Impact of an accessory cavitated uterine mass on fertility: case presentation and review of the literature

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Objective: We report a case of an accessory cavitated uterine mass (ACUM) in a patient with infertility and chronic pelvic pain. In addition, we summarize the literature to better characterize ACUM diagnosis and management.

Design: A comprehensive literature search using the PubMed database was performed through April 2023. Historical ACUM diagnostic criteria were applied as inclusion criteria. Descriptive statistics and statistical evaluation were reported.

Results: A 31-year-old nulligravid woman presented with chronic pelvic pain, dysmenorrhea, primary infertility, and history of endometriosis. Three-dimensional ultrasonography identified an ACUM and laparoscopic excision provided complete resolution of symptoms. Subsequently, she conceived without assistance twice with uncomplicated vaginal deliveries. A total of 154 articles were identified, 34 papers met inclusion criteria and were individually reviewed, consisting of 70 reported cases. The most common presenting complaints were dysmenorrhea (81.4%), chronic pelvic/abdominal pain (54.1%), and refractory pain (34.3%). Diagnostic imaging included magnetic resonance imaging (62.9%) and transvaginal ultrasound (55.7%). Management included resection via laparoscopy (75.7%) or laparotomy (18.6%), or hysterectomy (5.7%). Of cases with reported outcomes, 90.7% had complete relief of symptoms after surgery.

Conclusion: ACUM often presents with dysmenorrhea, chronic pelvic pain, or abdominal pain and is identifiable on magnetic resonance imaging as a hyperenhancing mass. Three-dimensional transvaginal ultrasound can also accurately identify ACUM. A total of 90.7% of patients had complete relief of symptoms after intervention. It is important to identify ACUM early to relieve pain and reduce unnecessary interventions. Like our patient, other reports have demonstrated concomitant infertility and endometriosis. However, further investigation is needed to explore the association between infertility and ACUM. (Fertil Steril Rep® 2023;4:402-9. ©2023 by American Society for Reproductive Medicine.)

Key Words: Accessory cavitated uterine mass, infertility, chronic pelvic pain, case report, literature review

ccessory cavitated uterine mass (ACUM) is a rare Müllerian anomaly characterized by the presence of an accessory uterine cavity in close proximity with the uterus, but non-communicating with its main body. Initially described in

1912, these masses contain functional endometrial and myometrial components that can be found within the external myometrium and/or in the broad ligament in an otherwise normal uterus (1, 2). The etiology of ACUM is likely congenital and is thought to

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involve duplication and persistence of Müllerian ductal tissue and possibly gubernaculum dysfunction (3). Although approximately 7% of women will be diagnosed with a reproductive tract anatomic anomaly in their life (4), the exact prevalence of ACUM is unknown. Furthermore, it has been postulated that ACUM may be more prevalent than previously thought, with the theory that cases of ACUM may have been misclassified in the past as a uterine-like mass, adult adenomyosis, or juvenile cystic adenomyoma (JCA) (5).

ACUM most commonly present clinically with dysmenorrhea and cyclic pelvic pain because of the functional endometrium and bleeding within the lesion during the menstrual cycle. The diagnosis of ACUM is challenged by the broad differential diagnosis including rudimentary and cavitated uterine horns, cystic adenomyosis, endometriosis, degenerated leiomyomas, and primary dysmenorrhea (5). Rudimentary and cavitated uterine horns result from failure of Müllerian ducts to develop a normal uterine cavity, whereas an ACUM is a remnant of an embryologic structure associated with an otherwise structurally normal uterus (3, 6). Cystic or cavitated adenomyosis constitute a heterogeneous group with likely different etiologies (7). It was proposed that these lesions are further classified on the basis of their epithelial lining (8).

For the diagnosis of ACUM, the following criteria are proposed: an isolated accessory cavitated mass; a normal uterus, fallopian tubes, and ovaries; a surgical case with an excised mass and a pathological examination; an accessory cavity lined by endometrial epithelium with glands and stroma; a chocolate-brown-colored fluid content; and no adenomyosis, although there could be small foci of adenomyosis in the myometrium adjacent to the accessory cavity (3). The recommended treatment of ACUM typically involves excision of the mass resulting in resolution of associated symptoms. As a result of the broad differential diagnosis and relatively low index of suspicion among clinicians, the diagnosis of ACUM may be significantly delayed. Furthermore, the impact of ACUM on infertility has not been described. Herein, we present a case of ACUM in a patient with infertility and chronic pelvic pain with subsequent spontaneous conception and uncomplicated delivery after excision of the mass. In addition, we present a literature review investigating ACUM presentation, diagnosis, imaging findings, and treatment.

