

Accessory Cavitated Uterine Mass with Associated Chocolate Cyst and Ureteric Endometrioma: A Case Report

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ABSTRACT

Accessory Cavitated Uterine Malformation (ACUM) is a Müllerian anomaly characterised by a non communicating uterine cavity lined with functional endometrium and surrounded by myometrium. ACUM is an uncommon cause of pelvic pain and dysmenorrhoea. This condition was associated with a ureteric endometrial deposit and endometrioma, highlighting an unusual presentation. A 19-year-old unmarried female patient presented with complaints of right flank pain during her menstrual cycles, as well as irregular menstrual cycles alternating between hypomenorrhoea and polymenorrhagia since the onset of menstruation five years prior. Ultrasound and Magnetic Resonance Imaging (MRI) revealed a well-defined solid lesion on the right lateral wall of the uterine fundic region, located near the attachment of the round ligament. The right ovary displayed a cyst with multiple internal echoes, exhibiting a ground-glass appearance on ultrasound and a T2 shading effect on MRI, suggestive of an endometriotic cyst. Notably, right-sided hydronephrosis was observed due to a hypoechoic solid lesion near the ureterovesical junction on ultrasound, which appeared T2 hypointense on MRI, indicative of an endometriotic deposit. The present case illustrates the utility of ultrasound and MRI in diagnosing ACUM, along with the associated chocolate cyst and ureteric endometrioma, resulting in obstructive uropathy—an exceptionally rare combination.

Keywords: Magnetic resonance imaging, Non communicating uterine horn, Uterine cavity

CASE REPORT

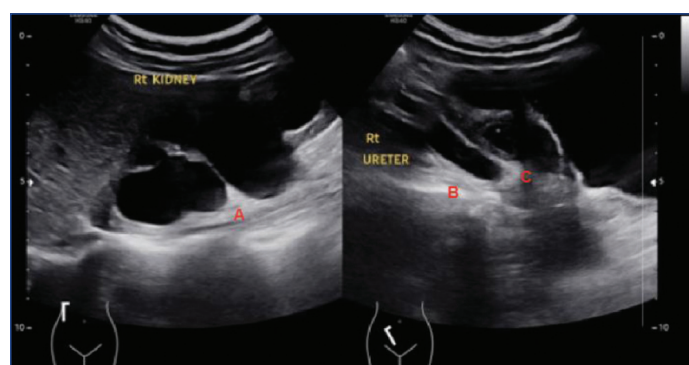
A 19-year-old unmarried female patient presented to the study Institute with complaints of right flank pain, pain during menstrual cycles, and irregular cycles, alternating between hypomenorrhea and polymenorrhagia since the onset of menstruation five years ago.

On physical examination, no significant abnormalities were detected. Pervaginal and per speculum examinations were not performed due to her unmarried status. An abdominal examination revealed no tenderness, guarding, or rigidity.

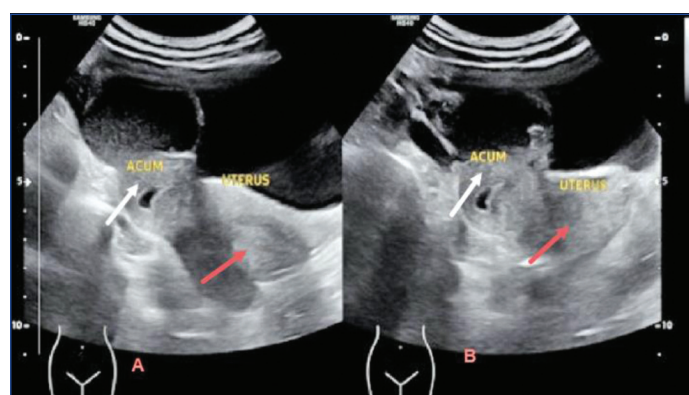
Her routine blood investigations—including Complete Blood Count (CBC), serum electrolytes, serum proteins, urine routine and microscopy with urine culture analysis, routine Renal Function Tests (RFT), Liver Function Tests (LFT), random blood sugar, and blood serology—were all normal.

Ultrasound of the abdomen and pelvis revealed marked hydronephrosis in the right kidney with hydroureter [Table/Fig-1]. Narrowing was noted at the lower end of the ureter, with a small hypoechoic solid lesion measuring 8×4 mm adjacent to it, likely representing an endometriotic deposit involving the lower ureter [Table/Fig-1]. A well-defined, solid, round to oval-shaped lesion measuring 28×29×26 mm Craniocaudal length x Anteroposterior length transverse length (CC×AP×TR) was observed on the right lateral surface of the uterine fundus [Table/Fig-2]. This lesion appeared isoechoic to the uterine myometrium, with its central portion showing a linear echogenic endometrium and a thin anechoic fluid collection that did not communicate with the uterine cavity [Table/Fig-2].

