Accessory and Cavitated Uterine Mass: A Rare and Unclassified Mullerian Anomaly

Maria Zenaida B. Lu, MD and Marian Capco-Dichoso, MD, FPOGS, FPSRM
Department of Obstetrics and Gynecology, De La Salle University Medical Center

Accessory and cavitated uterine mass is a rare developmental mullerian anomaly theorized to be related to gubernaculum dysfunction. It presents typically in young women as severe dysmenorrhea and chronic pelvic pain refractory to medical therapy. It is an accessory cavity lined by functional endometrium and surrounded by myometrium-like smooth muscle located in an otherwise normal uterus, typically located at the right anterior wall at the level of the round ligament attachment. Ultrasound, hysterosalpingography and magnetic resonance imaging are helpful tools to diagnose and distinguish this entity from a wide array of differential diagnoses. Surgical excision and histopathologic studies confirm the diagnosis and effectively relieves severe dysmenorrhea and chronic pelvic pain. This is a case of a 39 year old nulligravid who presented with severe dysmenorrhea initially diagnosed as rudimentary horn versus myoma. Excision revealed a cavitated mass containing chocolate-colored fluid within located at the right postero-fundal area. Histopathology revealed a diagnosis of accessory and cavitated uterine mass.

Key words: accessory and cavitated uterine mass, mullerian anomaly, dysmenorrhea

Introduction

Dysmenorrhea is a cyclic, painful cramping sensation in the lower abdomen during or just before menses. Identifiable causes of dysmenorrhea include gynecologic pathologies and non-gynecologic conditions such as mental, gastrointestinal and genitourinary disorders.[1] Accessory and Cavitated Uterine Mass (ACUM) is a rare Mullerian anomaly that causes severe dysmenorrhea refractory to medical management.

Acien, et al. in 20102 first suggested that most published cases of juvenile cystic adenomyomas, noncommunicating accessory uterine cavities, as well as uterus-like masses are the same pathology representing a new type of Mullerian anomaly called accessory and cavitated uterine mass. This accessory mass with a cavity lined by a functional endometrium and filled by chocolate-colored fluid is typically located at the anterior wall at the level of insertion of round ligament. It is an entity that is underdiagnosed due to its broad differential diagnoses that include rudimentary horn and other uterine malformations, as well as cystic degeneration of adenomyosis, adenomyoma or leiomyoma.[2] To the best of the authors' knowledge, less than a hundred cases have been published as ACUM and none has been reported in the Philippines.

This report aimed to raise awareness of this pathology and its appropriate term, Accessory and Cavitated Uterine Mass (ACUM), and contribute to its clinical evidence. In doing so, the ultimate goal is to help clinicians in making the correct diagnosis in a timely manner and select the best surgical treatment for the patient.

The Case

This is a case of a 39 year old nulligravid who presented with dysmenorrhea. She had her menarche at 14 years old and has had regular cycles lasting for 4 to 5 days, using 2-3 moderately-soaked pads per day. She noted dysmenorrhea since her menarche that started 4 days prior to her menses and lasted until 4 days after her menses. Intake of celecoxib provided some relief. The dysmenorrhea remained uninvestigated until two years prior to admission when she noted increase in the intensity of the pain. Upon consult, a transrectal ultrasound was requested revealing the presence of a well-circumscribed hypoechoic mass described as subserous <50% intramural myoma measuring 3.83cm x 3.68cm x 3.63cm. She was offered surgical management but opted for medical management hence given unrecalled hormonal therapy in the form of combined oral contraceptive pills. The pills along with celecoxib provided little relief and the patient was lost to follow-up.

A year prior to admission, the patient noted persistence and worsening of dysmenorrhea leading to another consult wherein a repeat ultrasound (Figure 1) noted two well-circumscribed hypoechoic masses 1) posterior uterine wall (subserous <50% intramural measuring 1.18cm x 0.92cm x 0.71cm) 2) attached to the right uterine corpus measuring 3.67 cm x 3.76 cm x 2.92 cm with an endometrial like cavity lined by a 0.77 cm thick endometrium and dilated to 1.18 cm containing fluid of low-level echoes. These were diagnosed as myoma uteri and bicornuate uterus with a non-communicating right uterine horn and hematometra. She was then referred to a reproductive medicine specialist who suggested magnetic resonance imaging for confirmation of the uterine anomaly and surgical management but the patient was still hesitant and opted to continue her combined oral contraceptive pills and celecoxib which provided little relief.

