

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 a normally functioning uterus. This entity is problematic because of a broad differential diagnosis, including rudimentary and cavitated uterine horns; and is generally underdiagnosed. Magnetic Resonance Imaging (MRI) forms the mainstay of diagnostic imaging. The other modalities for diagnosis include Hysterosalpingography (HSG) and Ultrasonography (USG).*

Keywords: Accessory and Cavitated Uterine Mass, chronic pelvic pain, dysmenorrhea, MRI diagnosis, developmental Mullerian anomaly

1. Introduction

Accessory and Cavitated Uterine Mass (ACUM) is a rare, newly recognized Mullerian anomaly. It is an accessory cavity lined by functional endometrium within a normally functioning uterus, whereas in other Mullerian anomalies 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) [1, 2, 3]

ACUM is frequently observed in young, nulliparous women presenting with severe dysmenorrhea and recurrent pelvic pain despite taking analgesics or oral contraceptive pills (OCP). Some present with infertility. It is common among women under 30 years of age [4, 5]. ACUM is a diagnostic challenge and is often under - diagnosed. Differential diagnoses include rudimentary and cavitated uterine horns, adenomyosis with cystic or degenerated areas, and degenerating fibroids [2, 6, 7] The entity can be easily picked up on initial routine pelvic USG. Magnetic resonance imaging (MRI) is highly accurate in making diagnosis [8].

2. Case Presentation

A 22 - year - old nulligravida woman presented with severe dysmenorrhea and chronic pelvic pain since menarche which aggravated in the preceding 2 years. Pain was not relieved with repeated intake of analgesics. She attained menarche at the age of 13 years and began experiencing dysmenorrhea

since then which got worsened progressively in the recent years. She was hospitalized previously for IV analgesics for the same reason. She had normal menstrual history. Her general physical and per abdomen examination was normal. There was no history suggestive of pelvic inflammatory disease.

Transabdominal / transvaginal scan report revealed normal sized and anteverted uterus and both the ovaries showed multiple follicles. Myometrium showed a well - defined heterogenous cystic lesion with fluid - fluid level, posterior acoustic enhancement, and no internal/peripheral vascularity. Minimal fluid was noted in pouch of Douglas.

MRI pelvis was done to further characterize the adnexal mass. On MRI, the uterus appeared normal with a well - defined rounded altered signal intensity lesion measuring ~ 1.6 x 1.7 x 2.0 cm (TV x CC x AP) showing fluid level appears T1 hyperintense and central T2 shading effect suggest hemorrhagic component involving left lateral wall of myometrium in body of uterus causing mild displacement of endometrial cavity. However, lesions show no obvious communication with endometrial cavity. This lesion shows areas of blooming on GRE with mild restricted diffusion on DWI and low signal on ADC maps. Endometrial appears thickened and measures ~ 14 mm. Multiple T2 hyperintense tiny periphery arranged follicles with central echogenic stroma noted in bilateral ovaries. Mild fluid noted in pouch of Douglas. Based on the above findings, a diagnosis of ACUM was considered.

