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Case Report

Accessory Cavitated Uterine Mass: A Diagnostic Dilemma Illustrated by Three Case Reports

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Abstract

Background: Accessory cavitated uterine mass (ACUM) is a rare, unclassified Müllerian anomaly characterized by distinct imaging features. It is typically located within the uterus, close to the round ligament, and has a uterus-like structural arrangement. The patient may present with pelvic pain or dysmenorrhea. Most of these cases are misdiagnosed because of a lack of awareness about this unusual entity.

Case Presentation: Three cases of ACUM in young patients who experienced prolonged symptoms and had incomplete family structures were reported in this paper. Initially, two of these cases were misdiagnosed during ultrasound examinations (USG). The subsequent magnetic resonance imaging (MRI) revealed characteristic imaging features consistent with ACUM, which provided significant psychological relief to both the patients and their families. Two patients received hormonal therapy, both of whom were unmarried. The third patient, however, indicated a wish to conceive and was therefore initiated on analgesics. All three patients chose to forgo surgical intervention, opting instead for medical management despite its limited success in alleviating their symptoms. This decision was made to minimize obstetric risks associated with surgical interventions in potential future pregnancies.

Conclusion: Laparoscopy or open surgery is the mainstay treatment for a permanent relief from the symptoms. However, surgical treatment should be offered with caution as no data are available in medical literature regarding the effect of surgically induced myometrial scarring on patients' reproductive outcomes. Since most of these patients were young and nulligravida, the therapy had to be personalized in accordance with the patient's preference and family status.

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Introduction

ACUM is a rare uterine anomaly portrayed by the presence of an accessory cavity within the uterus that does not show a communication with the main uterine cavity and is lined by endometrium, which is further surrounded by uterine smooth muscle (1). ACUM is associated with a normal uterine cavity in contrast to other Müllerian anomalies, in which the uterus is malformed. The patient may present with chronic pelvic pain or dysmenorrhea. Although the diagnosis can be reliably achieved using transvaginal ultrasound (TVS) or transrectal ultrasound (TRUS), most cases are misdiagnosed on ultrasound, mainly due to a lack of awareness of this unusual Müllerian anomaly among clinicians. MRI is the most accurate imaging tool in diagnosing ACUM and also acts as a complementary exam to rule out adenomyosis or other Müllerian malformations.

Case Presentation

Three case reports of patients diagnosed with ACUM in our institution from July 2023 to April 2024 were presented. Informed consent was obtained from each patient for the publication of radiological imaging. All three patients were symptomatic for a variable duration, and two were misdiagnosed on their initial ultrasound. Pelvic ultrasound was performed as an initial investigation, followed by MRI in all three patients for confirmation of the diagnosis and for better delineation of the uterine and pelvic anatomy. None of our patients had associated urinary tract or vertebral anomalies.

Case 1: A 21-year-old, unmarried nulligravida presented with a one-year history of mild intermittent left iliac fossa pain, which worsened in severity over the last three months. There was also a history of intermittent foul-smelling vaginal discharge for four years. She had attained menarche at the age of 14, and her menstrual cycles were normal. The general physical and abdominal examinations were normal. On ultrasound of the pelvis (Figure 1A), the possibility of a Robert's uterus (asymmetric septate uterus, wherein an oblique septum asymmetrically divides the uterine cavity) was considered, and an MRI was advised. On MRI (Figure 1B), a well-circumscribed mass