MATERIALS AND METHODS

Informed consent was obtained from our patient to share deidentified information regarding her case presentation. For the literature review, PubMed was searched using the terms: "uterine-like mass," "accessory cavitated uterine mass," "uterus-like mass," and "broad ligament endometriosis" including all studies through April 2023. Inclusion was based on existing historical diagnostic criteria of ACUM established by Acien et al. (3): an isolated accessory cavitated mass; a normal uterus, fallopian tubes, and ovaries; a surgical case with an excised mass and a pathological examination; an accessory cavity lined by endometrial epithelium with glands and stroma; and chocolate-brown-colored fluid content. However, patients with concomitant adenomyosis were included in the review. Because of ongoing debate regarding whether JCA should be grouped with ACUM, it was unclear whether JCA met the true diagnostic criteria and were therefore excluded. All publicly available cases that met the above criteria were included in the final analysis, which evaluated ACUM presentation, diagnosis, radiologic modalities and findings, management, outcomes, and association with other findings such as infertility and endometriosis. Descriptive statistics were reported, and statistical evaluation were

conducted using Fisher's exact test or Chi-square analysis, with statistical significance defined as P<.05.

CASE REPORT Patient History

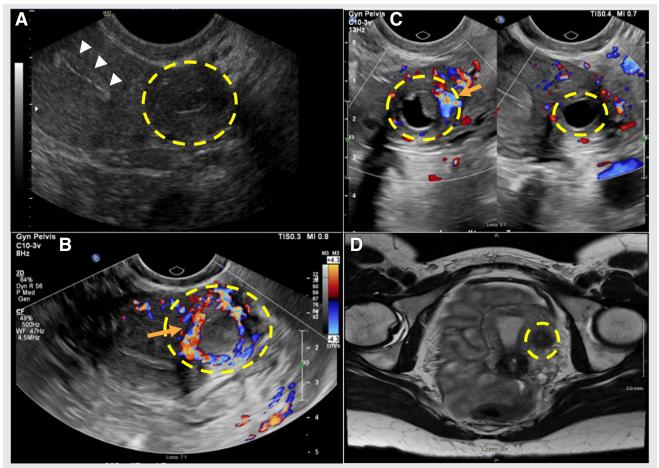
A 31-year-old nulligravid woman sought consultation because of primary infertility that had persisted for 7 years. She had no significant past medical history except for her gynecological history and had a normal body mass index. Her partner had a history of Stage IV-B, Nodular Sclerosing Hodgkin Lymphoma 2 years before her presentation and received 6 monthly cycles of chemotherapy with remission. Because of late-stage diagnosis, gamete cryopreservation was not considered by the patient or her partner at the time of diagnosis.

The patient had a complex history of chronic pelvic pain and endometriosis. She reported 10 years of cyclic left-sided, sharp, stabbing pain occurring in coordination with menses that significantly impacted her functional capacity. She had multiple visits to several emergency departments for pain control. At her initial presentation to an Obstetrician-Gynecologist 9 years prior, pelvic ultrasound imaging was performed and revealed a thick-walled 1.8-cm mass with a central cystic component located in the left adnexa between the uterus and ovary (Fig. 1A).

She received empiric treatment with leuprolide acetate for suspected endometriosis for 1 year with some improvement in her symptoms during this treatment period. She underwent diagnostic laparoscopy, notable only for 3 small peritoneal implants along the left ovarian fossa. All other pelvic anatomy was reportedly normal without mention of a lesion in the left adnexa corresponding to prior imaging. The patient's symptoms were persistent, and she elected to restart leuprolide acetate treatment. On cessation of this treatment, her symptoms returned resulting in several emergency department visits. During these visits, ultrasonographic features of the lesion, although overall similar in location/size, revealed changes of the cyst contents. Specifically, the previously demonstrated anechoic cystic lesion now contained areas of echoic material within the cyst (Fig. 1B). Furthermore, vascularity surrounding the cyst was temporal in nature, where an acute pain episode displayed increased vascularity on the basis of Doppler flow and the cyst was completely replaced by anechoic material (Fig. 1C). Pelvic magnetic resonance imaging (MRI) favored an atypical exophytic fibroid without endometrial cavity involvement (Fig. 1D). In the interim, the patient underwent a second diagnostic laparoscopy with excision of a left fundal implant; pathologic analysis only revealed a dense serosal fibrous adhesion.