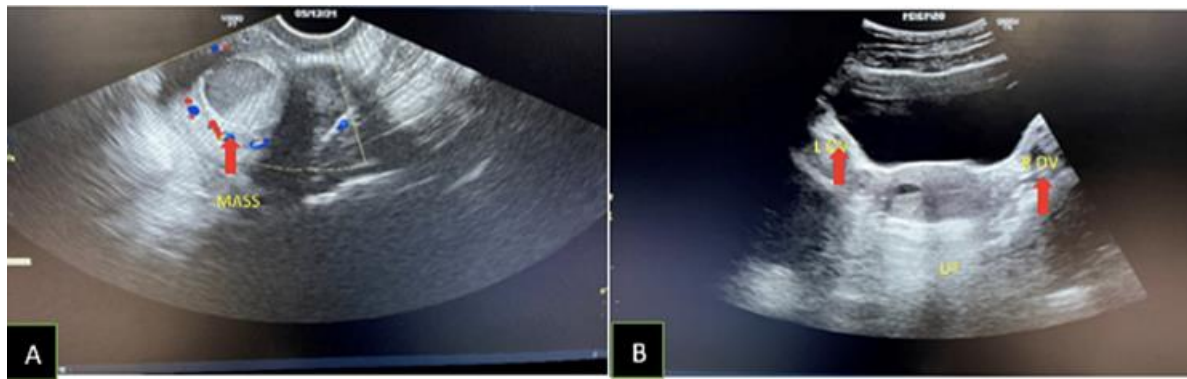


Figure 1: (A - B): TVS/TAS pelvis transverse images

TVS/TAS 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, Red arrow = ovaries.

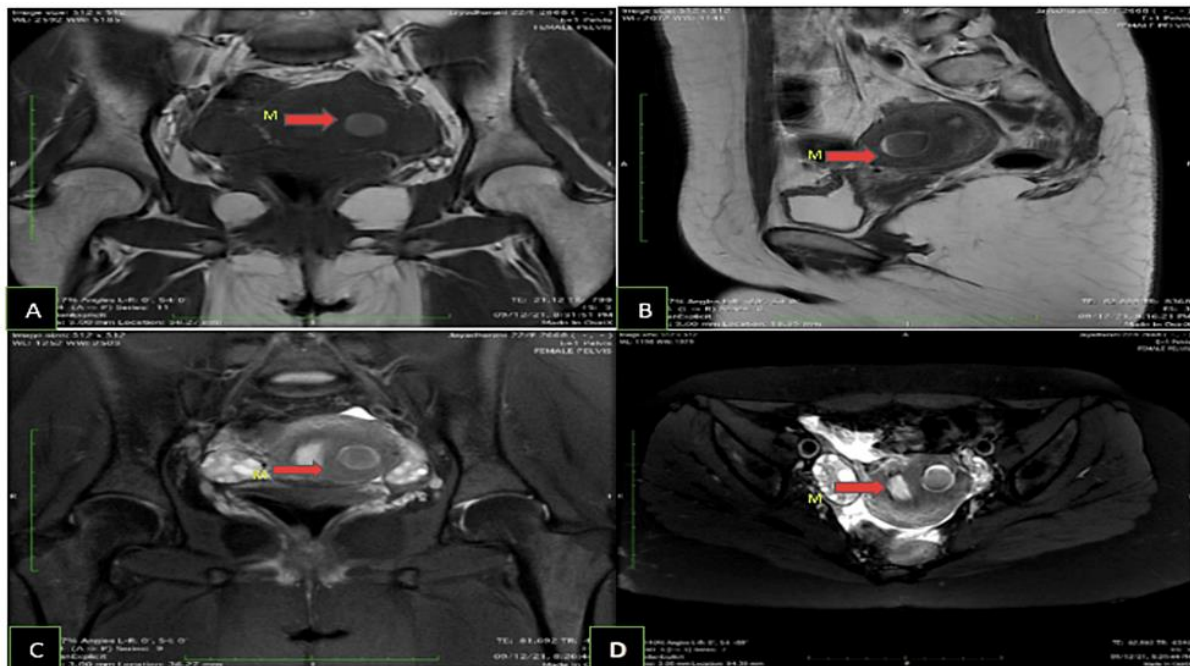


Figure 2: (A - D): Axial FS T2W images (A B) and coronal T2W images (C D) normal - shaped uterine cavity with both cornua (arrows) attached normally. Bilateral ovaries (*) are seen separate from the mass (M)

3. Discussion

An ACUM is a rare newly recognized, Mullerian anomaly. It is an accessory cavity lined with functional endometrium in addition to a normal uterine cavity. An ACUM is a cavitated mass confined to the myometrium; is encapsulated; and has uterus - like histologic organization [3]. The ACUM is lined by endometrial glands and stroma, and is surrounded by irregularly arranged smooth - muscle cells that show positivity for estrogen receptors and progesterone receptors, resembling myometrium [4]. The ACUM has both a macroscopic and microscopic resemblance to the uterus.

