International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Elhabashy A et al. Int J Reprod Contracept Obstet Gynecol. 2025 Mar;14(3):698-702
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20250498

Original Research Article

Accessory cavitated uterine malformation: an underdiagnosed Mullerian anomaly

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Received: 01 January 2025
Revised: 07 February 2025
Accepted: 10 February 2025

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ABSTRACT

Background: Accessory cavitated uterine malformation (ACUM) is an extremely rare Mullerian anomaly that is not yet classified by the international societies as a type of uterine malformations. Due to its rarity, it is usually misdiagnosed as adenomyoma. Objective was to provide specific sonographic features of ACUM and differentiate it from the more common adenomyoma.
Methods: Our study is a comparative retrospective study. We presented the sonographic features of 3 cases of ACUM and compared these features with the sonographic features of 10 cases of adenomyoma. All cases had scanned by 2D and 3D luteal TVUS at Habashy 4D scan centre (Alexandria; Egypt) between June 2019 and June 2024. All the 13 cases in our study were had chronic pelvic pain and dyspareunia with a lesion in the myometrium that was not matched with the whorly echotexture of myoma. We had described the sonographic characteristics of the lesion and matched them with the final diagnosis after histopathology.
Results: TVUS features of the 3 cases who had ACUM were cystic lesion within the lateral myometrial wall. Its content has ground glass echotexture. TVUS features of the 10 cases of adenomyoma were heterogeneous ill-defined solid myometrial lesion with translesional minimal flow.
Conclusions: ACUM is a rare uterine malformation that is usually misdiagnosed as adenomyoma. Distinction between these two pathologies is important as the treatment of both differ. ACUM is suggested in patients with pelvic pain when there is a myometrial cystic lesion separable from a normal uterine cavity and contained a ground-glass material.

Keywords: Accessory cavitated uterine malformation, Adenomyoma, Sonographic features

INTRODUCTION

Accessory cavitated uterine malformation (ACUM) or juvenile cystic adenomyosis (JCA) is an extremely rare Mullerian anomaly with less than 150 cases reported in literature.[1] Three definitions had been proposed till now for ACUM by Takeuchi et al, Acien et al and Naftalin et al.[2-4] Summary of these descriptions is ACUM represents a cystic myometrial lesions (>1 cm) with echogenic contents and is separable from a normal uterine cavity in a case complaining of dysmenorrhea and her age is <30 years old.[1-4]

Three pathogenesis for ACUM had been proposed which are: metaplasia, mullerianosis and Mullerian duct fusion anomaly.[5-8] Though the last pathogenesis is the most acceptable one; ACUM is not yet classified by uterine anomaly classification of the European Society of Human Reproduction and Embryology (ESHRE)/the European Society of Gynecological Endoscopy (ESGE) nor the revised American Society of Reproductive Medicine (rASRM). ACUM is included in the embryological–clinical classification of female genital malformations proposed by Acién. ACUM could be due to duplication and persistence of a Mullerian duct segment at the level of round ligament attachment, possibly related to gubernaculum dysfunction.[9-13]

Cases who had ACUM present mainly with severe dysmenorrhea that rarely respond to medical treatment. Many case reports and case series had described ACUM as a cystic mass within the lateral myometrial wall that contained blood products. This mass is adjacent to a normal shaped uterine cavity. Intraoperative and postoperative assessment revealed cystic cavity containing thick brown fluid consistent with old blood, lined by endometrium and surrounded by myometrium.[14,15]

Due to its rarity, it is usually misdiagnosed as adenomyoma. Other differential diagnosis of ACUM beside adenomyoma are four conditions; which are: myoma, unicornuate uterus with functioning non-communicating horn, Robert's uterus and interstitial ectopic pregnancy.[16-18] The clinical awareness of ACUM as a possible cause of severe dysmenorrhea is unfortunately confined to few practitioners and our study is aiming to achieve better understanding of ACUM, including symptoms, diagnosis and management of this rare uterine malformation.

Objective

The objective of our study is to provide specific sonographic features of ACUM and differentiate it from the more common adenomyoma.

METHODS

Our study is a comparative retrospective descriptive study that was approved by the ethical committee of medical research at Faculty of Medicine; Alexandria University; Egypt. Patients' consents had been signed.

We presented the sonographic features of 3 cases of ACUM that was proved by histopathology as such postoperatively (after excisional biopsy). Then we will compare these features with the sonographic features of 10 cases of adenomyoma that was proved by histopathology as such post-hysterectomy.