The right ovary was bulky due to a well-defined anechoic cyst measuring 46×28 mm, which exhibited multiple internal echoes with a ground-glass appearance, suggestive of an endometriotic cyst (also known as a chocolate cyst) [Table/Fig-3]. The cyst appeared adherent to the ACUM and the adjoining uterine fundus. Mild anechoic free fluid was noted in the pouch of Douglas, likely due to retrograde menstruation.

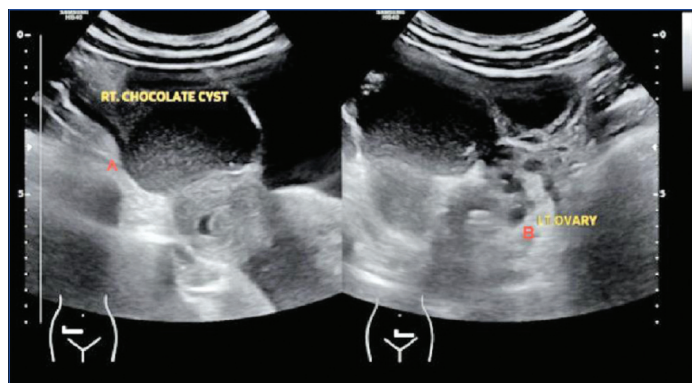


[Table/Fig-1]: Ultrasonography (USG) image showing a) Hydronephrosis in the right kidney; b) Right hydroureter in its entire extent; c) due to hypoechoic endometrial deposit in the right lower ureter.



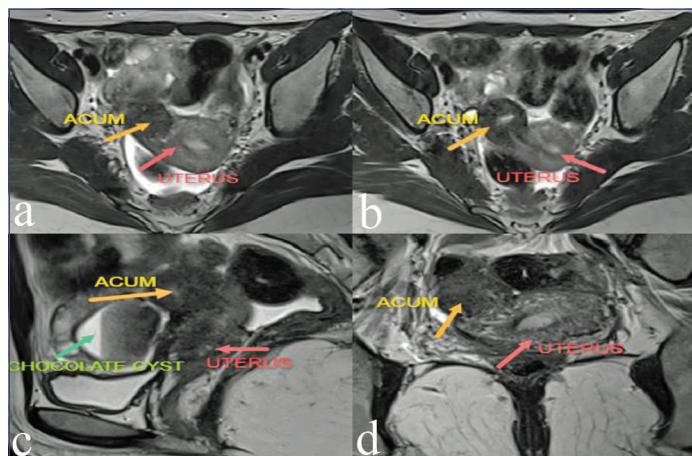
[Table/Fig-2]: USG image a) (axial oblique); b) (axial) showing ACUM (white arrows) with thin anechoic fluid content in the center at the right side of uterine fundus and normal uterus (red arrows).

Based on the ultrasound findings, a diagnosis of accessory cavitated uterine mass with right ovarian endometrioma and an endometriotic deposit in relation to the right lower ureter just proximal to the ureterovesical junction, resulting in proximal obstructive uropathy, was made.



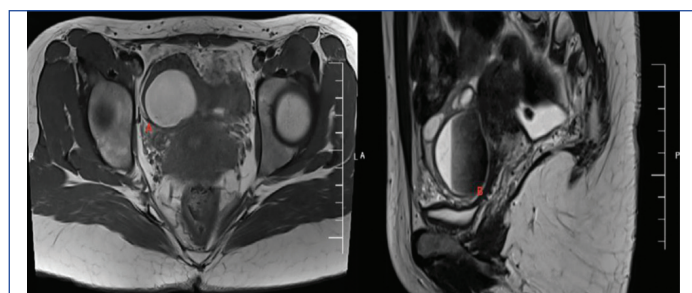
[Table/Fig-3]: USG showing chocolate cyst in right ovary filled with multiple internal echoes a) with ground glass appearance b) with normal left ovary.

The MRI of the pelvis and abdomen was performed for further evaluation and confirmation. A well-defined solid lesion measuring approximately 31×24×25 mm (AP×TR×CC) was noted on the right lateral wall in the fundic region [Table/Fig-4]. It appeared T1 isointense and T2 hypointense, with its central portion being hyperintense on T2-weighted Imaging (T2WI) [Table/Fig-4]. There was no diffusion restriction observed on Diffusion-Weighted Imaging (DWI). The uterus was deviated towards the left [Table/Fig-4].



