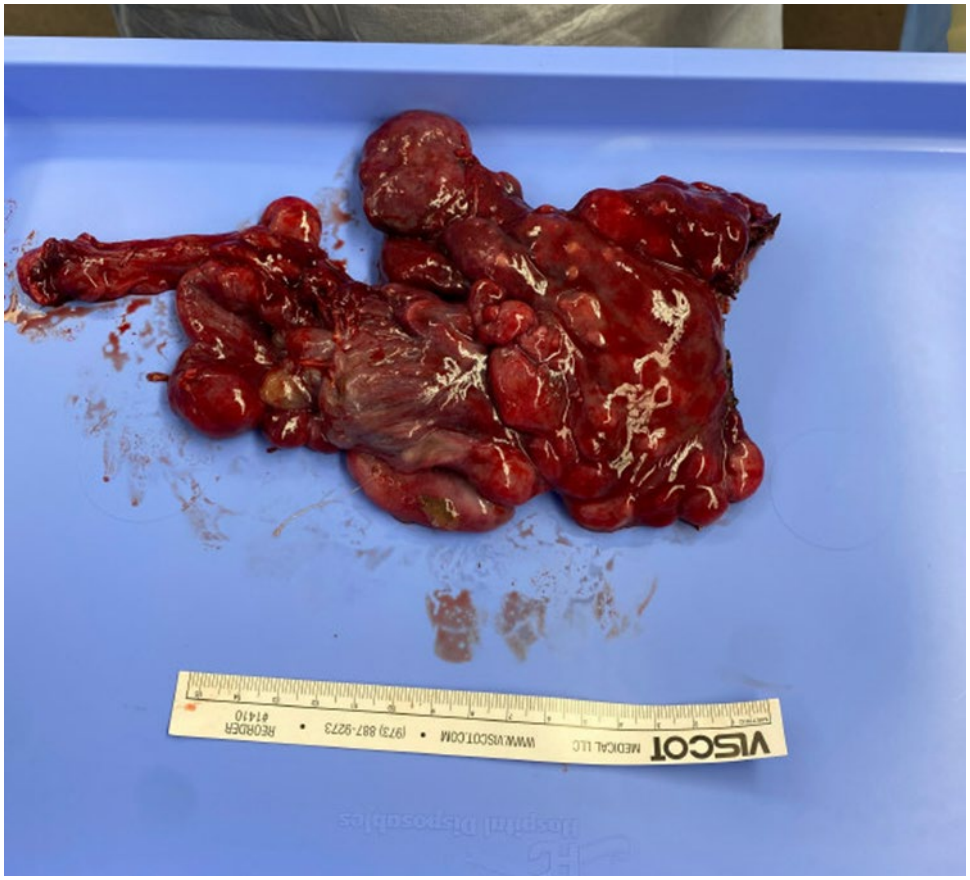

Figure 4: Gross appearance of anterior portion of the resected uterine mass, showing villous, finger-like projections and cotyledonoid nodules varying from 0.5 cm-5cm in their greatest dimension.

Microscopic examination showed a tumor that is composed of spindled cells that are arranged in broad, sweeping fascicles (A; 10x) with intervening hydropic changes and dilated blood vessels (B; 10x). There is no cytological atypia, tumor necrosis or increased mitotic activity (up to 1 mitosis per 10 high power fields). By immunohistochemistry, the constituent cells are positive for desmin (C; 20X) and smooth muscle specific actin (D; 20X) Figure 5.

Due to the benign clinical course of this tumor, no further treatment was recommended at the time. At the recommendation of the gynecologic oncology tumor board, the patient will be initially followed with pelvic CT scans every 6 months for surveillance.