At this point, the patient had presented for infertility evaluation. Work up at that time did demonstrate male factor infertility with semen analysis parameters consistent with oligoasthenoteratozoospermia. Her ovarian reserve testing was within normal limits (AMH 2.2 ng/mL). Hysterosalpingography (HSG) revealed bilateral tubal patency and normal uterine contour. Given severe male factor, in vitro fertilization was recommended. However, the patient desired to pursue ovulation induction-intrauterine insemination cycles because of financial constraints. In midcycle ultrasound, the internal contents appeared markedly similar to the endometrium.

FIGURE 1



Diagnostic imaging of accessory cavitated uterine mass before patient presentation to treating clinical center. (A) Mass measuring 1.8 cm between the left ovary and uterus with central cystic component and relatively thick wall. (B) Complex cystic mass identified to the left of the endometrium measuring approximately 2.0 cm \times 1.9 cm \times 1.8 cm. (C) Cystic area with increased echogenicity and vascularity. (D) Pelvic magnetic resonance imaging favored an atypical exophytic fibroid without endometrial cavity involvement. Dashed circle indicates the accessory cavitated uterine mass, Doppler flow is indicated by yellow arrows, and white arrow heads indicate the endometrial cavity. *Strug. Impact of an accessory cavitated uterine mass on fertility. Fertil Steril Rep 2023.*

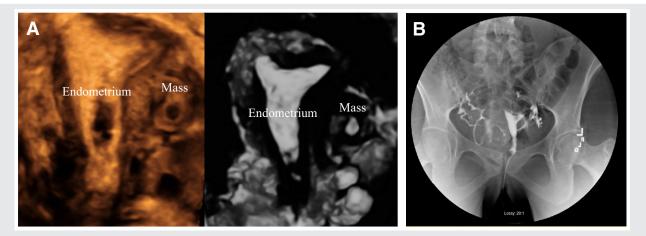
She completed 2 ovulation induction-intrauterine insemination cycles with very low sperm total motile counts without conception. The patient was referred to a specialist with experience in diagnosing uterine anomalies given a high index of suspicion the adnexal mass had functional endometrium, suggestive of an ACUM. Saline-infusion sonography (SIS) with three-dimensional (3D) ultrasound reconstruction demonstrated a left uterine mass in the mid-portion of the uterus, 13 mm in greatest dimension with a central hypoechoic area and internal trilaminar appearance during the patient's midcycle (Fig. 2A). On the basis of the imaging findings, there was a high index of suspicion for ACUM rather than a theoretical atypical non-communicating rudimentary uterine horn. The latter was less likely given HSG findings of bilateral tubal patency (Fig. 2B).

Laparoscopic Excision

Given continued acute pain, the patient elected to pursue surgical management of the uterine mass. The patient

underwent robotic-assisted laparoscopic excision of the left parametrial mass with excision of endometriosis present on the left posterior broad ligament (Fig. 3A). Red endometriotic implants were observed overlying the mass extending to the left broad ligament. The broad ligament peritoneum in this area was thickened and on dissection contained brown chocolate fluid. A solution of dilute vasopressin was injected around the identified broad ligament mass. An incision was made over the mass parallel to the round ligament. The incision was extended into the mass and there was no capsule suggestive of leiomyoma present. The mass was grasped and sharply dissected away from the surrounding fibrosis with minimal electrocautery while maintaining hemostasis. The defect was closed in 2 layers with O-polydioxanone barbed suture with the second layer incorporating serosa. The surgery was overall uncomplicated with minimal blood loss, and the patient was discharged home the same day.

FIGURE 2



Uterine cavity evaluation during assessment of accessory cavitated uterine mass. (A) Three-dimensional saline-infused sonogram with reconstruction revealed (left and right panel) a left-sided mass near the mid-portion of the uterus with a central hypoechoic area and normal fundal uterine contour. (B) Hysterosalpingogram confirmed bilateral tubal patency suggesting low likelihood the associated mass represented a rudimentary uterine horn with theoretical possibility of an atypical non-communicating rudimentary horn.