All cases had scanned by two-dimensional (2D) and three-dimensional (3D) transvaginal ultrasound (TVUS) assessment in the luteal phase of a spontaneous cycle at Habashy 4D scan center (Alexandria; Egypt), using: GE Voluson S-10 Expert RIC5-9A probe. Study had conducted between June 2019 and June 2024; and the long duration explained by the rarity of ACUM. Pre-scanning beta human chorionic gonadotropin (β-hCG) was negative. All the 13 cases in our study were had chronic pelvic pain & dyspareunia with a lesion in the myometrium that was not matched with the whorly echotexture of the myoma. We had described the sonographic characteristics of the lesion and matched them with the final diagnosis after histopathology.

Data were fed to the computer and analyzed using IBM statistical package for the social sciences (SPSS) software package version 20.0. (Armonk, NY: IBM Corp). Categorical data were represented as numbers and percentages. Quantitative data were expressed as mean and standard deviation and median. Student t-test was used to compare two groups for normally distributed quantitative variables.

RESULTS

There was statistically significant difference between the mean age of ACUM cases versus that of adenomyoma cases (35 years versus 43 years respectively). Parity was also higher in adenomyoma cases in comparison to ACUM cases. All cases of ACUM and adenomyoma had complained of: chronic pelvic pain (CPP); dysmenorrhea and dyspareunia. All the 10 cases in our study who had adenomyoma had complained of abnormal uterine bleeding (AUB). One of the 3 cases in our study who had ACUM had complained of AUB.

Table 1: Demographic data of the studied groups.

Variables	ACUM (n=3)	Adenomyoma (n=10)	P value*
Age (years) (mean±SD)	35±5.21	43±5.4	0.008
Parity (mean±SD)	1±2	3±1	0.001
AUB	1 case (33.33%)	10 cases 100%	<0.001

*P value for comparing between both groups (significant if p<0.05)

TVUS features of the 3 cases who had ACUM were rounded cyctic lesion within the lateral wall of the corpus of the uterus. This cystic lesion has a myometrial mantle and inner smooth surface. Contents of the cyst has ground glass echotexture similar to what is present in ovarian endometrioma. The lesion is separable from the uterine cavity that was normal in shape and geometry. There was minimal vascularity in the myometrial mantle of the lesion on Doppler mapping with no vascularity in its central core. Mild tenderness noted on targeted probing of the lesion. No associated adenomyosis nor endometriosis. In all the 3 cases of ACUM in our study the size of the lesion was about 25×30×25 mm. 2 cases of them was on the left side and one case was on the right side. Figures 1 and 2 showed the previously mentioned sonographic description for ACUM.

Patients counselled about the various therapeutic modalities of ACUM and they opted surgical intervention. Laparoscopy was done for them and revealed a small rounded lesion in the lateral wall of the uterus near the cornu between the insertion of the round ligament and the origin of the fallopian tube. The lesion had a cystic sensation on manipulation. Laparoscopic excision of the lesion had been done safely and specimen sent for pathology. During dissection of the lesion; brownish fluid (chocolate like) had been expelled. The cystic nature of the myometrial lesion together with the presence of chocolate like material within it confirm the diagnosis of ACUM.

Figure 3 showed the laparoscopic findings of one of our ACUM cases.

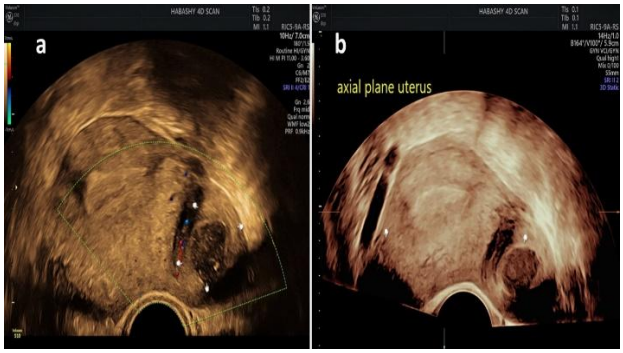

Figure 1: Axial plane of the uterus in ACUM (a) 2D-TVUS: cystic myometrial lesion near the left cornu, separable from the endometrium and partially extrudes the serosa, myometrial mantle noted around the lesion with smooth inner lining, contents showed ground glass echotexture, minimal vascularity noted at the lesion's periphery with no vascularity in its central core, and (b) 3D-TVUS; minimum mode.

