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CASE REPORT

# Role of 3D Coronal Ultrasound in Diagnosis of Accessory and Cavitated Uterine Mass: A rare Mullerian Anomaly

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## Abstract

Accessory and cavitated uterine mass is rare developmental Mullerian anomaly. There is a non-communicating uterus-like mass that occurs contiguously along wall of uterus often underdiagnosed and needs expertise to identify. To raise awareness, provide information about this pathology and emphasize role of coronal 3D ultrasound in its diagnosis. A 28-year-old married female presented with dysmenorrhea and chronic pelvic pain. On ultrasound, a homogeneously isoechoic mass was noted in right lateral wall of uterus with central echogenicity. On 3D reconstruction, the main uterine cavity was normal and both cornu were visualized without any recognized Mullerian anomaly. No communication with the main endometrial cavity seen. On laparoscopy, mass was located under right round ligament insertion. Sectioning revealed chocolate colored fluid. ACUM is non-communicating uterus-like mass. It resembles uterus both macroscopically and microscopically. It represents a cavitated mass lined by endometrial glands and stroma surrounded by irregular smooth muscle cells. Criterias for diagnosing ACUM are (1) accessory cavitated mass located under round ligament; (2) normal uterus, fallopian tubes, and ovaries (3) surgical case with excised mass and pathological examination; (4) accessory cavity lined by endometrium with glands and stroma; (5) chocolate-brown fluid contents. On ultrasound, they appear solid isoechoic masses with central cystic areas separate from ovaries. 3D reconstruction can be used to rule out Mullerian anomaly. ACUM is a rare surgically treatable cause of dysmenorrhea, often underdiagnosed due to lack of knowledge about entity. 3D ultrasound can be highly accurate in making the diagnosis.

## Introduction

An accessory and cavitated uterine mass (ACUM) is an uncommon newly recognized developmental Mullerian anomaly. It is defined as an isolated mass lined by functional endometrium and encapsulated by smooth muscle. It is locally defined to myometrium (unlike diffuse adenomyosis), encapsulated (unlike myoma), and bears uterus-like histological organization (unlike adenomyoma) [1]. It is often underdiagnosed and needs expertise to diagnose.

The objectives of this case report are to raise awareness and provide information about this interesting pathology, to emphasize the role of 3D coronal ultrasound in the diagnosis of this entity and to provide information regarding the importance of determining the characteristics and localization of the ACUM for rapidly choosing the most appropriate surgical option.

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## Case Report

A 28-year-old married female presented with complaints of dysmenorrhea and chronic pelvic pain, which had got worse for the past two years. She had regular menstrual cycles, with a normal flow and duration. There was no other significant medical or surgical history. The patient had undergone various clinical, laboratory and microbiological exams and transabdominal ultrasound to rule out possible causes of dysmenorrhea.

She underwent a detailed 2D and 3D transvaginal ultrasound assessment. The scan showed a large echogenic mass measuring 32 × 30 × 27 mm in size, within the right lateral myometrium. The mass had an echogenic rim and a cystic core with dense internal echoes. There was no evidence of endometrial indentation or communication with the endometrial cavity (Fig. 1). On color Doppler, vascularity was seen in the periphery of the mass without any intralesional vascularity (Fig. 2). The right ovary was separate from the mass. No pelvic endometriotic deposits were seen and there was no evidence of hematosalpinx.

On 3D coronal ultrasound, a well-defined, ovoid, non-communicating cavitated mass was noted along the right lateral uterine wall inferior to the level of right cornu. The main uterine cavity was normal in shape and size, and both the cornu were visualized and normal. There was no communication of this lesion with the endometrial cavity of the main uterus (Fig. 3). On the basis of clinical features and ultrasound findings, a diagnosis of accessory and cavitated uterine mass ( ACUM) was established.

Based on the ultrasound diagnosis of ACUM, the mass was approached laparoscopically rather than hysteroscopically. On laparoscopy, the mass was seen as a bulge along the right lateral myometrium below the attachment of round ligament (Fig. 4). Characteristically, this mass had an outer shell-like capsule of fibroid and when a nick was made over the anterior wall of the mass, chocolate colored endometriotic fluid started oozing out. This mass thus had characteristic laparoscopic features of outer shell-like capsule of a fibroid and inner core of an endometriotic cyst (Fig. 5). The cyst cavity was drained and the mass was removed in toto. The myometrial defect was sutured. The main uterine cavity was not entered and remained unopened at surgery. There was no communication seen with the main uterine cavity. The patient had an uneventful postoperative course and asymptomatic surgical outcome.