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Pathology and Follow-Up

Once removed, the mass was cut in half revealing a cavity of brown fluid. Microscopic examination revealed endometrial glands and stroma with surrounding smooth muscle proliferation consistent with ACUM (Fig. 3B). Her postoperative course was uncomplicated, and the patient reported complete resolution of her pain. Four months later, the patient spontaneously conceived. Her pregnancy was complicated by dietcontrolled gestational diabetes. She presented in spontaneous labor at 38 weeks gestation with an uneventful spontaneous vaginal delivery and postpartum course. The patient has also had an additional pregnancy by spontaneous conception with successful vaginal delivery.

Results of Literature Review

During review of the literature, a total of 154 unique articles were identified on the basis of the above-mentioned search criteria. After application of ACUM inclusion criteria, 34 papers were individually reviewed, consisting of 70 reported cases applied for analysis (Supplemental Fig. 1, available online).

The average patient age was 24.9 (\pm 9.3) years and most common presenting complaints were dysmenorrhea (81.4%), chronic pelvic or abdominal pain (54.1%), and pain refractory to analgesics or oral contraceptive pills (34.3%). ACUM was associated with endometriosis (10.0%), adenomyosis (5.7%), and infertility (4.3%). Infertility was mentioned within 2 case reports. Similar to the current case, one of the patients successfully conceived after surgical removal. The other patient had a 5-year history of primary infertility but her fertility status after resection was not reported. Another report described an extrauterine pregnancy within an ACUM with associated spontaneous miscarriage, but ultimately required surgical removal (Table 1).

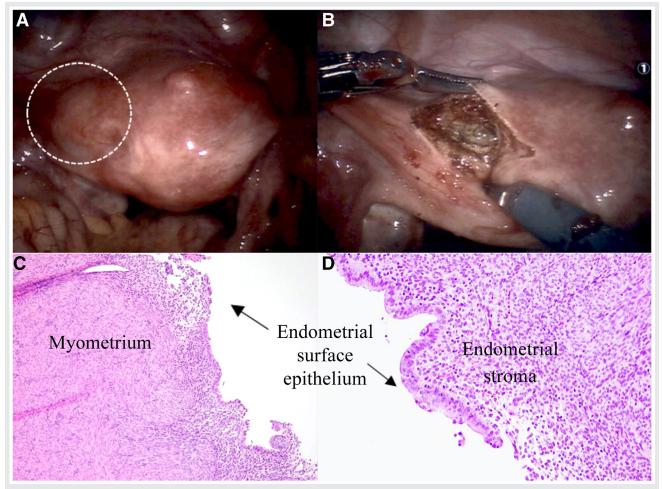
ACUM was identified at distant extrauterine sites (4.3%), near the adnexa (7.1%), in the broad ligament (15.7%), and located anterior intrauterine (2.9%), posterior intrauterine (1.4%), right intrauterine (28.6%), and left intrauterine (38.6%). The average mass size was 3.9 (\pm 2.0) cm. ACUM evaluation by imaging studies (Table 1) consisted of MRI (62.9%), transvaginal ultrasound (55.7%), transabdominal ultrasound (21.4%), transrectal ultrasound (8.6%), and computed tomography (7.1%). Many studies included dual imaging modalities but few studies reported the use of 3D transvaginal ultrasound.

Surprisingly, MRI findings of a hyperenhancing mass in T1-weighted imaging was more prevalent than T2 (odds ratio 8.0, 69.4% vs. 22.2%, respectively; P < .001). Ultrasound findings identified a hypoechoic mass more often (63.2%) than a hyperechoic mass (36.8%), but this difference was not statistically significant (P=.194). Of described cases, patients were statistically more likely to have undergone laparoscopy (75.7%) than laparotomy (18.6%) or more invasive surgery such as hysterectomy (5.7%) (P<.001). Of the 77.1% of cases that reported outcomes, 90.7% had complete relief of symptoms after definitive management (Table 1).

