

Imaging diagnosis of accessory and cavitated uterine mass, a rare mullerian anomaly

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Abstract

Accessory and Cavitated Uterine Mass (ACUM) is a rare form of developmental Mullerian anomaly seen in young females, which presents as chronic recurrent pelvic pain and severe dysmenorrhea. It is an accessory cavity lying within an otherwise normal uterus. It is lined by functional endometrium and surrounded by myometrium-like smooth muscle cells; hence, it bears striking macroscopic and microscopic resemblance to the uterus. Hysterosalpingography (HSG), Ultrasonography (USG), and Magnetic Resonance Imaging (MRI) form the mainstay of diagnostic imaging. The entity is often under diagnosed; therefore, a high index of suspicion combined with HSG and MRI imaging can help in making an accurate diagnosis.

Key words: Accessory and cavitated uterine mass; juvenile cystic adenomyoma; mullerian anomalies; unicornuate uterus; uterus-like mass

Introduction

Accessory and Cavitated Uterine Mass (ACUM) is a rare, newly recognized Mullerian anomaly. It is an accessory cavity lined by functional endometrium within an otherwise normal uterine cavity, in contrast to the other Mullerian anomalies in which the uterus is malformed. The cavitated mass is locally defined to myometrium (unlike diffuse adenomyosis), is encapsulated (unlike myoma), and bears uterus-like histological organization (unlike adenomyoma). The entity needs expertise to diagnose as it is a rare but treatable cause of severe dysmenorrhea and chronic pelvic pain in young females with a wide range of differential diagnosis. The entity can be easily picked up on initial routine pelvic USG. Magnetic resonance imaging (MRI) is highly accurate in making diagnosis.

Case Report

A 24-year-old unmarried female presented with severe dysmenorrhea and chronic pelvic pain since menarche, which aggravated 2 years prior to presentation. Her menstrual cycles were regular with normal flow. There was history of recurrent renal calculi. There was no history suggestive of pelvic inflammatory disease. She was treated with non-steroidal anti-inflammatory drugs (NSAIDs) earlier and with oral contraceptive pills (OCP) for the last few months. Her general physical and per abdomen examination was normal.

Previous serial USG reports revealed a 3.0 × 3.6 cm, solid right adnexal mass abutting the right ovary with likely ovarian origin. On repeat USG pelvis, a homogeneous isoechoic well-defined mass was noted in the right adnexa between uterus and right ovary, with central echogenicity [Figure 1]. Uterus and bilateral ovaries appeared normal and there was no fluid in pouch of Douglas. A differential diagnosis of rudimentary horn with unicornuate uterus was considered. On USG upper abdomen, multiple non-obstructing right renal calculi and gall bladder (GB) calculi were noted. Serum calcium, phosphate, parathyroid hormone (PTH), uric acid, liver

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and renal function tests, urine analysis, and urine calcium were within normal limits.

MRI pelvis was done to further characterize the adnexal mass. On MRI, the uterus appeared normal with a well-defined, rounded, non-communicating cavitated mass measuring 3 × 4 cm noted along the right anterior uterine wall just below the insertion of round ligament. The cavity was lined by T2-hyperintense endometrium with hemorrhagic contents within, which appeared hyperintense on T1 and hypointense on T2W images [Figure 2]. The junctional zone of the cavity was thickened (13 mm) and endo-myometrial interface was indistinct. The main uterine cavity was normal in shape and size, and both the cornua were visualized normally [Figure 3] which ruled out USG diagnosis of rudimentary horn. The junctional zone, endomyometrial interface, and myometrial signal intensity of the main uterine cavity were normal. Both ovaries were normal. No pelvic endometriotic deposits were seen and there was no evidence of hematosalpinx. Based on the above findings, a diagnosis of ACUM was considered.