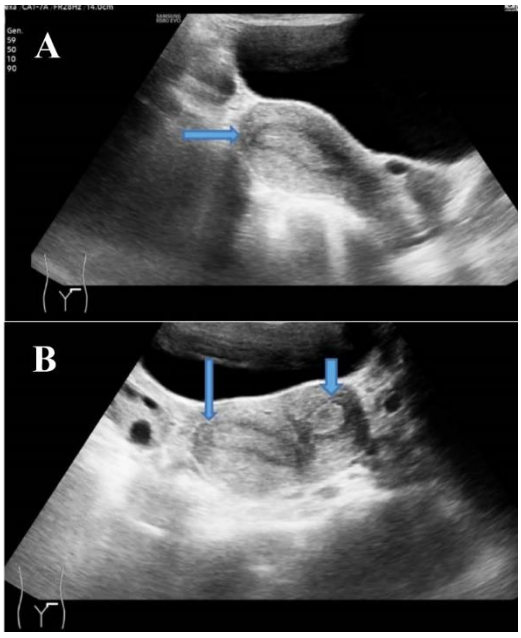

Figure 1A. A 21-year-old nulligravida presenting with pain in the left iliac fossa. USG (A) sagittal image shows a normal uterus (arrow) and (B) axial image shows a normal uterus with echogenic endometrium (long arrow) and ACUM in the left lateral wall of the uterus (short arrow)

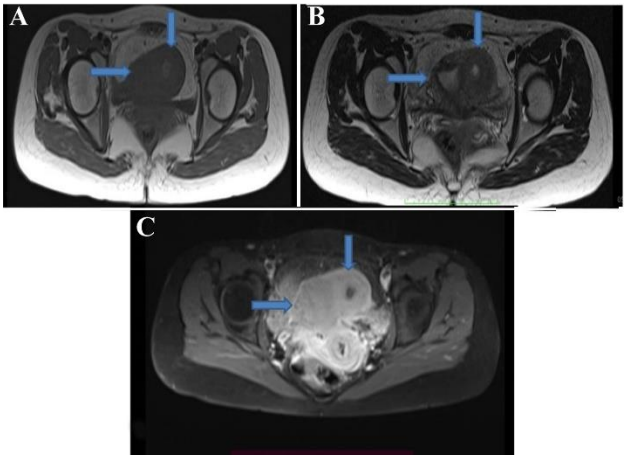

Figure 1B. MRI axial images: (A) T1-weighted, (B) T2-weighted, and (C) post -contrast T1 fat-saturated sequences show the uterus (horizontal arrow) and ACUM (vertical arrow). The lesion shows hyperintense contents on both T1- and T2-weighted images, consistent with blood products

was seen within the left wall of the uterus. Imaging revealed a round cystic area demonstrating hyperintense signal on T1-weighted, fat-saturated T1-weighted, and T2-weighted sequences, indi cating hemorrhagic contents. This cavity showed no communication with the main uterine cavity and was lined by endometrium, surrounded by a hypointense junctional zone on T2-weighted imaging. Bilateral ovaries and fallopian tubes were normal. The left fallopian tube showed normal insertion into the left uterine cornu. With these typical MRI imaging findings, a diagnosis of ACUM was confirmed. The patient was prescribed oral contraceptive pills, and after a six-month follow-up, she was demonstrating a positive response to the treatment. Notably, she experienced the onset of dysmenorrhea in the absence of the medication.

Case 2: A 20-year-old, unmarried nulligravida presented with mild chronic pelvic pain and severe dysmenorrhea two years after the onset of menarche, which occurred at the age of 13. She was initially treated with analgesics, which were not effective in relieving the pain. She was subse quently shifted to oral contraceptive pills for the past year, resulting in partial symptom relief. There was no history of vaginal discharge. Ultrasound was performed, revealing a well-defined mass embedded in the uterine myometrium near the left cornu, showing peripheral vascular flow. This mass had a central fluid-filled cavity and an echogenic stripe lining it, corresponding to the endometrium, which was further surrounded by

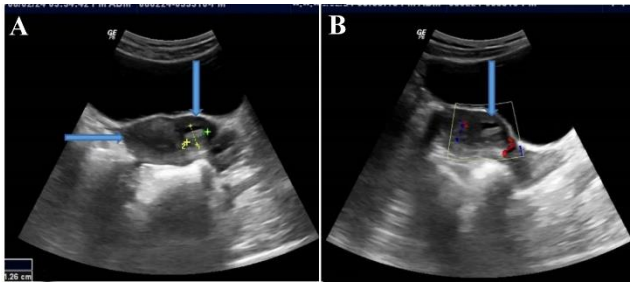

Figure 2A. A 20-year-old nulligravida presenting with chronic pelvic pain and dysmenorrhea. USG (A) axial image shows normal uterine cavity (horizontal arrow) and ACUM in the left lateral wall of the uterus, showing fluid in the accessory cavity (vertical arrow) and (B) left parasagittal image shows ACUM and peripheral vascular flow