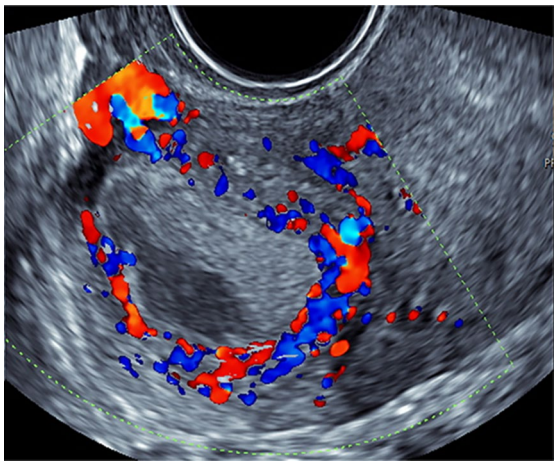

**Fig. 2** 2D Transvaginal color doppler showing vascularity along the periphery of the mass without any intralesional vascularity

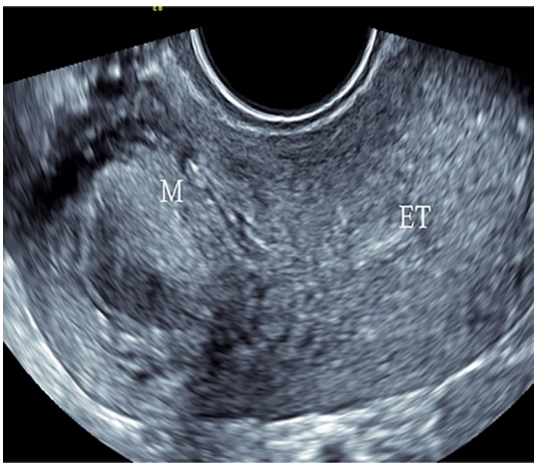


**Fig. 1** 2D Transvaginal transverse section of the uterus showing an echogenic mass with a central hypoechoic area and a visible non communicating separate endometrial lining ( *M* Mass, *ET* Endometrium).

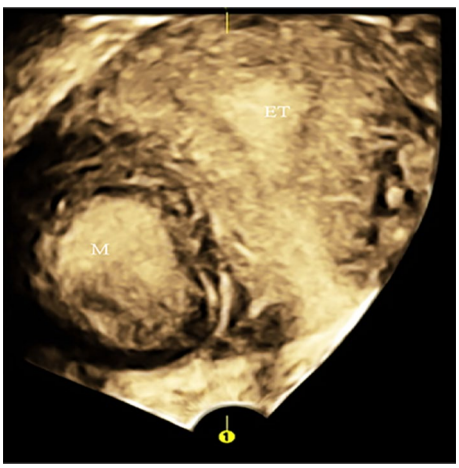


**Fig. 3** 3D Coronal Ultrasound showing a non-communicating cavitated mass along the right lateral uterine wall below the right cornu. Normal main uterine cavity with both the cornu visualized. (*M* Mass, *ET* Endometrium)

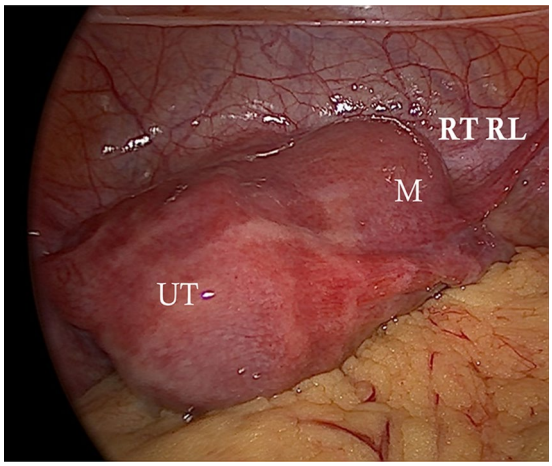


**Fig. 4** Laparoscopic image showing a bulging nodule on the right lateral surface of the uterus below the attachment of round ligament (*M* Mass, *UT* Uterus, *RT RL* Right round ligament

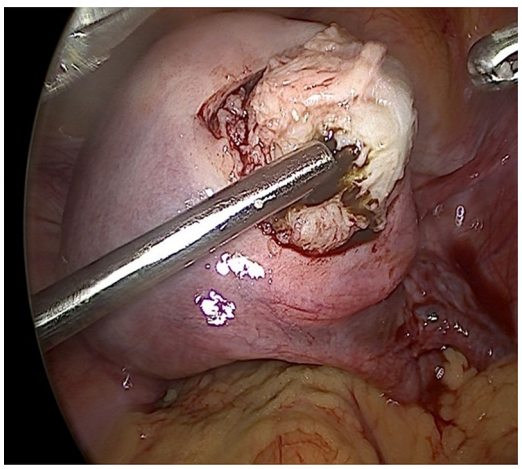

**Fig. 5** Laparoscopic image showing mass having an outer shell-like capsule similar to a fibroid and spillage of internal chocolate colored fluid

On histopathology, a cavitated mass lined by functional endometrium with glands and stroma surrounded by irregularly arranged smooth muscle cells was seen.