As HSG is best avoided in young unmarried patients, laparoscopy was done. On laparoscopy, the mass was found to be attached to the right anterior uterine wall just below the attachment of right round ligament. Bilateral fallopian

tubes and ovaries were normal with no endometriotic deposits. A transverse incision was made over the anterior wall of the mass and 10-12 ml of chocolate-colored fluid was drained. The uterine cavity was not entered into and the myometrial defect was closed. There was no communication with the main uterine cavity. The postoperative course was uneventful and the patient gradually improved symptomatically.

Histopathology revealed a cavitated mass lined by functional endometrium with glands and stroma surrounded by irregularly arranged smooth muscle cells. Foci of adenomyosis were also noted within the myometrium of the mass [Figure 4]. The smooth muscle cells stained positive for desmin, estrogen receptor (ER), and progesterone receptor (PR).

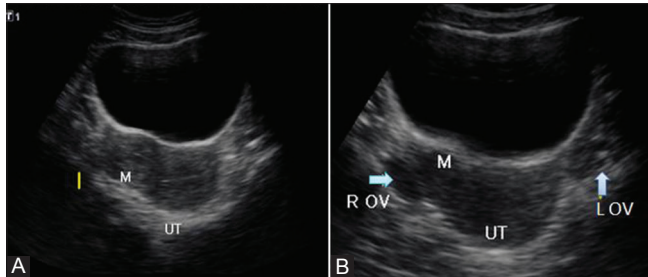


Figure 1 (A-B) : USG pelvis transverse images showing well-defined isoechoic uterus like mass with central echogenic endometrium in the right adnexa. Both ovaries are seen separately. UT= uterus, M = mass, arrow (⇒) ovaries

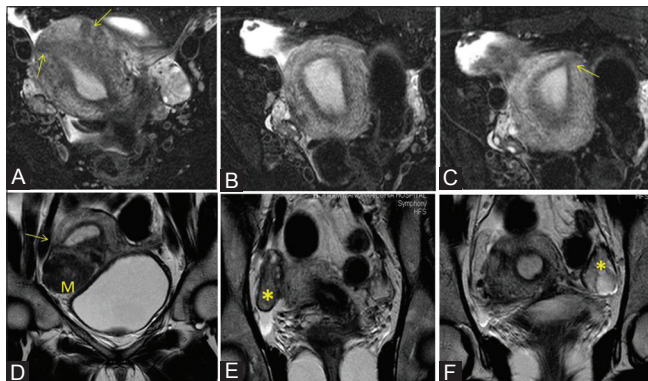


Figure 3 (A-F) : Axial FS T2W images (A-C) and coronal T2W images (D-F) showing normal-shaped uterine cavity with both cornua (arrows) attached normally. Bilateral ovaries (*) are seen separate from the mass, M

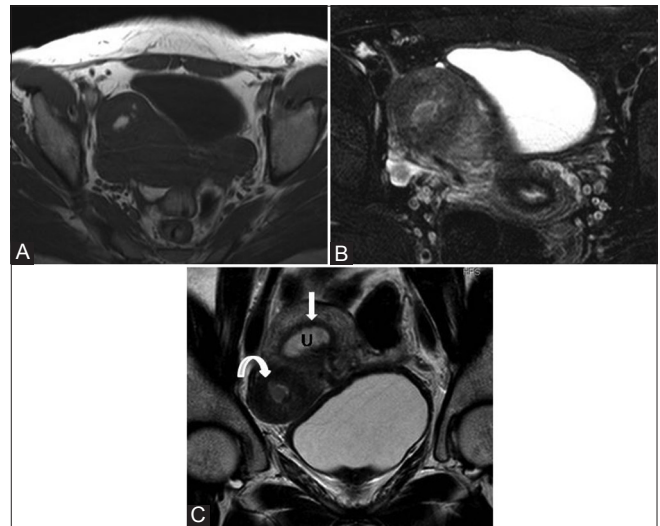


Figure 2 (A-C) : (A) Axial T1W, (B) fat-saturated axial T2W, and (C) coronal T2W images showing a cavitated mass attached just below the round ligament containing T1-hyper and T2-hypointense contents suggestive of blood products (A and B) and lined with T2-hyperintense endometrium (C). The junctional zone is thickened with ill-defined endo-myometrial interface (curved arrow). Note thin (4 mm) junctional zone with sharp endo-myometrial interface (straight arrow) in normal uterine cavity, U