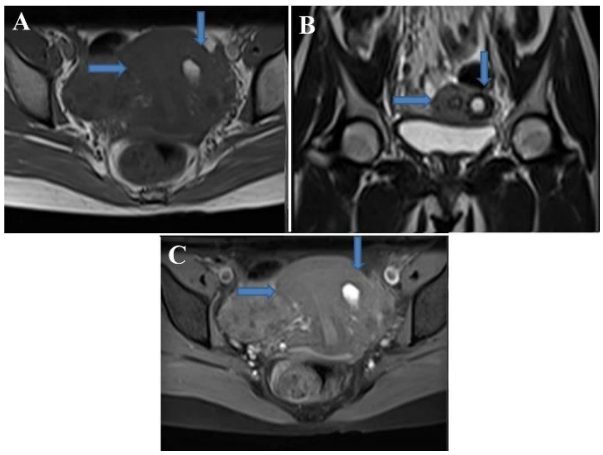

Figure 2B. MRI images: (A) axial T1-weighted image shows uterus (horizontal arrow) and hyperintense cavity of ACUM (vertical arrow) suggestive of blood products, (B) coronal T2-weighted image shows uterus (horizontal arrow) and ACUM (vertical arrow) with a hyperintense signal in accessory cavity, and (C) axial post-contrast T1 fat-saturated image shows normal enhancement of uterine myometrium (horizontal arrow) and normally enhancing myometrium surrounding cavity of ACUM (vertical arrow)

soft tissue with an echotexture similar to that of the myometrium. Based on the findings, ACUM was proposed as the diagnosis (Figure 2A). An MRI of the pelvis was then performed to confirm the diagnosis and to rule out other associated uterine pathologies (Figure 2B). The patient continued to use oral contraceptive pills; however, the response had not been entirely satisfactory. Nevertheless, the patient expressed a desire to avoid surgical intervention.

Case 3: A 32 -year- old, para 1, married woman presented with recurrent left lower abdominal pain for the past three years. The patient had already been investigated for a similar complaint and had been misdiagnosed twice on an ultrasound performed in a remote area. Initially, she was erroneously diagnosed with a heterotopic pregnancy based on a first-trimester ultrasound that demonstrated a single intrauterine gestational sac alongside an additional sac in the left adnexa. Subsequently, she received a second misdiagnosis of chronic left cornual ectopic pregnancy. There was no past history of surgery. Urine analysis for pregnancy yielded a negative result. Finally, she underwent an ultrasound (Figure 3A) and MRI (Figure 3B) in our institution, which diagnosed her as a case of ACUM. In her desire to conceive

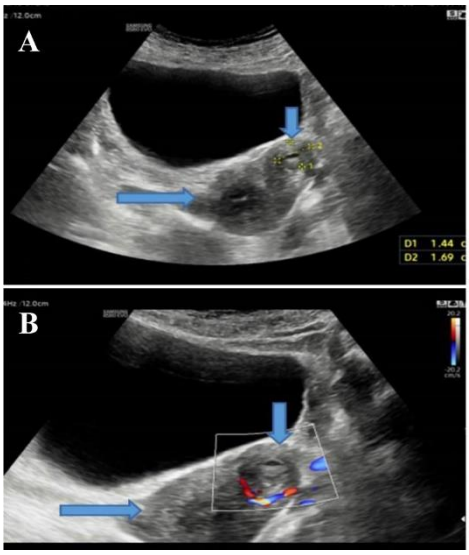

Figure 3A. A 32 -year- old multiparous presenting with recurrent left lower abdominal pain. (A) Axial grayscale image and (B) color Doppler image show the uterus (horizontal arrow) and ACUM (vertical arrow) with peripheral vascular flow in the left lateral wall of the uterus