## Discussion

Uterus-like masses (ULMs) are rare lesions that resemble the uterus both macroscopically as well as microscopically [1]. Accessory and cavitated uterine masses (ACUM) are non-communicating ULMs that occur contiguously along the wall of the uterus. ACUM is described in literature as an accessory isolated cavitated uterine mass, lined with endometrial endothelium but with no communication to the normal uterine cavity. The uterine cavity in ACUMs is normal unlike other Mullerian anomalies and hence needs to be described more accurately. The condition is not included in the ESHRE classification systems [2] for uterine malformations, and thus not commonly diagnosed.

The etiology of these conditions is poorly understood. One theory is that ACUM is a uterine malformation resulting from a different etiology than common Mullerian uterine malformations described in the ESHRE/ESGE consensus [2]. The current explanation is that a gubernaculum dysfunction may be responsible for duplication or persistence of paramesonephric tissue leading to accessory uterine tissue at the attachment level of the round ligament.

The criteria [3] used to include a case as accessory and cavitated uterine mass were: (1) an isolated accessory cavitated mass; (2) normal uterus (endometrial cavity), tubes, and ovaries; (3) surgical case with excised mass and with pathological examination; (4) accessory cavity lined by endometrial epithelium with glands and stroma; (5) chocolate-brown-colored fluid content; and (6) no adenomyosis (if uterus removed), but there could be small foci of adenomyosis in the myometrium adjacent to the accessory cavity.

The key-point in diagnosis is clinical suspicion based on symptoms, patient age and knowledge of this entity. ACUM's present in young women under 30 years and have common but distressing clinical manifestations of chronic pelvic pain, severe dysmenorrhea and abdominal cramps resistant to common drug therapy. Repeated bleeding within the mass during the menses causes distension of cavity and thus chronic pelvic pain.

With a systematic 2D and 3D Transvaginal ultrasound examination and the knowledge of ACUM, detection should be possible in most cases. Ultrasound can identify these masses as solid isoechoic to predominantly cystic masses resembling an endometrioma arising within the myometrium of uterus, separate from the ovaries. The fluid inside the mass has "ground-glass" echogenicity and surrounded by myometrium. On 3D coronal ultrasound, the uterine cavity has a normal shape and there are two regular fallopian tubes [3]. Visualization of normal size and shape of the uterus and both cornu rules out traditional Mullerian anomalies. The mass is seen within the myometrium below the level of cornu. Thus, coronal 3D ultrasound not only clinches the diagnosis but also differentiate from differential diagnoses of Mullerian anomalies. Histopathology reveals a cystic adenoma like mass with an internal cavity lined with endometrium and an external muscular wall.

ACUMs are difficult to diagnose because of the broad differential diagnosis, of which obstructed cavitated rudimentary horn [4] with unicornuate uterus being the commonest. 3D coronal ultrasound in those cases reveals an isolated cavitated mass but with absent ipsilateral uterine cornu and a banana-shaped contralateral uterine cavity. It is important to differentiate between these two closely mimicking entities

because ACUM has a far better obstetrical and gynecological prognosis.

The misery of severe, chronic symptoms is rapidly relieved with laparoscopic uterine-sparing enucleation of the mass. Patients show a good recovery and asymptomatic post-surgical course.

## Conclusion

ACUM is a newly recognized, rare Mullerian anomaly that is probably related to a dysfunction of the female gubernaculum. It is often underdiagnosed or commonly misdiagnosed as obstructed cavitated rudimentary horn with unicornuate uterus due to lack of knowledge and awareness about the entity. Coronal 3D ultrasound is modality of choice not only to confidently diagnose ACUM but also to reliably differentiate from other recognized Mullerian anomalies. 3D ultrasound represents a cost-effective and noninvasive way to diagnose patients with ACUM. Clinical suspicion of this entity based on the symptoms, patient age, knowledge and awareness of this entity will help to indicate a preoperative diagnosis of ACUM more frequently and accurately.

### Declarations

**Conflict of interest** The authors declare that they have no conflict of interest.

**Human and Animal rights** No human or animal participants.

**Informed consent** Informed consent taken from patient–Yes.

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