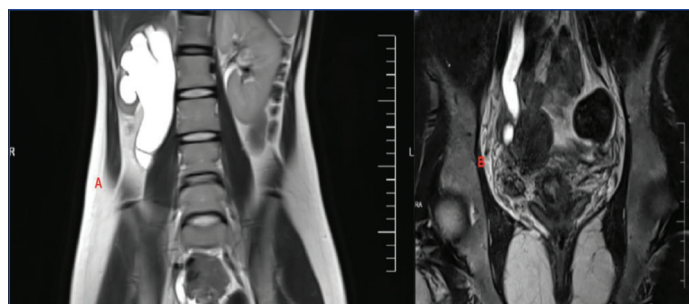
[Table/Fig-4]: a,b) (Axial T2WI); c) (Sagittal T2WI); and d) (Coronal T2WI) images show ACUM arising from the right anterolateral uterine wall in uterine fundus region. Yellow arrows=ACUM, Red arrows=Normal uterus, Green arrow=Chocolate cyst.

Additionally, a well-defined lesion was noted in the right ovary, measuring approximately 42×29×40 mm (CC×AP×TR). This lesion was hyperintense on T1-weighted imaging (T1WI) and T2WI, with T2 shading. It was not bright on T1DIXON F and not nullified on T1DIXON W, exhibiting mild blooming on Gradient Echo (GRE) sequences [Table/Fig-5]. The lesion was indenting the dome of the urinary bladder and was suggestive of a chocolate cyst [Table/Fig-5]. The right kidney showed severe hydronephrosis with thinned renal parenchyma [Table/Fig-6]. The entire ureter was dilated and tortuous [Table/Fig-6].



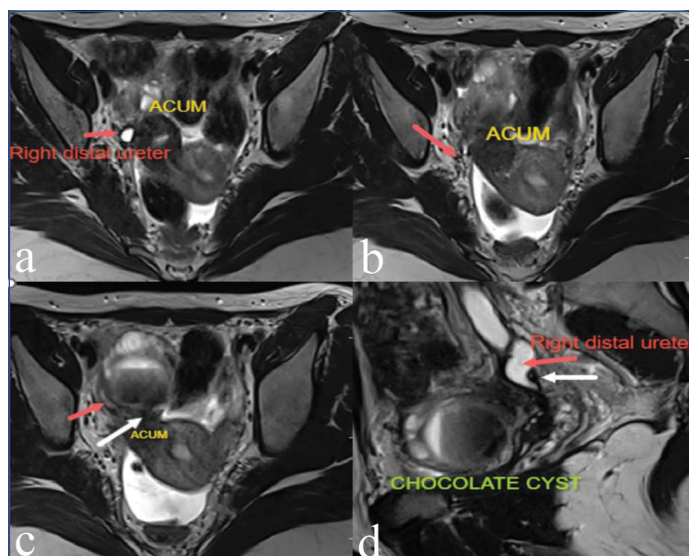
[Table/Fig-5]: (a) T1W Axial and (b) T2W Sagittal showing chocolate cyst in right ovary appearing hyperintense on T1W Axial; showing T2 shading on T2W Sagittal.

The lower ureter exhibited mild circumferential wall thickening (2-3 mm) proximal to the right Ureterovesical (UV) junction, along with a well-defined T2 isointense-hypointense lesion measuring



[Table/Fig-6]: Coronal MRI Abdomen (a) and Coronal MRI Pelvis (b) showing right-sided marked hydronephrosis up to the pelvic portion.

approximately 12×10 mm in the vicinity just proximal to the right UV junction, likely representing an endometriotic deposit [Table/Fig-7]. These findings confirmed the ultrasound diagnosis.