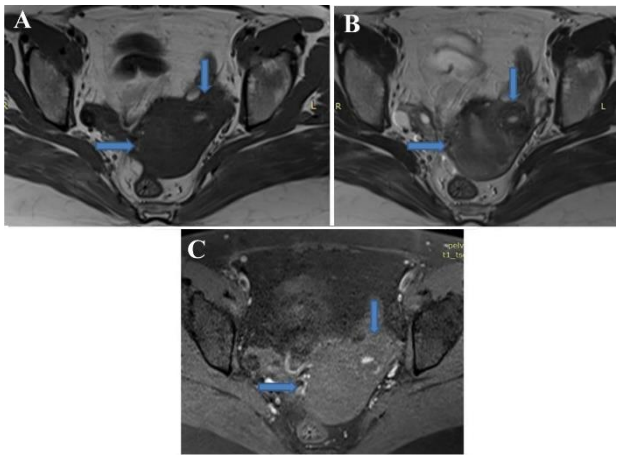

Figure 3B. MRI axial images: (A) T1-weighted; (B) T2-weighted; and (C) post- contrast T1 fat-saturated images show uterus (horizontal arrow) and ACUM (vertical arrow) in left lateral uterine wall, having hyperintense signals on both T1- and T2-weighted images, consistent with hemorrhagic contents

a second child, she opted for analgesics to alleviate her pain instead of pursuing hormonal therapy. After a six-month follow-up, she had demonstrated symptomatic improvement; however, conception had not yet occurred.

None of our patients had associated adenomyosis. All three of our patients refused surgical treatment at this point in time, as the long-term surgical effects on the patient's reproductive life are still not known, and it may be an obstetric risk to the patient. They agreed to proceed with medical therapy, even though it brought only a temporary relief to the symptoms.

Discussion

ACUM is a well-circumscribed uterine lesion characterized by a cystic cavity containing blood products, lined by normal endometrial epithelium and surrounded by myometrium, without communication with the normal uterine cavity. It protrudes from a normally shaped and functional uterus, most commonly localized on the lateral uterine wall, caudal to the round ligament insertion (2). It is considered as a rare Müllerian anomaly that is yet to be fully explored and is currently classified as U6 under the European Society of Human Reproduction and Embryology (ESHRE), and the European Society for Gynecological Endoscopy (ESGE) classification system for congenital anomalies of the female genital tract. According to the American Society for Reproductive Medicine Müllerian Anomalies Classification (ASRM MAC 2021), it is considered a variant (3, 4). Although unclear, there are two hypotheses regarding its etiopathogenesis. First, it is postulated that gubernaculum dysfunction during embryogenesis may be responsible for the duplication or persistence of paramesonephric tissue, leading to the development of accessory uterine tissue (4, 5). Hence, it is considered a developmental uterine anomaly with a typical uterus-like structural arrangement (6). The second hypothesis suggests that duplication or persistence of the Müllerian duct tissue may be associated with the development of round ligament insufficiency. Müllerian remnants that may give rise to ACUM are thought to develop similarly to rudimentary uterine horns or uterine appendages. When anomalous Müllerian ducts are present, they typically participate in normal fusion and resorption processes in coordination with their anatomically symmetrical contralateral Müllerian duct counterparts. However, the endometrial tissue fails to undergo resorption, resulting in the formation of isolated, non-communicating cavities. This leads to the emergence of a new Müllerian duct malformation, characterized as ACUM (7, 8).