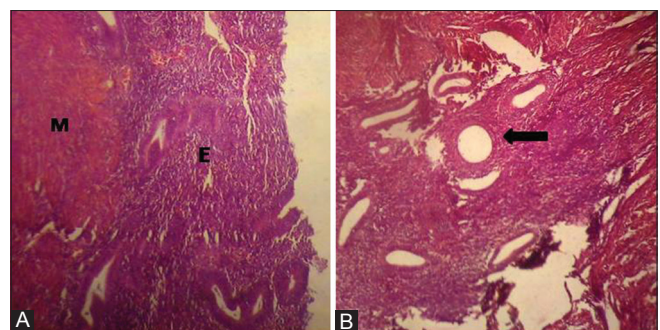


Figure 4 (A-B) : Photomicrograph (A) showing functional endometrium E, with glands and stroma lining the cavity wall and surrounded by irregularly arranged smooth muscle cells M, resembling myometrium. Foci of adenomyosis (B) are seen within the myometrium of the resected specimen (arrow)

Discussion

Uterus develops from embryonic fusion of two Mullerian ducts. Seven classes [Table 1] of Mullerian anomalies have been described.^[1] Septate uterus is the most common type, which is followed by unicornuate uterus.

Uterus-like mass (ULM) is an uncommon distinct entity described in literature which represents cavitated mass lined by endometrial glands and stroma that are surrounded by irregularly arranged smooth muscle cells, which in addition to other smooth muscle markers also show positivity for ER and PR, resembling myometrium.^[2] Hence, these ULMs bear both macroscopic and microscopic resemblance to the uterus and can arise anywhere within and beyond the uterus at any age.

ACUM is a non-communicating ULM arising in the uterus itself. The entity needs to be classified separately as the uterine cavity is otherwise normal unlike other Mullerian anomalies. It characteristically presents at a younger age, usually <30 years, with severe dysmenorrhea and chronic pelvic pain due to distention of the cavity caused by repeated bleeding. Various authors have previously described such masses with different names such as juvenile cystic adenomyoma (JCA), cavitated adenomyoma, accessory cavitated masses, etc., essentially representing the same entity now termed as ACUM. The condition is separate from cystic adenomyosis which is seen in middle-aged females due to diffusely spread adenomyotic foci in the uterus. The cysts are typically small, usually less than 5 mm, due to periodic hemorrhage in ectopic endometrium, and on histopathologic examination (HPE), they lack typical endometrial lining and uterus-like smooth muscle organization. ACUM, on the other hand, is seen in adolescents and resembles uterus on microscopy. The main uterine cavity and myometrium is otherwise normal; however, myometrium of ACUM itself may develop adenomyosis due to increased intracystic pressure.

There are three theories^[2] of development: (1) congenital anomaly theory, (2) heterotopias theory, and (3) metaplasia theory. Most of the authors accept ACUM as a congenital anomaly.^[3] The proposed mechanism says that the

accessory mass could be caused by duplication of ductal Mullerian tissue in the critical area at the level of attachment of round ligament, possibly related to gubernaculum dysfunction.^[3] Association with genitourinary and gastrointestinal anomalies have been described.^[2]

The first case^[4] of ULM was reported by Cozzutto in 1981. Around 36 cases of ACUM have been reported in literature with various terminologies, of which 22 have been reported after 2010 probably due to increased awareness. Beginning from 2010, the greatest number of cases ($n = 9$) has been reported by Takeuchi *et al.*,^[5] although they limited the inclusion criteria to women under 30 years of age. Kriplani *et al.*^[6] reported four cases in 2011, and one case each was reported by Akar *et al.*^[7] in 2010 and Chun *et al.*^[8] in 2011. In 2012, Jain^[9] reported two cases of JCA, simulating Mullerian anomalies. In 2013, another case with its laparoscopic management has been reported by Bedaiwy *et al.*^[10] Till now, around 31 cases of ULM arising outside uterus have been described.^[2] The most common extrauterine site is ovary; however, such masses have been seen in broad ligament, small bowel, mesentery, appendix, colon, conus medullaris, and utero-sacral ligament.