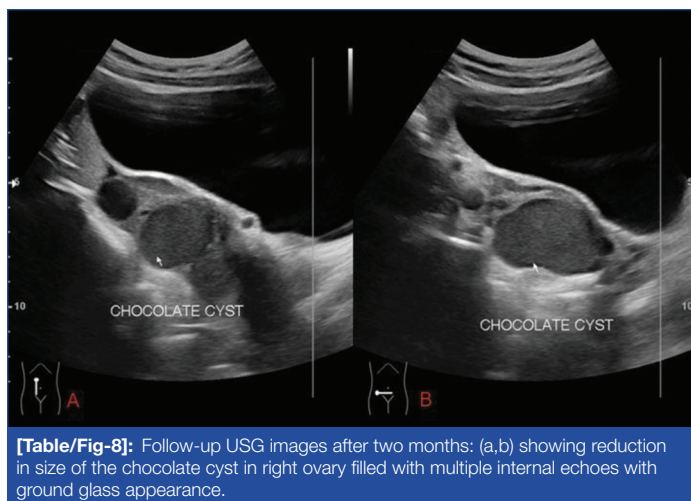
[Table/Fig-7]: a,b) (Axial T2WI) shows dilated right ureter with right distal ureteric wall thickening occurring due to endometriotic deposit at its lower end indicated by white arrow in images c) (Axial T2WI) and d) (sagittal T2WI). Red Arrows=Right distal ureter, White Arrows=Distal Ureteric endometriotic deposit (appearing isointense to hypointense on T2WI).

The lesion appears to lie within the right lateral uterine wall (intramyometrial) and is seen to be continuous with the right anterolateral uterine wall near the fundus, beneath the insertion of the round ligament attachment. It did not appear to extend outside the normal uterine contour, with no vaginal or renal anomalies observed. Therefore, based on radiological imaging, it is classified as ACUM rather than a non communicating uterine horn.

The patient underwent cystoscopy with the placement of a right Double-J (DJ) stent for right hydronephrosis, followed by hormonal treatment for endometriosis (injection of Lupride Depot 3.75 gm intramuscularly, monthly). Follow-up ultrasound after two months revealed marked regression of the right-sided hydronephrosis and hydronephrosis, along with minimal regression in the size of the chocolate cyst [Table/Fig-8]. The patient is currently still on hormonal treatment, as she has not given consent for surgical excision of the ACUM, considering her age.

DISCUSSION

The ACUM is a relatively under-recognised entity, defined by the presence of a non-communicating accessory cavity lined with endometrium and surrounded by uterine smooth muscle [1]. It is a rare Müllerian anomaly and represents an auxiliary uterine chamber that is closely related to the uterus but appears unable to interact with its main body [2,3]. The bilateral ovaries and fallopian tubes, along with the endometrial cavity, are normal [3,4]. In 2021, ACUM was officially recognised based on its specific location, symptoms, and characteristic imaging findings [1].



The ACUM is typically observed in the lateral portion of the myometrium directly beneath the insertion of the round ligament. It is a diagnosis of exclusion and an uncommon cause of dysmenorrhoea. ACUM is primarily observed in women of reproductive age, particularly those under 30, and is infrequently recorded in multiparous women. The signs and symptoms of ACUM might appear as soon as menarche or shortly thereafter [1]. Due to the functioning endometrium and bleeding within the lesion during the menstrual cycle, ACUM clinically manifests as dysmenorrhoea and cyclic pelvic pain [5].

The main symptoms include pelvic discomfort, dyspareunia, and dysmenorrhoea that do not improve with medication. While it can be asymptomatic in some individuals, pain typically worsens after menstruation begins [6]. Although 7% of women will be diagnosed with a reproductive tract anatomical abnormality, the prevalence of ACUM is unknown due to its rarity, which may lead to significant underestimation and underdiagnosis [7].

Commonly accepted diagnostic criteria for ACUM, as proposed by Acién P et al., include: an isolated accessory cavitated mass, a normal uterus with a functional endometrial-lined cavity, normal fallopian tubes and ovaries, pathological confirmation after surgical excision, a cavity filled with chocolate-coloured fluid, no true adenomyosis, an accessory cavity lined with functional endometrium containing glands and stroma, and a concentric smooth muscle arrangement in the myometrial mantle [6].

However, the fluid within the endometrial cavity can be anechoic in 8.7% of cases on ultrasound, hypointense on T1-weighted imaging (T1WI) in 4.5% of cases, and hyperintense on T2WI in 32.6% of cases [8].

The ACUM is most commonly found in the left intrauterine region (38.6%), followed by the right intrauterine region (28.6%) and the broad ligament (15.7%). Rare extrauterine cases (4.3%) have been reported in locations such as the left uterosacral ligament, mesoappendix, lower lumbar area near the jejunum, and the descending colon [8]. Extrauterine ACUM suggests a higher likelihood of Müllerian duct malformation or remnant, rather than gubernaculum dysfunction [9-11].

Ultrasound, especially Transvaginal Ultrasound (TVS), is the primary tool for diagnosing ACUM. Its diagnosis often remains uncertain due to a lack of awareness. It is frequently misdiagnosed as a subserosal fibroid, degenerated fibroid, or endometrioma [3,4,12].