DISCUSSION

We report a unique case of delayed ACUM diagnosis in a patient experiencing chronic pelvic pain and infertility and summarize the current literature, which remains relatively scant in defining diagnostic criteria and management of ACUM. In the current patient, delayed diagnosis might be attributed to the more common alternative diagnoses: degenerating or atypical leiomyoma, endometriosis, and

FIGURE 3



Surgical and pathologic evaluation of accessory cavitated uterine mass. (A) Enlargement of left broad ligament of the uterus (dashed circle). (B) Incision along broad ligament revealing accessory cavitated uterine mass. (C) Histologic examination of removed mass demonstrated endometrial luminal and glandular epithelium with stroma surrounded by proliferative myometrium. (D) Higher powered histologic image demonstrating endometrial luminal epithelium and stroma.

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non-communicating rudimentary horn. Many of these diagnoses share common symptoms with ACUM such as dysmenorrhea and pain resistant to medical therapy, but closer scrutiny of imaging studies and a high level of suspicion led to the ultimate diagnosis and treatment of ACUM. Early consideration may lead to a timelier diagnosis, treatment, and allocation of medical resources and prevent symptom-free years lost to ineffective treatment measures. On the basis of literature review, endometriosis has a relatively high prevalence (10.0%), whereas other pathologies such as adenomyosis, fibroids, and Müllerian anomalies were relatively infrequent.

Etiologies

The 2 hypotheses regarding the emergence of ACUM are an isolated Müllerian duct malformation or dysfunction of the female gubernaculum (3). Müllerian remnants that may lead to the development of ACUM could develop in a similar

matter to rudimentary uterine mass or appendix (3). However, many cases were previously reported ACUM located at the insertion area or pathway of the round ligament from the uterine horn. Therefore, it was inferred that there may be some relationship with the attachment of the female gubernaculum and its dysfunction during embryologic development (3).

Recently, controversy has emerged regarding the inclusion of JCA as an entity of ACUM. Previously, JCA has been defined as a cystic lesion >1 cm in diameter; independent of the uterine cavity; covered by hypertrophic myometrium as indicated on radiologic imaging; associated with severe dysmenorrhea; and age <30 years (9). Previous studies conclude that JCA is a rare variant of adenomyosis rather than a congenital abnormality with distinguishing criteria from ACUM including the presence of denser areas of adenomyosis surrounding the cystic area (9, 10). However, other publications suggest that JCA in young women should be

TABLE 1

Patient demographics, imaging modalities, and associated findings with diagnosis of accessory cavitated uterine mass on the basis of literature review

Demographics	% (n/total)	Demographics	% (n/total)
Presenting complaint		Location	
Dysmenorrhea	81.4% (57/70)	Left intrauterine	38.6% (27/70)
Chronic pelvic or abdominal pain	54.1% (36/70)	Right intrauterine	28.6% (20/70)
Refractory to medications	34.3% (24/70)	Broad ligament	15.7% (11/70)
Cyclic pelvic pain	15.7% (11/70)	Adnexal	7.1% (5/70)
Other symptoms	12.9% (9/70)	Distant extrauterine	4.3% (3/70)
Additional findings	`````	Anterior intrauterine	2.9% (2/70)
Endometriosis	10.0% (7/70)	Posterior intrauterine	1.4% (1/70)
Adenomyosis	5.7% (4/70)	Imaging modality	
Infertility	4.3% (3/70)	MRI	62.9% (44/70)
Fibroids	1.4% (1/70)	TVUS	55.7% (39/70)
Uterine Anomalies	0.0% (0/70)	TAUS	21.4% (15/70)
Management		TRUS	8.6% (6/70)
Laparoscopy	75.7% (53/70)	HSG	8.6% (6/70)
Laparotomy	18.6% (13/70)	СТ	7.1% (5/70)
Invasive surgery	5.7% (4/70)	MRI findings	
Outcome		Hyperenhancing mass T1	69.4% (25/36)
Complete relief of symptoms	70.0% (49/70)	Hyperenhancing mass T2	22.2% (8/36)
Not reported	22.9% (16/70)	Hypoenhancing mass T2	8.3% (3/36)
Partial relief of symptoms	7.1% (5/70)	Ultrasound findings	
		Hypoechoic mass	63.2% (12/19)
		Hyperechoic mass	36.8% (7/19)
CT = computed tomography; HSG = hysterosalpingography; MRI = magnetic resonance imaging; TAUS = transabdominal ultrasound; TRUS = transrectal ultrasound; TVUS = transvaginal ultrasound.			