It characteristically presents in nulligravida and at a younger age, usually less than 30 years. One of our patients experienced dysmenorrhea that started soon after menarche, along with chronic pelvic pain that continued throughout the menstrual cycle, caused by the pressure effect of an enlarged accessory uterine cavity. This distension results from cyclic hemorrhage of the functional endometrial lining (6, 9). The pain, similar to that in endometriosis, is usually refractory to medical treatment (4). Prior to 2012, when Acien et al. established diagnostic criteria and introduced the term ACUM, this anomaly was variably described in literature as juvenile cystic adenomyoma, isolated adenomyoma, or cavitated adenomyoma. The diagnostic criteria for ACUM, as established by Acien et al., comprise: (1) an accessory cavitated uterine mass exhibiting uterus-like tissue architecture; (2) absence of communication with the uterine cavity; (3) coexistence with normal uterine anatomy, bilateral fallopian tubes, and ovaries; and (4) no histological evidence of adenomyosis (10, 11). This can be confirmed through histopathological examination or imaging. Histopathological examination of the excised ACUM specimen revealed a uterus-like structure with a central cavity lined by endometrium containing hemorrhagic material, surrounded by myometrial tissue comparable to that of a normal uterus. Microscopically, the cavity of the ACUM was lined by an endometrial gland and stroma. Histologically, these lesions demonstrate immunohistochemical staining patterns identical to normal endometrium, exhibiting strong positivity for CD10, estrogen receptors (ER), and progesterone receptors (PR) (12).

Even with typical imaging features, it remains a diagnostic dilemma, and most cases are misdiagnosed on ultrasound, mainly due to limited knowledge about this seemingly rare entity with an unknown incidence. Ultrasound is the first-line imaging modality for ACUM, which shows a non-communicating, small cystic cavity with homogeneous echoes. This cavity is lined by an echogenic endometrial layer with variable thickness depending on the phase of the menstrual cycle, and surrounded by tissue having an echotexture similar to that of the myometrium. The location is typically

near the uterine cornu and inferior to the insertion of the round ligament (9). The rest of the uterus, including the ovaries and fallopian tubes, appear normal, as better depicted on 3D/4D ultrasound. This advanced imaging modality aids in excluding other Müllerian anomalies and visualizes the interstitial portion of the fallopian tube, which helps to locate these lesions as they present caudal to the round ligament at the tubal insertion site (12). MRI offers advantages over ultrasonography by providing precise lesion localization, detailed morphological assessment, and accurate tissue characterization. MRI also has the advantage of providing a comprehensive evaluation of the pelvic cavity. The presence of hyperintense signal on T1-weighted, fat-saturated T1-weighted, and T2-weighted sequences reflects a hemorrhagic content within the accessory cavity. A fluid-fluid level may also be observed within the cavity, corresponding to bleeding contents of different ages. The surrounding tissue's hypointense signal on T2-weighted imaging corresponds to junctional zone myometrium (4). Saline-infused ultrasound or contrast-enhanced hysterosalpingography is no longer required, as MRI can precisely locate the ipsilateral fallopian tube insertion and normal uterine cornu (3, 9).

The primary differential diagnosis was Robert's uterus (U4 class-a cavitated rudimentary uterine horn), which was initially misclassified in our first case. Robert's uterus represents a rare variant of the asymmetric septate uterus, characterized as a resorption defect wherein an oblique septum asymmetrically divides the uterine cavity. The uterine isthmus was well-developed; however, there was a noted aplasia of the ipsilateral cervix. The diagnostic triad of Robert's uterus comprises: a unicornuate uterus, a contralateral obstructed hemicavity often containing hematometra, and a normal uterine fundus that may exhibit a slight external indentation (13). The key element to differentiate was the normal insertion of the ipsilateral fallopian tube, as it never opens in ACUM. The presence of a normal uterine architecture in ACUM contrasts with the banana-shaped configuration of the developed horn in hemiuterus, providing key discriminatory features between these entities (6). Similarly, the functional rudimentary horn in Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome can be easily identified due to associated aplasia or hypoplasia of the uterus and other Müllerian duct derivatives. Cornual ectopic pregnancy, as seen in our third case, is located in the adnexa and not within the uterine myometrium; it typically lacks an endometrial lining surrounding the cavity. Focal cystic adenomyosis, another differential diagnosis for ACUM, typically occurs in multiparous women aged 35–40 years, appears ill-defined on imaging, and is situated outside the junctional zone. It has no significant mass effect on the endometrial cavity and lacks the typical endometrial lining. The rest of the uterus may exhibit features of focal or diffuse adenomyosis (14). Uterine leiomyomas exhibiting cystic or hemorrhagic degeneration may also mimic ACUM on imaging. However, the larger size, multiplicity of lesions, and mass effect on the endometrial cavity usually help differentiate it from ACUM (9, 15). A definitive diagnosis of ACUM may require laparoscopy and histopathology to confirm the presence of endometrial lining, a hallmark feature.