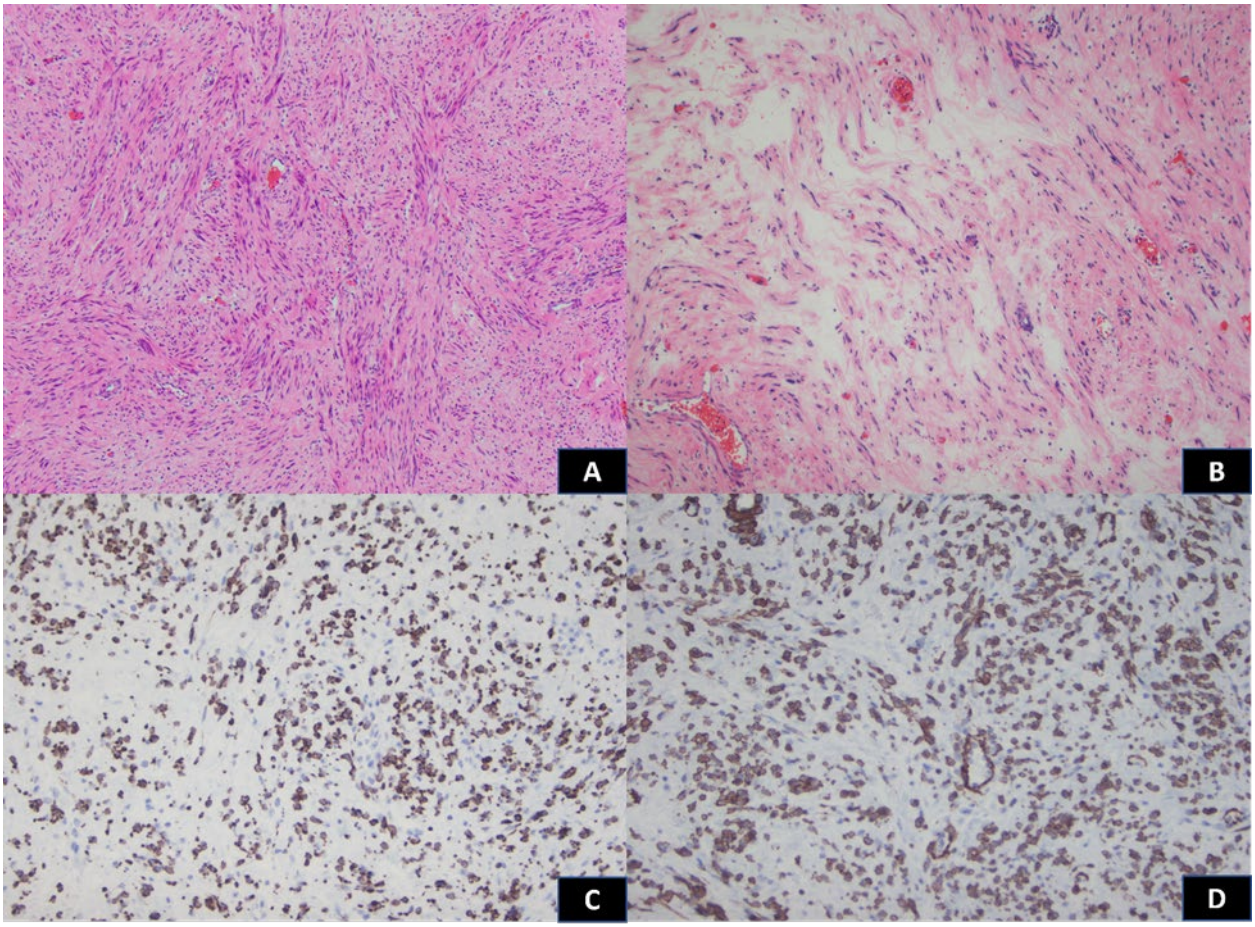

Figure 5: Microscopic examination showed a tumor that is composed of spindled cells that are arranged in broad, sweeping fascicles (A; 10x) with intervening hydropic changes and dilated blood vessels (B; 10x). There is no cytological atypia, tumor necrosis or increased mitotic activity (up to 1 mitosis per 10 high power fields). By immunohistochemistry, the constituent cells are positive for desmin (C; 20X) and smooth muscle specific actin (D; 20X).