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grouped with ACUM to avoid nosological framework overlap and that adenomyosis surrounding may result from increased intra-cystic menstrual pressure (2, 7, 9). In support of this argument, both identified entities have similar outcomes after surgical treatment (9, 10).

In our literature review, ACUM were commonly identified in the left intrauterine region (38.6%), right intrauterine region (28.6%), and within the broad ligament (15.7%). There were also 3 reported cases (4.3%) meeting the criteria for ACUM that had extrauterine locations. The extrauterine locations included the left uterosacral ligament adherent to the sigmoid colon, the mesoappendix, and the left lower lumbar region near the jejunum and descending colon (11–13). Identification of ACUM in an extrauterine location makes gubernaculum dysfunction less likely as the etiology and Müllerian duct malformation or remnant more reasonable. In one of these cases, ACUM developed adjacent to bowel approximately 6 years after laparoscopic hysterectomy with morcellation, suggesting local seeding of uterine tissue during morcellation as an iatrogenic etiology (13).

Interestingly, 2 cases reported ACUM in association with malignancy. One case reported a small ACUM appearing as a cystic mass with normal uterine epithelium and myometrium in association with a moderately differentiated endometrioid ovarian carcinoma (14). In this case, ovarian stromal metaplasia was hypothesized to result in development of an ACUM from the developing malignancy. Another case reported associated clear cell carcinoma within a uterine-like mass in the right retroperitoneal space (15). Pathogenesis of clear cell carcinoma within an ACUM likely reflects similar mechanisms associated with clear cell carcinoma-associated with endometriosis (16). These cases raise a clinical concern that ACUM may develop as a reactive process to an underlying malignancy or may have inherent malignant potential because of their histologic origin. As a result, prudent diagnosis and surgical excision are of the utmost importance.

Imaging Findings

In the current case, 3D SIS clearly differentiated ACUM from other causes of chronic pelvic pain and dysmenorrhea. This methodology demonstrated the ACUM as a distinct mass within the myometrium, independent of the endometrial cavity that was otherwise normal in contour. Imaging criteria for ACUM are focused on the detection of functional endometrium requiring a skilled sonographer (3). Cycle-dependent changes of ACUM are evident as represented by progression from trilaminar to a hyperechoic appearance similar to the changes in the endometrium during progression from the proliferative to secretory phase (17). The patient's cycle day should be noted and correlated with ultrasound findings. Color Doppler flow may also show cycle-dependent changes in vascularity as highlighted in Figure 1A and B. Furthermore, additional imaging modalities may be used to differentiate ACUM from other etiologies. For example, HSG demonstrating bilateral tubal fill and spill assisted in distinguishing between ACUM and a non-communicating rudimentary horn. In the literature, 8.6% of patients with ACUM had a reported HSG. In the literature, MRI hyperenhancement on T1 was reported significantly more than hyperenhancement on T2. On ultrasound, hypoechoic masses were more commonly reported (63.2%) than hyperechoic masses (36.8%), but this was not statistically significant. This is likely because of the functional nature of the endometrium in the ACUM leading to cyclic changes in the ultrasound findings.

The reported imaging findings may present in the setting of etiologies other than ACUM. The most important distinguishing factor of an ACUM is the identification of functional endometrium within the mass. The cyclic changes identified within the mass on transvaginal ultrasound suggested functional endometrium. In addition, increased Doppler flow and hypoechoic internal contents were noted during acute pain episodes. SIS highlighted ultrasonographic features of the mass along with the capacity to evaluate the mass in relation to the endometrial cavity, which otherwise appeared normal in contour making rudimentary horn less likely. Although leiomyoma may also contain Doppler flow, they are typically solid in appearance and do not present as a fluid-filled cysts. Focal adenomyosis may appear as a myometrial cyst but typically additional ultrasonographic features of adenomyosis may be present such as globular uterus, irregular, junctional zone, and asymmetric thickening (18). Although endometriomas may display similar internal contents as an ACUM, they are extrauterine within ovaries.