ACUM is a non - communicating uterus - like mass arising in the uterus itself. But this entity is different from other mullerian anomalies as the uterine cavity in ACUM is otherwise normal. Patients with ACUM are of younger age, usually <30 years, presenting with severe dysmenorrhea and chronic pelvic pain due to distention of the cavity caused by repeated bleeding. Previously such masses were described 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 can be misdiagnosed as cystic adenomyosis which is seen in middle - aged females. However, cystic adenomyomas consist of endometrial glands surrounded by endometrial - type stroma and smooth muscle but lack the uterus - like tissue found in ACUM. The cysts are typically small, usually less than 5 mm, due to periodic hemorrhage in ectopic endometrium [5, 6].

There are three theories of development: (1) congenital anomaly theory, (2) heterotopias theory, and (3) metaplasia theory [3]. Most of the authors accept ACUM as a congenital anomaly [4]. 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].

The first case of ULM was reported by Cozzutto in 1981.5 Malhotra V et al6 reported a case in a 30 - year - old multiparous female who presented with chief complaints of pain in her lower abdomen for 8 years. Her MRI revealed a

well - defined, rounded lesion of 2 cm in diameter which had a hyperdense center with peripheral enhancement that invaded the body of the uterus on the left side and the bilateral adnexa and ovaries appeared to be normal. Supermaniam et al [2]. reported two cases, first case was a 22 year old female presenting with 3- month history of severe pain after menses. Transabdominal ultrasound scan (TAUS) showed a cystic mass measuring 2.65×3.62 cm on the right side of the uterus, which appeared like a right endometrioma. Second case was a 36 - year - old married nulligravida woman presented with a history of chronic pelvic pain and severe dysmenorrhea. TVUS and TAUS revealed a cavitated right - sided intramural mass measuring 3.29×3.28 cm. The cavitated mass contained echogenic homogenous material resembling an endometrioma measuring 1.01×1.77 cm. Uterus was normal sized in both the cases.

Azuma et al [7]. reported ACUM in a 37 year old woman with similar complaints. Acien. P et al [4]. reported four cases of ACUM. Jain N et al.1 reported one case of ACUM in a 24 - year - old unmarried female presented with severe dysmenorrhea and chronic pelvic pain since menarche. Initially USG was done, later MRI revealed 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. In 2013, another case with its laparoscopic management has been reported by Bedaiwy et al [9].

The criteria 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 [10].

For diagnostic management, Ultrasound 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 [1]. But for further characterization of the mass, MRI is to be done. It is a non - invasive procedure and hence preferred over HSG in younger and unmarried females.

In this study, as MRI revealed a normal sized uterus, other mullerian anomalies can be ruled out. Cystic degeneration in adenomyoma and fibroid will not show T2 - hyperintense endometrial lining and hemorrhagic contents and are not usually seen in adolescents. However, at times, the differentiation may not be possible on MRI and laparoscopy remains the only option available for confirmation and treatment.

In most of the studies, therapeutic management, included laparoscopic excision of the mass. Most of the cases were misdiagnosed preoperatively as other Mullerian anomalies, cystic degeneration in adenomyoma and leiomyoma, and broad ligament fibroids. Awareness of such ACUM cases can help the radiologist to make accurate pre - operative diagnosis of ACUM.

4. Conclusions

ACUM, a rare Mullerian anomaly related to dysfunction of gubernaculum, is a treatable cause of severe dysmenorrhea in young females. Because of awareness the entity is not as rare as before. USG, MRI and HSG are preferred for diagnosis. But for younger and unmarried females MRI is the preferred procedure over HSG and is also highly accurate in making the diagnosis. The MRI findings of an accessory cavitated ULM with hemorrhagic contents, and usually non - communicating with the endometrial cavity with 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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