Discussion

Uterine leiomyomas, colloquially known as “fibroids,” are the most common benign smooth muscle neoplasm of the uterus with well-established treatment regimens and known effects on conception and pregnancy. Unique and rarely observed growth patterns of uterine leiomyomas, such as cotyledonoid dissecting leiomyomas, may present a diagnostic and interventional challenge to clinicians, especially when concurrent with pregnancy.[6]

Cotyledonoid dissecting leiomyoma (sometimes referred to as a Sternberg Tumor) was first described by Roth et al. in 1996 as a grape-like, exophytic smooth muscle neoplasm that resembles the cotyledons of the placenta.[7] To date, approximately 70 cases of CDL have been described in the literature, with only one other present in pregnancy. These tumors arise in a wide age range of 21-73 years, with a median size of 12 cm.[3] Patients with CDL endorse typical fibroid mass-effect symptoms such as pelvic fullness, abdominal pain, and abnormal uterine bleeding.[3,8] In this case, the patient's tumor was found incidentally on an initial antenatal ultrasound, and later confirmed with MRI.

Imaging of CDL may concern the diagnostician for malignancy, as CDL often presents as a heterogeneous, irregular, large, and highly vascularized mass with potential invasion into the retroperitoneum or pelvic sidewall.[11] Upon surgical exploration, the exophytic, villous appearance, large tumor size, and widespread infiltrative growth within the pelvic cavity may deepen concern for malignancy. Histologically, however, these tumors are generally bland, mitotically inactive leiomyomas that lack the typical features of sarcoma growth such as mitotic activity, nuclear atypia, cellular pleomorphism, or necrosis.[3,6] As in this case, CDL is microscopically characterized by uniform smooth muscle nodules of diverse sizes which are rich in vessels.

Surgical resection is the best treatment for CDL, however due to the sarcomatoid appearance of the lesion, overtreatment may occur. Most patients with CDL undergo non-conservative treatment with total hysterectomy (80%), while a minority undergo myomectomies (20%).[3,12] Frozen sectioning should be performed intraoperatively to prevent overtreatment if possible. For older patients or those who do not desire future fertility, hysterectomy is a suitable option, as was the case in this patient. For patients desiring future fertility, myomectomy, even for larger tumors (25+ cm), has been shown to be an effective treatment, but has been associated in certain cases with recurrence of the tumor likely secondary to incomplete resection.[9,12,13] Despite sparse recurrence with myomectomy, there have been no reported cases of malignant transformations or metastases, with the longest follow-up being 41 years.[8] In our case, due to the proximity of implants to the rectal mucosa, there was concern for subtotal excision. As such, radiographic surveillance for recurrence was chosen as the follow-up plan.

Typical leiomyomas have been known to be estrogen dependent and grow in pregnancy, as was seen in the above case. They are known to increase the risk of spontaneous abortion, fetal malpresentation, placenta previa, preterm birth, peripartum hemorrhage, and cesarean section.[5] However, there is a paucity of data on rarer leiomyoma growth patterns in pregnancy, with only one prior case of CDL being described in pregnancy. The earlier reported case was managed with myomectomy at 14 weeks, with an uncomplicated C-section at term. The above case is the first reported case of a CDL that was left untreated during pregnancy and was only complicated by breech presentation and later cesarean section. This case helps add to sparse data of CDL in pregnancy and demonstrates the possibility of successful conservative management of

the pregnant patient with suspected CDL.

While cotyledonoid dissecting leiomyomas are an exceedingly rare growth pattern of a common tumor, their sarcomatous appearance may pose significant diagnostic and management challenges for clinicians, especially when encountered in pregnancy. The literature demonstrates that CDL is a benign disease with a great prognosis that can be definitively managed with surgical excision.

References

1. Zimmermann A, Bernuit D, Gerlinger C, Schaefers M, Geppert K. Prevalence, symptoms and management of uterine fibroids: an international internet-based survey of 21,746 women. BMC Womens Health. 2012 Mar 26;12:6. https://doi.org/10.1186/1472-6874-12-6. PMID: 22448610; PMCID: PMC3342149.