Given the rarity of the pathology, there is a lack of standardized treatment strategies and long-term follow-up data. The management of this condition may involve conservative approaches, primarily utilizing medical therapy, which was the preferred choice among all three patients in our study. Analgesics, such as non-steroidal anti-inflammatory drugs (NSAIDs), along with hormonal therapies, including oral contraceptive pills, progesterone-only pills, the levonorgestrel-releasing intrauterine system (Mirena), and gonadotropin-releasing hormone agonists are viable options. The continuous implementation of hormone suppressive therapies impedes ovarian steroidogenesis, thereby preventing the shedding of the endometrium. This mechanism prevents blood accumulation within the accessory uterine cavity, thereby avoiding cavity distension as the primary source of pain in this condition (12). However, it provides only temporary and partial relief from the symptoms. Moreover, the hormonal therapy cannot be given to patients who want to conceive, as was observed in our third case. The alternative therapeutic approach involves complete surgical excision of the lesion. Minimally invasive laparoscopic surgery or open surgery (14, 16) are common approaches. While intraoperative ultrasound improves myometrial lesion localization and reduces uterine trauma, it risks iatrogenic scarring that may adversely affect fertility (10). A more recent management approach involves TVS-guided alcohol sclerotherapy, which can be performed under local anesthesia. However, this technique remains in its early developmental stages (3, 4). The rationale

behind this treatment is the destruction of the functional endometrium, thereby preventing the monthly accumulation of menstrual blood. In addition, the use of lauromacrogol, which has been used for the sclerotherapy of hepatic cysts, was recently introduced for the sclerotherapy of ACUM, functioning as a local anaesthetic. Ultrasound-guided drainage, in conjunction with hormonal suppression of the menstrual cycle to mitigate further bleeding, may provide symptomatic relief. However, this approach appears to be a temporary intervention, as the ectopic endometrium continues to maintain its functional activity (17).

Hence, it is imperative to establish a customized treatment plan that corresponds with the patient's clinical condition. Due to the uncertain reproductive outcomes following surgical intervention, evaluating the effects on maternal and fetal health remains a significant challenge. Therefore, medical treatment may be recommended as the primary line of therapy, even if it yields only partial symptom relief and aligns with the patient's preferences. Alternatively, minimally invasive surgical options may be considered with the expectation that they will not adversely influence future pregnancies. The uncertain prognosis associated with this condition makes it vital for clinicians to engage in thorough long-term monitoring of patients diagnosed with ACUM. Comprehensive research efforts, including population-based longitudinal observational studies and a deeper understanding of this entity, are essential for advancing knowledge in this area.

Conclusion

Ultrasound enables real-time imaging while MRI offers excellent soft tissue contrast and detailed anatomic visualization of the ACUM lesion and surrounding pelvic cavity. The combination of these complementary imaging modalities enhances the diagnostic accuracy of ACUM. An appropriate individualized management strategy should be opted for, taking into account the unforeseen effects of surgery on the patient's fertility and reproductive outcome.

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Ethics approval and consent to participate: The ethical approval was waived off by the institutional ethical committee 'Indira Gandhi Medical College and Hospital, Shimla, HP' as the index cases were de-identified. A verbal informed consent was taken from the patients.

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Conflict of Interest

The authors declare that they have no competing interests.