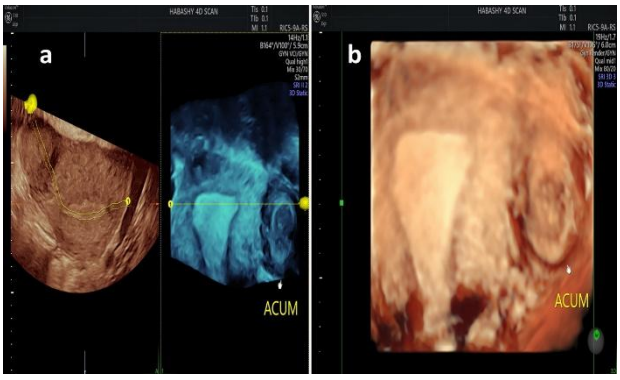

Figure 2: Coronal view of the uterus in ACUM using 3D-TVUS; showing cystic myometrial lesion separable from a normal uterine cavity (a) omni-view mode, and (b) HD-live surface rendering mode.

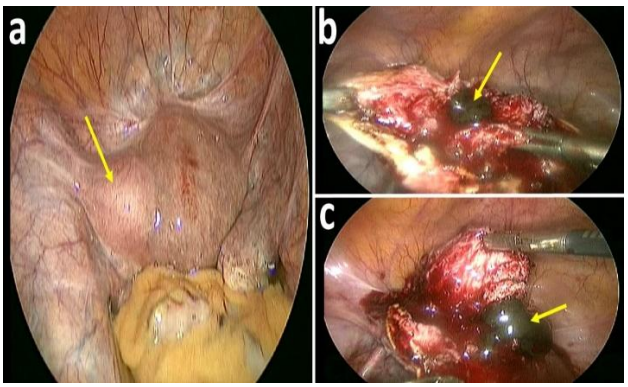

Figure 3: Laparoscopy for ACUM (a) rounded lesion at the left cornu of the uterus; yellow arrow, (b and c) chocolate like material extruded from the ACUM; yellow arrows.

Histopathological assessment of the 3 cases revealed myometrial tissue with endometrial covering that showed endometrial glands and stroma. This histopathology confirms the diagnosis of ACUM. No atypia noted in both myometrial and endometrial tissues assessed. Dramatic symptomatic improvement had been reported by the 3 cases on follow up a month after laparoscopy and is maintained for 7 months without added medical treatment. Figure 4 showed both the sonographic and laparoscopic findings of one of our case.

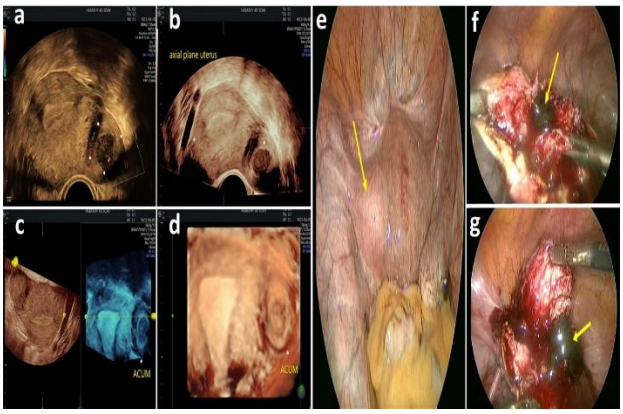

Figure 4: Accessory cavitated uterine malformation (ACUM) (a) 2D-TVUS axial plane of the uterus, (b) 3D-TVUS; minimum mode: axial plane of the uterus, (c) 3D-TVUS; omni-view mode, (d) 3D-TVUS: surface mode, HD-live, (e) laparoscopy for ACUM before myometrial dissection, showing a rounded lesion at the left cornu of the uterus (the yellow arrow), (f and g) chocolate like material extruded from the ACUM after dissection (the yellow arrows).

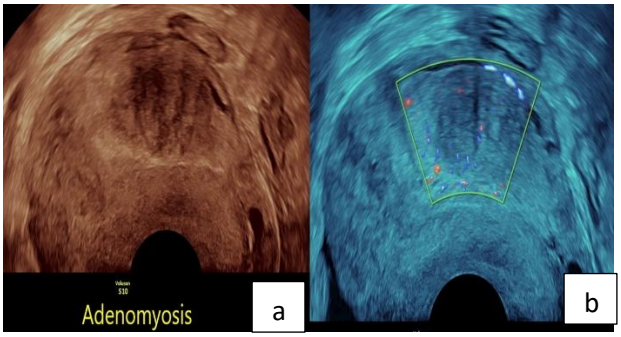

Figure 5 (a and b): Sonographic findings in adenomyoma. Hetrogenous ill-defined solid lesion within the myometrium with translesional minimal flow on Doppler mapping. Associated diffuse adenomyosis.