ACUM and Fertility

This case demonstrates a unique association between ACUM and infertility, which is not well established, and based on literature review, only 2 other reported cases described with infertility. Our case and one additional prior case describe unassisted conception and resolution of symptoms after surgical resection of an ACUM, indicating a possibly significant association between ACUM and infertility (19). Another report described an extrauterine (ectopic) pregnancy and subsequent miscarriage within an ACUM ultimately requiring surgical removal (20). An additional case report described a patient identified with ACUM after a 5-year history of primary infertility. However, her fertility status after ACUM resection was not reported (21). Regarding pathophysiology, the effect of an ACUM on fertility has not been established. Adenomyosis and endometriosis are associated with a reduction in likelihood of pregnancy through in vitro fertilization (2, 22, 23); therefore, it is reasonable for a similar disease process, such as ACUM, to also have a negative impact on fertility resulting from abnormal endometrial function or promoting a proinflammatory environment preventing embryo implantation. Additional research is needed to determine if a relationship with infertility exists and explore the pathophysiologic mechanisms.

Management, Safe Delivery, and Multidisciplinary Collaboration

After definitive diagnosis, treatment options should be approached with shared decision making. A majority (75.7%) of cases in the literature underwent laparoscopic excision. To preserve fertility in young people, resection of the ACUM is preferred vs. total hysterectomy. With fertility-sparing

techniques, there is an increased risk of future pregnancy and perinatal complications including uterine rupture (24). In the setting of myomectomies, the risk of uterine rupture is greater in laparoscopic myomectomies compared with laparotomies (24). In the present case, fertility was preserved after uterine-sparing robotic-assisted laparoscopic excision of the para-uterine mass. This patient's complex presentation warranted a multidisciplinary approach at our institution. The cyclic changes of the endometrium within the ACUM were identified by the patient's Reproductive Endocrinology and Infertility specialist. A Maternal Fetal Medicine specialist with experience in evaluation of Müllerian anomalies further characterized and identified the mass using SIS with 3D reconstruction. Finally, her surgery was performed by a Minimally Invasive Gynecologic Surgeon. Her pregnancy care transitioned to her primary Obstetrician-Gynecologist with an overall uncomplicated pregnancy and delivery. A multidisciplinary team approach proved vital in the favorable outcome for our patient. This case adds to the paucity of evidence that vaginal delivery after ACUM excision without myometrial entry may be attempted safely, which has only been described once previously in the literature (19).

CONCLUSION

This case highlights key points regarding evaluation and management of ACUM. Our patient experienced a significant delay in diagnosis with symptoms and imaging findings attributed to more common etiologies. As a result of this delay, the patient experienced significantly reduced quality of life. In addition, she received unnecessary medical interventions (hormonal contraception, lupron) for her symptoms, visited the emergency room on multiple occasions for pain, and underwent several diagnostic laparoscopies that could have been avoided with an earlier suspicion for ACUM. Prompt pain relief is expected after excision. Of the 77.1% of published cases that reported outcomes, 90.7% had complete relief of symptoms after definitive management. In addition, excision was associated with spontaneous conception after 7 years of infertility. Making the diagnosis of ACUM is important to coordinate surgical excision as medical treatments are likely to only provide temporary relief while surgery is definitive. If mistaken for adenomyosis or endometriosis, patients may receive unnecessary medical therapies with significant side effects associated with hormonal blockade and often further reduction of fertility with an ultimate delay in what could be definitive care.

This case demonstrates the utility of each imaging study performed, with key architectural features of the ACUM dynamically present depending on menstrual cycle phase and SIS demonstrating close association but independence of the ACUM from the endometrial cavity. A strength of this literature review is the comprehensive nature using distinct criteria for ACUM diagnosis. However, this review was limited by reporting within the reviewed case reports and series, where many articles did not report patient outcomes. Ultimately, ACUM should be considered in the differential of patients with chronic, cyclic pelvic pain, dysmenorrhea, and infertility. With expertise from multiple specialties, successful excision may not only resolve symptoms but also improve fertility and allow for uncomplicated delivery.

Please see supplemental document for the patient's perspective.