2. Saeed AS, Hanaa B, Faisal AS, Najla AM. Cotyledonoid dissecting leiomyoma of the uterus: a case report of a benign uterine tumor with sarcomalike gross appearance and review of literature. Int J Gynecol Pathol. 2006 Jul;25(3):262-7. https://doi.org/10.1097/01.pgp.0000186542.19434.ea. PMID: 16810065.

3. Buonomo F, Bussolaro S, Fiorillo CA, Giorda G, Romano F, Biffi S, Ricci G. The Management of the Cotyledonoid Leiomyoma of the Uterus: A Narrative Review of the Literature. Int J Environ Res Public Health. 2021 Aug 12;18(16):8521. https://doi.org/10.3390/ijerph18168521. PMID: 34444270; PMCID: PMC8393327.

4. Mathew M, Gowri V, Al Hamdani A, Machado L, Rao K, Shabnam S. Cotyledonoid leiomyoma in pregnancy. Obstet Gynecol. 2007 Feb;109(2 Pt2):509-11. https://doi.org/10.1097/01.AOG.0000222359.34860.91. PMID: 17267876.

5. Parazzini F, Tozzi L, Bianchi S. Pregnancy outcome and uterine fibroids. Best Pract Res Clin Obstet Gynaecol. 2016 Jul;34:74-84. https://doi.org/10.1016/j.bpobgyn.2015.11.017. Epub 2015 Nov 25. PMID: 26723475.

6. Jamal I, Gupta RK, Sinha RK, Bhadani PP. Cotyledonoid dissecting leiomyoma: an uncommon form of a common disease. Obstet Gynecol Sci. 2019 Sep;62(5):362-366. https://doi.org/10.5468/ogs.2019.62.5.362. Epub 2019 Jul 26. PMID: 31538081; PMCID: PMC6737059.

7. Roth LM, Reed RJ, Sternberg WH. Cotyledonoid dissecting leiomyoma of the uterus. The Sternberg tumor. Am J Surg Pathol. 1996 Dec;20(12):1455-61. https://doi.org/10.1097/00000478-199612000-00004. PMID: 8944038.

8. Smith CC, Gold MA, Wile G, Fadare O. Cotyledonoid dissecting leiomyoma of the uterus: a review of clinical, pathological, and radiological features. Int J Surg Pathol. 2012 Aug;20(4):330-41. https://doi.org/10.1177/1066896912450315. Epub 2012 Jun 18. PMID: 22710314.

9. Roth LM, Kirker JA, Insull M, Whittaker J. Recurrent cotyledonoid dissecting leiomyoma of the uterus. Int J Gynecol Pathol. 2013 Mar;32(2):215-20. https://doi.org/10.1097/PGP.0b013e318257dff4. PMID: 23370645.

10. Buonomo F, Bussolaro S, Giorda G, Romano F, Biffi S, Ricci G. Cotyledonoid Leiomyoma Clinical Characteristics, Imaging Features, and Review of the Literature. J Ultrasound Med. 2021 Jul;40(7):1459-1469. https://doi.org/10.1002/jum.15510. Epub 2020 Sep 21. PMID: 32955750.

11. Preda L, Rizzo S, Gorone MS, Fasani R, Maggioni A, Bellomi M. MRI features of cotyledonoid dissecting leiomyoma of the uterus. Tumori. 2009 Jul-Aug;95(4):532-4. https://doi.org/10.1177/030089160909500422. PMID: 19856670.

12. Xu T, Wu S, Yang R, Zhao L, Sui M, Cui M, Chang W. Cotyledonoid dissecting leiomyoma of the uterus: A report of four cases and a review of the literature. Oncol Lett. 2016 Apr;11(4):2865-2868. https://doi.org/10.3892/ol.2016.4314. Epub 2016 Mar 8. PMID: 27073566; PMCID: PMC4812148.

13. Özdemir Ö, Sağır G, Akbaş B, Güven S, Reis, A. A case report on recurrent cotyledonoid dissecting leiomyoma. J Clin Obstet Gynecol. 2019;29(4):148-150. https://doi.org/10.5336/jcog.2019-71191