In the interim, she noted intermittent right upper quadrant pain. A whole abdomen triple contrast CT-scan was done, which revealed gas-containing gallbladder stones and a right adnexal mass. Both kidneys were visualized and were normal. There was also note of persistence of dysmenorrhea hence repeat pelvic ultrasound was done noting a normal sized anteverted uterus with intact endometrium and

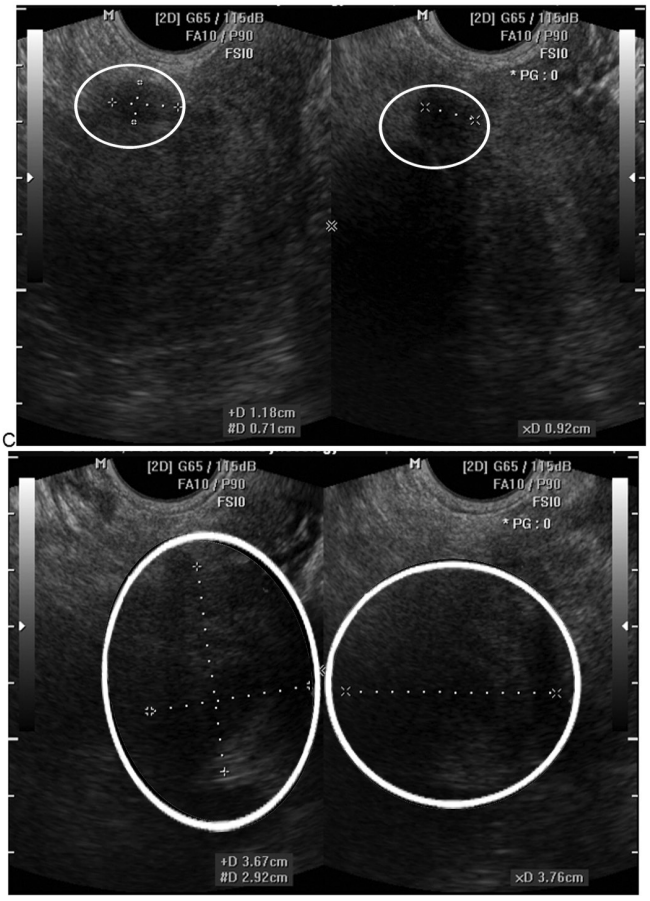

Figure 1. Sonographic pictures of the myoma uteri (top) and right uterine horn (bottom).

a right rudimentary horn versus subserous myoma; the cervix and right ovary were normal while the left ovary was not visualized. After an episode of severe pain which prompted consult at the emergency room, the patient consented for surgical management of both the cholelithiasis and the right adnexal mass.

The patient has a history of bronchial asthma (well-controlled), as well as allergy to NSAIDS. Both her parents are asthmatic. Her father is a known hypertensive while her mother is diabetic. The patient is a lawyer in a regional trial court. She is a non-smoker and a non-alcoholic beverage drinker. She has had no history of sexual contact or previously diagnosed gynecologic disease.

At the time of physical examination, the patient was on the second day of her menstrual cycle and had severe dysmenorrhea. She had a soft abdomen with no direct or rebound tenderness. Pelvic examination revealed normal appearing external

genitalia and presence of hymen. Bimanual rectal exam revealed an asymmetrically enlarged (more on the right) tender mass around 5 cm from the anal opening that was compatible with a retroverted uterus approximately 12 weeks age of gestation in size. No bulging mass was palpated on the anterior rectal wall. No adnexal mass or tenderness was noted. The rectovaginal septum was smooth and non-tender.

The initial impression was subserous myoma with degeneration vs. noncommunicating rudimentary horn in a unicornuate uterus; Cholelithiasis. The surgical plan was laparoscopic cholecystectomy followed by myomectomy or excision of rudimentary horn.

After the laparoscopic cholecystectomy, a diagnostic pelvic laparoscopy was performed revealing dense adhesions between the posterior wall of the uterus and rectum. Bilateral adnexa were grossly normal. The uterine mass could not be visualized properly hence a laparotomy was performed. The uterus was asymmetrically enlarged on the right with smooth serosal surface. There was a firm, cream-tan mass noted on the right posterolateral wall of the uterus, just below the insertion of the fallopian tube, measuring 5.5cm x 4.7cm x 4.5cm (Figure 2). Intraoperatively, this was thought to be a subserous myoma. The mass was removed as if doing a myomectomy with series of sharp and blunt dissection. On cut section, within the uterine mass, there was note of a doughy cream-tan mass measuring 1.5cm x 1.0cm x 1.0cm with cavity measuring 1 cm in length containing brownish fluid within and what seemed to be an endometrial lining that was 0.7 cm thick (Figure 3). The gallbladder measured 7.0 cm x 3.0 cm, filled with bile and a pigment stone measuring 1.5cm x 1.5cm.