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REFERENCES

- 1. Oliver J. An accessory uterus distended with menstrual fluid enucleated from the substance of the right broad ligament. Lancet 1912;179:1609.
- Peyron N, Jacquemier E, Charlot M, Devouassoux M, Raudrant D, Golfier F, et al. Accessory cavitated uterine mass: MRI features and surgical correlations of a rare but under-recognised entity. Eur Radiol 2019;29:1144–52.
- Acien P, Acien M, Fernandez F, Jose Mayol M, Aranda I. The cavitated accessory uterine mass: a Mullerian anomaly in women with an otherwise normal uterus. Obstet Gynecol 2010;116:1101–9.
- Saravelos SH, Cocksedge KA, Li TC. Prevalence and diagnosis of congenital uterine anomalies in women with reproductive failure: a critical appraisal. Hum Reprod Update 2008;14:415–29.
- Acien P, Bataller A, Fernandez F, Acien MI, Rodriguez JM, Mayol MJ. New cases of accessory and cavitated uterine masses (ACUM): a significant cause of severe dysmenorrhea and recurrent pelvic pain in young women. Hum Reprod 2012;27:683–94.
- Jayasinghe Y, Rane A, Stalewski H, Grover S. The presentation and early diagnosis of the rudimentary uterine horn. Obstet Gynecol 2005;105: 1456–67.
- Acien P, Acien M. Accessory and cavitated uterine mass versus juvenile cystic adenomyoma. F S Rep 2021;2:357–8.
- Brosens I, Gordts S, Habiba M, Benagiano G. Uterine cystic adenomyosis: a disease of younger women. J Pediatr Adolesc Gynecol 2015;28:420–6.
- Arya S, Burks HR. Juvenile cystic adenomyoma, a rare diagnostic challenge: case reports and literature review. F S Rep 2021;2:166–71.

- Martin DC, Koninckx PR. Juvenile cystic adenomyomas: acquired adenomyosis variant or congenital Mullerian defects? F S Rep 2021;2:145.
- 11. Kim HC, Yang DM, Kim SW, Kim GY, Choi SI, Park SJ. Uterus-like mass involving the appendix: US and CT findings. J Clin Ultrasound 2012;40: 518–21.
- Matsuzaki S, Murakami T, Sato S, Moriya T, Sasano H, Yajima A. Endomyometriosis arising in the uterosacral ligament: a case report including a literature review and immunohistochemical analysis. Pathol Int 2000; 50:493–6.
- Bakshi N, Dhawan S. Extrapelvic "Uterus like mass" following laparoscopic morcellation hysterectomy – a consequence of iatrogenic implantation? Int J Surg Pathol 2023;31:791–4.
- Rahilly MA, al-Nafussi A. Uterus-like mass of the ovary associated with endometrioid carcinoma. Histopathology 1991;18:549–51.
- 15. Nakakita B, Abiko K, Mikami Y, Kido A, Baba T, Yoshioka Y, et al. Clear cell carcinoma arising from a uterus-like mass. Pathol Int 2014;64:576–80.
- Kvaskoff M, Mahamat-Saleh Y, Farland LV, Shigesi N, Terry KL, Harris HR, et al. Endometriosis and cancer: a systematic review and meta-analysis. Hum Reprod Update 2021;27:393–420.
- Reed BG, Carr BR. The normal menstrual cycle and the control of ovulation. In: Feingold KR, Anawalt B, Blackman MR, Boyce A, Chrousos G, Corpas E, et al., editors. Endotext; 2000, South Dartmouth (MA).
- Van den Bosch T, de Bruijn AM, de Leeuw RA, Dueholm M, Exacoustos C, Valentin L, et al. Sonographic classification and reporting system for diagnosing adenomyosis. Ultrasound Obstet Gynecol 2019; 53:576–82.
- Supermaniam S, Thye WL. Diagnosis and laparoscopic excision of accessory cavitated uterine mass in young women: two case reports. Case Rep Womens Health 2020;26:e00187.
- Alkhateeb HM, Yaseen EM. Twin pregnancy in an accessory cavitated noncommunicating uterus. Int J Surg Case Rep 2015;10:45–8.
- Mondal R, Bhave P. Accessory cavitated uterine malformation: enhancing awareness about this unexplored perpetrator of dysmenorrhea. Int J Gynaecol Obstet 2023;162:409–32.
- Bulletti C, Coccia ME, Battistoni S, Borini A. Endometriosis and infertility. J Assist Reprod Genet 2010;27:441–7.
- Macer ML, Taylor HS. Endometriosis and infertility: a review of the pathogenesis and treatment of endometriosis-associated infertility. Obstet Gynecol Clin North Am 2012;39:535–49.
- 24. Gil Y, Badeghiesh A, Suarthana E, Mansour F, Capmas P, Volodarsky-Perel A, et al. Risk of uterine rupture after myomectomy by laparoscopy or laparotomy. J Gynecol Obstet Hum Reprod 2020;49:101843.