TVUS features of the 10 cases of adenomyoma were hetrogenous ill-defined solid lesion within any area of the myometrium with translesional minimal flow on Doppler mapping. Associated diffuse adenomyosis was present in all of our cases. All cases also have enlarged uterus (uterine length >10 cm). The size of adenomyomas in our cases was variable ranging from 2×2 cm to 7×6 cm. Figure

5 showed the previously mentioned sonographic description of adenomyoma. The 10 cases who had adenomyosis had underwent hysterectomy due to persistence of symptoms that did not respond to conservative measures. Diagnosis of adenomyoma with adenomyosis was confirmed in these cases postoperatively.

DISCUSSION

ACUM is an extremely rare Mullerian anomaly with less than 150 cases reported in literature.[1] Due to its rarity; it is commonly misdiagnosed as adenomyoma. Figure 6 showed diagrammatic depiction of ACUM with list of its seven sonographic features that we had found.

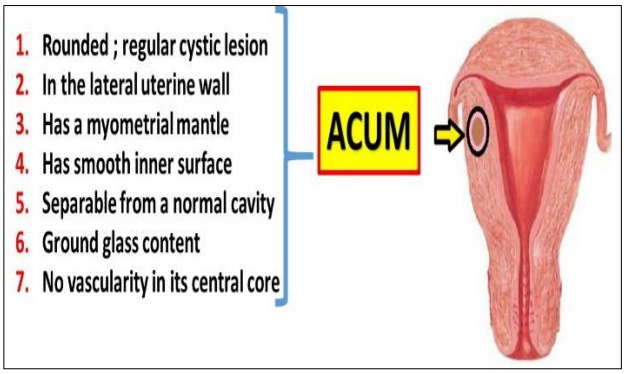

Figure 6: Diagram of ACUM with the seven sonographic features for its diagnosis.

Table 1: Differential diagnosis of ACUM and adenomyoma.

Variables	ACUM	Adenomyoma
Epidemiology	Rare	More common
Nature	Cystic	Solid
Shape	Rounded	Ill-defined
Outline	Well-defined	Ill-defined
Contour	Smooth	Ill-defined
Rim (edge)	Echogenic myometrial mantle	No rim
Core echogenicity	Ground glass	Mixed echogenicity
Vascularity	Circumferential	Translesional
Endomyometrial junctional zone (EMJZ)	Regular (distinct)	Irregular (ill-defined; indistinct)
Associated adenomyosis	No	Common
Treatment	Excision is curative	Excision is useless
	Rarely need hysterectomy	Commonly need hysterectomy

In our cases the delay between the start of symptoms and definitive diagnosis was about 2 years; and this delay is commonly reported in literature for ACUM due to rarity of this pathology which leads that many gynaecologist and sonographers did not know it and misdiagnose it as other pathology.[1] Interestingly noted that the age of our 3 cases of ACUM was >30 years and this is contradictory with Takeuchi et al definition of ACUM who stated that it occurs in females <30-year-old.[2]

Due to its rarity it is usually misdiagnosed as adenomyoma. Other differential diagnosis of ACUM beside adenomyoma are four conditions; which are: myoma, unicornuate uterus with functioning non-communicating horn, Robert's uterus and interstitial ectopic pregnancy.[16-18]

Backow and Tokgoz et al had described the sonographic features of ACUM in 13-year-old and 17-year-old adolescents respectively presented with disabling dysmenorrhea.[14,15] They described it as a cystic mass within the lateral myometrial wall that contained blood products. This mass was adjacent to a normal shaped uterine cavity. Intraoperative and postoperative assessment revealed cystic cavity containing thick brown fluid consistent with old blood, lined by endometrium and surrounded by myometrium. The findings of Backow and Tokgoz et al are comparable to our findings as regard the sonographic description, intraoperative and postoperative assessment.

Peyron et al described the histopathological evaluation of ACUM in eleven cases who were diagnosed by MRI. They described ACUM as a concentric organization of smooth muscle around a cavity lined by ectopic endometrium. Their pathological description of ACUM specimen was matched with our findings.[17]

Limitations

We have two limitations in our study. Firstly; we had included only three cases of ACUM. This limitation can be explained by the extreme rarity of this condition; as less than 150 cases had been reported in literature till now.[1] The second limitation is that we did not classify the type of focal adenomyosis in the 2nd group of our study according to the five types that published by Haesen et al.[19] This was due to that classification had been published after completion of our study.

CONCLUSION

Accessory cavitated uterine malformation is a rare uterine malformation that is usually misdiagnosed as adenomyoma. Distinction between these two pathologies are important as the treatment of both differ. ACUM is suggested in patients with pelvic pain when there is a myometrial cystic lesion seperable from a normal uterine cavity and contained a ground-glass material.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

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Cite this article as: Elhabashy A, Elhabashy M, Salem H. Accessory cavitated uterine malformation: an underdiagnosed Mullerian anomaly. Int J Reprod Contracept Obstet Gynecol 2025;14:698-702.