The postoperative diagnosis was Obstructive uterine anomaly, to consider accessory cavitated uterus; cholelithiasis.

The postoperative course of the patient was unremarkable and she was discharged after 2 days. Histopathology confirmed a mass consistent with accessory and cavitated uterine mass with secretory phase endometrium and adenomyosis (Figure 4), and chronic cholecystitis with cholelithiasis. At present, 8 months after the surgery, the patient remains asymptomatic and has no recurrence of dysmenorrhea.

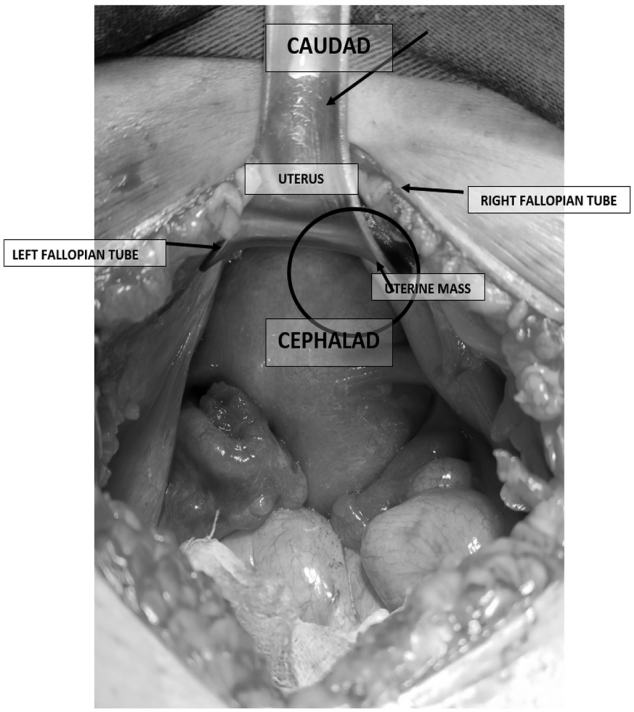

Figure 2. Intraoperative picture

Discussion

Accessory and cavitated uterine mass (ACUM) is a noncommunicating accessory mass consisting of myometrium lined with normal endometrium located within a normal functioning uterus. The pathogenesis of this entity remains controversial; however most authors agree that this is a congenital anomaly. When this anomaly was first reported, the theory was that there was failure of fusion in a small segment of the paired mullerian ducts. Normal fusion occurred both caudal and cephalad to this defect, leaving an isolated, non-communicating, accessory uterine cavity within a normal uterus.[1] Recently, after more cases have been investigated and reviewed, the most widely supported theory is that the accessory mass originated from a dysfunction of the female gubernaculum. The female gubernaculum is an embryonic structure that gives rise to the uterine round ligament and seems to be important in Müllerian development. ACUM is often found at the level of insertion of the round ligament and could be caused by the duplication and persistence of the ductal mullerian tissue in a critical area at the attachment level of the round ligament.[2]