Differential diagnosis include degenerating leiomyoma, cystic endometriosis, or a non communicating rudimentary horn, all of which share symptoms such as dysmenorrhoea and pain unresponsive to medication. Early recognition can lead to timely diagnosis, effective treatment, and better resource allocation, preventing years of unnecessary and ineffective management [5,13].

Cystic adenomyoma typically presents in older women and shows anechoic cysts within the myometrium, along with changes of adenomyosis in the rest of the uterus [4]. A non communicating rudimentary uterine horn features a cavity within the horn along with a fallopian tube that does not communicate with the uterus. It is usually observed in a unicornuate uterus, which is displaced off the midline [4]. A subserosal or degenerated fibroid consists of smooth muscle cells and fibrous connective tissue, typically seen in reproductive-age women. It appears hypoechoic compared to the myometrium on ultrasound and shows fluid collection only after degeneration [4].

There is very little evidence in the medical literature of ACUM occurring concurrently with either ovarian endometriomas or ureteric endometriosis. A high index of clinical suspicion, comprehensive imaging, and histopathological evaluation are needed for accurate and timely diagnosis.

The ACUM and endometriosis are distinct conditions with different mechanisms. Detailed research is required to identify a common pathway for their association. A comprehensive literature review has been conducted regarding the association of ACUM with chocolate cysts and/or ureteric endometriosis.

1. **Accessory Cavitated Uterine Malformation (ACUM) with chocolate cyst:** A 30-year-old woman, who was not pregnant, presented with rectal tenesmus and dysmenorrhoea that had been worsening over five years. Ultrasound revealed a well-defined, heterogeneous, thick-rimmed anechoic lesion in the lower-left anterior myometrium. MRI showed a circular lesion in the lower-left uterine wall on both T1- and T2-weighted images. It was isointense to the myometrium and displayed comparable features on contrast-enhanced scans [14]. During laparoscopy, a window-like defect in the left pelvic peritoneum, purple-blue endometriotic lesions on the right ovary, and a lesion on the left lateral myometrium near the inner side of the round ligament were discovered. The removed mass had thick walls, was spherical, grey, and contained chocolate-coloured fluid. Histopathology confirmed an ACUM surrounded by functioning endometrial glands and stroma within the myometrium [14].
2. **Ureteric endometriosis:**
 - a. Although some research indicates that endometriosis involving the ureter is relatively unusual, by the late 1980s, at least 121 cases had been described. The first cases were reported in the early 20th century, and the cumulative total of cases across various studies has varied [15].
 - b. It is quite uncommon for endometriosis to involve the ureters intrinsically. The present case report details the intrinsic involvement of the ureters by endometriosis. The patient was a 47-year-old woman, gravida 4, para 2, who had experienced hypermenorrhoea and dysmenorrhoea for four years. A right hydronephrosis was discovered during an intravenous pyelogram. She underwent right ureteroureterostomy and a total abdominal hysterectomy. Pathological investigation revealed intrinsic intramural endometriosis completely blocking the right ureter [16].

First identified by Cullen in 1917, ureteric endometriosis is an uncommon condition that accounts for 0.1% to 0.4% of genitourinary tract endometriosis. With a bladder/ureter/kidney/urethral endometriosis ratio of 40:5:1:1, the ureters are the second most prevalent site in the urinary tract affected by endometriosis. However, an increase in incidence has been noted over time, most likely due to heightened awareness and improved diagnosis [17]. In the literature review, we could not find any case of ACUM with the co-occurrence of chocolate cyst and ureteric endometriosis.

CONCLUSION(S)

The ACUM is rare and typically observed in younger individuals. It should be suspected in patients presenting with dysmenorrhoea,

chronic cyclic pelvic pain, and infertility. Both ultrasound and MRI serve as confirmatory tools for diagnosis. The simultaneous occurrence of ACUM with a chocolate cyst and ureteric endometrioma is extremely rare. Timely diagnosis and effective treatment can help avoid future complications. A high index of clinical suspicion, coupled with meticulous use of radiological imaging, is essential for early diagnosis.

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AUTHOR DECLARATION:

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

PLAGIARISM CHECKING METHODS: [Jain H et al.]

- Plagiarism X-checker: Mar 07, 2025
- Manual Googling: May 05, 2025
- iThenticate Software: May 07, 2025 (7%)

ETYMOLOGY: Author Origin

EMENDATIONS: 7

Date of Submission: Mar 04, 2025

Date of Peer Review: Mar 24, 2025

Date of Acceptance: May 09, 2025

Date of Publishing: Jun 01, 2025