References

1. Acién P, Acién M, Fernández F, José Mayol M, Aranda I. The cavitated accessory uterine mass: a Mullerian anomaly in women with an otherwise normal uterus. Obstet Gynecol. 2010;116(5):1101-9.
2. Betzler N, Brunes M, Anfelter P, Wedlund L, Persson J, Epstein E. Sonografic features of accessory cavitated uterine mass (ACUM) successfully treated with robotic assisted laparoscopic surgery-a case report. Clin Obstet Gynecol Reprod Med. 2019;5(6):1-4.
3. Zajaczkowska W, Kapczuk K. Accessory cavitated uterine mass (ACUM) as a miniature uterine anomaly causing severe lateralized dysmenorrhea: case series. Ginekol Pol. 2023;94(11):907-12.
4. Dekkiche S, Dubruc E, Kanbar M, Feki A, Mueller M, Meuwly JY, et al. Accessory and cavitated uterine masses: a case series and review of the literature. Front Reprod Health. 2023;5:1197931.
5. Acién P, del Campo FS, Mayol MJ, Acién M. The female gubernaculum: role in the embryology and development of the genital tract and in the possible genesis of malformations. Eur J Obstet Gynecol Reprod Biol. 2011;159(2):426-32.
6. Jain N, Verma R. Imaging diagnosis of accessory and cavitated uterine mass, a rare mullerian anomaly. Indian J Radiol Imaging. 2014;24(2):178-81.
7. Strug M, Christmas A, Schoonover A, Romero VC, Cordoba M, Leary E, et al. Impact of an accessory cavitated uterine mass on fertility: case presentation and review of the literature. F S Rep. 2023;4(4):402-9.
8. Deng F, Liu K, Huang Y, Chen Q, Wang L, Xiao X, et al. Successful treatment of a rare giant accessory cavitated uterine mass: a case report. J Int Med Res. 2024;52(5):3000605241252238.
9. Putta T, John R, Simon B, Sathyakumar K, Chandramohan A, Eapen A. Imaging manifestations of accessory cavitated uterine mass-a rare mullerian anomaly. Indian J Radiol Imaging. 2021;31(3):545-50.
10. Hu Q, Guo C, Chen Q, Zhang W, Wang H, Wei W. ACUM, an easily underdiagnosed cause of dysmenorrhea-a case report. Front Med (Lausanne). 2024;11:1308299.

11. HaiJing He, XuZhi Liang, Jiangtao Fan et al. Accessory and cavitated uterine mass (ACUM) in an 18-year-old woman: a case report and literature review, 04 January 2022, PREPRINT (Version 1) available at Research Square [https://doi.org/10.21203/rs.3.rs-1120568/v1].
12. Mondal R, Bhave P. Accessory cavitated uterine malformation (ACUM): a hidden face of dysmenorrhea. Indian J Obstet Gynecol Res. 2022;9(4):452-6.
13. Mahey R, Cheluvaraju R, Kumari S, Kachhawa G, Kumari A, Rajput M, et al. Robert's uterus versus Juvenile cystic adenomyoma–diagnostic and therapeutic challenges–case report and review of literature. J Hum Reprod Sci. 2023;16(1):79-86.
14. Takeuchi H, Kitade M, Kikuchi I, Kumakiri J, Kuroda K, Jinushi M. Diagnosis, laparoscopic management, and histopathologic findings of juvenile cystic adenomyoma: a review of nine cases. Fertil Steril. 2010;94(3):862-8.
15. Murase E, Siegelman ES, Outwater EK, Perez-Jaffe LA, Tureck RW. Uterine leiomyomas: histopathologic features, MR imaging findings, differential diagnosis, and treatment. Radiographics. 1999;19(5):1179-97.
16. Kriplani A, Mahey R, Agarwal N, Bhatla N, Yadav R, Singh MK. Laparoscopic management of juvenile cystic adenomyoma: four cases. J Minim Invasive Gynecol. 2011;18(3):343-8.
17. Timmerman S, Stubbe L, Van den Bosch T, Van Schoubroeck D, Tellum T, Froyman W. Accessory cavitated uterine malformation (ACUM): a scoping review. Acta Obstet Gynecol Scand. 2024;103(6):1036-45.