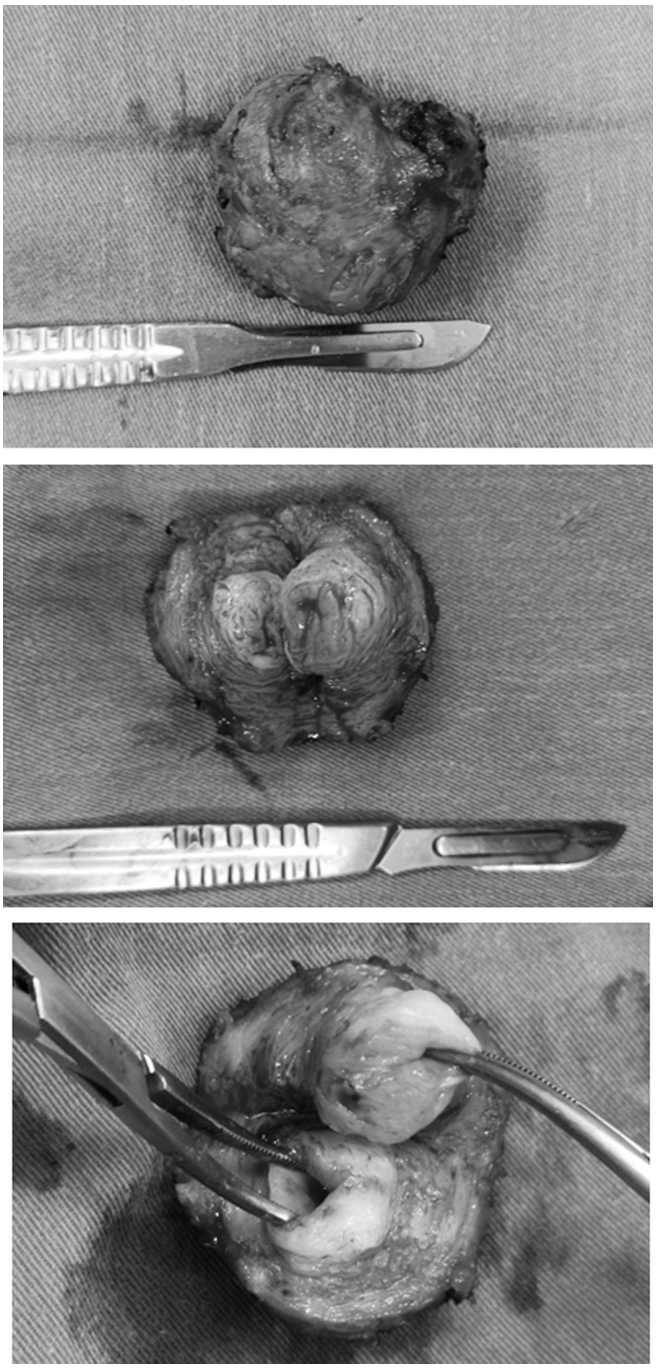

Figure 3. Accessory and cavitated uterine mass

ACUM is not present in both the traditional American Fertility Society and the more recent European Society of Human Reproduction and Embryology (ESHRE)/ European Society for Gynaecological Endoscopy (ESGE) classifications for malformations of the female genital tract. Both classifications are based on uterine anatomical deviations deriving from the same embryological origin. In 2011, Acien and Acien proposed an embryological-clinical classification system that would provide the most appropriate correlation with anatomical findings and clinical presentation and this classification included ACUM (Class 4).[3]

The criteria used to diagnose a case as ACUM was suggested by Acien et al.[2] This consists of 1) An accessory cavitated uterine mass with localization below the insertion of the round ligament, 2) Accessory cavity lined by normal endometrium, 3) Normal main uterus, endometrial cavity, fallopian tubes and ovaries, 4) Chocolate-coloured fluid content in the mass, 5) Surgical case with excised mass and with histopathological confirmation and 6) No signs of adenomyosis in the main uterus, however, tiny foci of adenomyosis may be seen in the myometrium adjacent to the accessory cavity due to increased intracystic pressure during menstruation. All these were present in the index patient.

ACUM was first reported in 1998 by Potter and Schenken who described a case of a 15 year old with non-communicating accessory uterine cavity within a normal uterus.[1] Since then, less than a hundred cases have been reported in literature. In 2020, a search done by Malhotra, et al found 68 cases that met the inclusion criteria for ACUM.[4] In most cases reported, the average age of women diagnosed with ACUM was less than 30 years old. However, several cases of women more than 35 years old were also found in literature.[5,6] All of these patients had their masses excised, and histopathologic evaluation revealed that they were cavities lined by endometrium and surrounded by myometrium. Majority of cases reported the ACUM to be located on the right lateral wall of the uterus adjacent to the round ligament, while other cases of ACUM were reported to be located near the attachment of the left round ligament, in the broad ligament, or in the posterior aspect of the uterus just above the cervix.[4] Almost all cases reported a solitary accessory cavitary mass except for two reported cases of ACUMs noted in the same unilateral area of the uterus. Almost all cases were also found in normal shaped uterus except for one case which reported ACUM in a bicornuate uterus.[5] None of the cases reported associations with urinary tract malformations. For the index case, the ACUM was noted in the right posterolateral uterine wall below the insertion of the round ligament.

There is a tendency to misdiagnose ACUM. Most common symptoms found in patients with ACUM are severe dysmenorrhea and chronic pelvic pain resistant to medical therapy. The non-communicating accessory uterine cavities present clinically with worsening pelvic pain due to progressive increase in the size of the mass, stretching of the endometrial cavity and intracystic bleeding.[3,4] Pelvic ultrasonography is often the initial imaging modality utilized. The uterine masses may appear as solid masses within the uterine wall with variable cystic component resembling an endometrioma. This may be confused as adenomyosis with cystic or degenerated areas, cystic degeneration in leiomyoma or unicornuate uterus with obstructed rudimentary horn. In the case presented, the uterine mass was initially thought to be a leiomyoma with degeneration vs a non-communicating rudimentary horn in a unicornuate uterus.