The criteria^[11] for diagnosing ACUM are: (1) an isolated accessory cavitated mass usually located under round ligament; (2) normal uterus, fallopian tubes, and ovaries; (3) a surgical case with excised mass and pathological examination; (4) an accessory cavity lined by endometrial epithelium with glands and stroma; (5) chocolate brown colored fluid contents; (6) no adenomyosis in the uterus (if resected), although there could be tiny foci of adenomyosis in the myometrium of the accessory cavity^[12] due to increased intracystic pressure. Although most of the cases fulfill all the above mentioned criteria, there have been few exceptions. There was one case showing two ACUM in the same patient, one case of ACUM in a patient with co-existing other Mullerian anomaly and one with accessory rudimentary tube attached with the mass.^[12]

USG is the initial imaging modality that can identify them as solid isoechoic to predominantly cystic masses resembling endometrioma arising within the uterus, visualized separately from the ovaries. On HSG, the mass may not be visualized at all. However, the most important role of HSG lies primarily in ruling out any Mullerian anomaly. MRI is the imaging modality of choice as it non-invasive and, hence, preferred over HSG in young unmarried females. It clearly shows the pelvic anatomy; cavitated mass with hemorrhagic contents; and the uterus, myometrium, and endo-myometrial interface. Hence, adenomyosis and pelvic endometriosis are best appreciated with MRI. Thin sections (3 mm) should be used as it will also help in ruling out Mullerian anomaly by demonstrating both cornua clearly.

Table 1: American Society of Reproductive Medicine classification of Mullerian anomalies

Class	Anomaly
1	Uterovaginal hypoplasia and agenesis
2	Unicornuate uterus
3	Uterus didelphys
4	Bicornuate uterus
5	Septate uterus
6	Arcuate uterus
7	Uterine anomalies related to diethylstilbestrol exposure

In our case, visualization of normal size and shape of the uterus and both cornua ruled out Mullerian anomalies. As bilateral ovaries were separate and appeared normal, ovarian tumors were excluded. Cystic degeneration in adenomyoma and fibroid will not show T2-hyperintense endometrial lining and hemorrhagic contents and are not usually seen in adolescents.

The entity closely mimics obstructed cavitated rudimentary horn^[9] with unicornuate uterus and differentiation may be difficult. HSG can be done to differentiate the two, but MRI is the preferred non-invasive modality. Both of them show cavitated mass lined by endometrium with hemorrhagic contents within it, but contralateral tilt of the uterus, banana-shaped small uterine cavity, and a single cornua favors obstructed horn^[13] over ACUM which was not seen in our case. However, at times, the differentiation may not be possible on MRI and laparoscopy remains the only option available for confirmation and treatment.

Regarding therapeutic management, most recent publications have included laparoscopic excision of the mass.

To the best of our knowledge, except a few cases, most of the cases were misdiagnosed preoperatively as other Mullerian anomalies, cystic degeneration in adenomyoma and leiomyoma, and broad ligament fibroids. Awareness and adequate knowledge of the entity can help the radiologist make accurate pre-operative diagnosis of ACUM.

Extensive search of literature did not show any association with renal and GB calculi. The findings are most likely incidental; however, further studies are needed.

Conclusion

ACUM, a rare Mullerian anomaly related to dysfunction of gubernaculum, is a treatable cause of severe dysmenorrhea in young females. The entity is not as rare as thought previously. MRI is highly accurate in making the diagnosis. The MRI findings of an accessory cavitated ULM located below the attachment of round ligament usually with hemorrhagic contents, an otherwise normal-shaped uterus with both cornua identified normally, without any evidence of adenomyosis, and bilateral normal tubes and ovaries should suggest the diagnosis of ACUM pre-operatively.

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