It is difficult to differentiate ACUM from a unicornuate uterus with obstructed rudimentary horn. Hysterosalpingography (HSG) can help in such cases and demonstrate a fallopian tube arising from an accessory uterine horn with the main uterine cavity having only one fallopian tube in cases of unicornuate uterus with a rudimentary horn. Pelvic magnetic resonance imaging (MRI) is the diagnostic modality of choice for malformations of the genital tract. It is non invasive, hence, is preferred over hysterosalpingography in unmarried females such as in the case of the patient presented. MRI will show the ACUM with hemorrhagic contents along the uterine wall under the round ligament. It will further demonstrate a normal uterus, cornua, and ovaries.[7] Even with the use of pelvic sonography, HSG or MRI, it still may be difficult to accurately diagnose ACUM. ACUM can only be suspected preoperatively and confirmed only after excision and histopathological examination.

Isolated or juvenile cystic adenomyomas (JCA) is another condition also described as a cystic lesion within the uterine wall that is independent of the normal uterine cavity. Takeuchi, et al.[8] established the following diagnostic criteria for JCA: 1) age of patient $\leq$ 30 years, 2) cystic lesion $\geq$ 1 cm in diameter independent of the uterine lumen and surrounded by myometrium, and 3) presence of severe dysmenorrhea.[8] The most important differential diagnosis for JCA is an obstructive uterine anomaly associated with severe dysmenorrhea such as ACUM. Histopathology will differentiate the two entities. On histopathologic examination, JCA lacks endometrial lining in the cystic cavity and the uterus like smooth muscle organization. There will also be presence of diffuse adenomyosis in the main uterus of patients with JCA.[5,7]

Search in local literature did not show any published cases of ACUM or JCA. This is the first local reported case of accessory cavitated uterus.

Early surgical treatment is recommended which includes adequate excision of the uterine mass either via laparotomy or laparoscopy. Acien, et al. who did surgery on numerous patients with ACUM indicated that although laparoscopic excisions can be performed, they preferred laparotomy to adequately access depth of penetration of the uterine mass for complete excision.[5] Excision is done by enucleation of the mass without entering the main endometrial cavity. After surgery, most patients have complete resolution of their dysmenorrhea.

Conclusion

ACUM is an emerging differential diagnosis in women presenting with severe and progressive dysmenorrhea that is refractory to medical therapy. It has remained a diagnosis of exclusion and often diagnosed post-operatively on histopathologic examination. A high index of suspicion with the aid of imaging modalities are necessary to clinch the diagnosis. Pelvic ultrasound and HSG are useful low-cost modalities but MRI remains as the diagnostic modality of choice as it is non-invasive and clearly visualizes the pelvic anatomy. Complete surgical excision is considered the best management and effectively resolves dysmenorrhea.

References

1. Potter DA & Schenken RS. Non-communicating accessory uterine cavity. Fertil Steril 1998; 70(6): 1165-6.
2. Acien P, Acien M, Fernandez F, Mayol M & Aranda I. The cavitated accessory uterine mass: A mullerian anomaly in women with an otherwise normal uterus. J Am Coll Obstet Gynecol 2010; 116(5): 1101-9.
3. Acién P, Acién M. The history of female genital tract malformation classifications and proposal of an updated system, Hum Reprod Update 2011; 17(5): 693–705.

4. Malhotra V, Dahiya S, Nanda S, Chauhan M and Bhuria V. Accessory and cavitated uterine mass. Is it a mullerian duct anomaly? J Gynecol Surg 2020; 36(6): 350-6.
5. Acien P, Bataller A, Fernandez F, Acien M, Rodriquez J & Mayol M. New cases of accessory and cavitated uterine masses (ACUM): A significant cause of severe dysmerrhea and recurrent pelvic pain in young women. Hum Reprod 2012; 27(3): 683-94.
6. Azuma Y, Taniguchi F, Wibisono H, Ikebuchi A, Moriyama M, Harada T. A case report of an accessory and cavitated uterine mass treated with total laparoscopic hysterectomy. Yonago Acta Med 2021; 64(2): 207-9.
7. Supermaniam S, Thye WL. Diagnosis and laparoscopic excision of accessory cavitated uterine mass in young women: Two case reports. Case Rep Women's Health 2020; 26: e00187.
8. Takeuchi H, Kitade M, Kikuchi I, Kumakiri J, Kuroda K, et al. Diagnosis, laparoscopic management, and histopathologic findings of juvenile cystic adenomyoma: a review of nine cases. Fertil Steril 2010; 94: 862-8.