

Diagnosis and laparoscopic excision of accessory cavitated uterine mass in young women: Two case reports

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ABSTRACT

Introduction: An accessory cavitated uterine mass (ACUM) is a rare congenital Mullerian anomaly where an accessory cavity with normal endometrial lining lies within a normally functioning uterus. It is common among young and nulliparous women presenting with severe dysmenorrhea and infertility.

Presentation of the Cases: We present two cases of ACUM. The first case was a 22-year-old woman who presented with severe dysmenorrhea and was initially misdiagnosed with non-communicating rudimentary horn. The second case was a 36-year-old woman who presented with primary infertility and dysmenorrhea. Gynecological examination and ultrasound scanning were done for both patients. Subsequently, laparoscopic excision of the ACUM was performed on both patients. Histopathological examination confirmed the diagnosis. Postoperatively, both patients did well, with no further dysmenorrhea. The second patient conceived spontaneously at the first attempt and at the time of writing was 33 weeks pregnant without any maternal or fetal problems.

Conclusion: The diagnosis of ACUM is often confused with non-communicating rudimentary uterine horn, true cavitated adenomyosis and degenerating fibroids. It is important to understand and distinguish ACUM. A thorough history, detailed gynecological examination and correct radiological modalities are critical to a proper diagnosis so that the correct surgery can be performed, especially when fertility is desired.

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1. Introduction

An accessory cavitated uterine mass (ACUM) is a rare congenital Mullerian anomaly where an accessory cavity with normal endometrial lining lies within a normally shaped and normally functioning uterus. This uterine malformation is different from the common Mullerian uterine malformation described in the ESHRE/ESGE consensus statement [1]. It occurs due to the duplication or persistence of ductal Mullerian tissue, which is believed to have originated from gubernaculum dysfunction, leading to accessory uterine tissue formation. 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 rare among women under 30 years of age. 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. Ultrasound scan, magnetic resonance imaging (MRI) and hysteroscopy are helpful in the diagnosis.

Here, we present two cases of ACUM diagnosed in line with the proposed ACUM criteria [2,3]. The first case was a 22-year-old woman who presented with severe dysmenorrhea and was initially misdiagnosed with non-communicating rudimentary horn. The second was a 36-year-old woman who presented with primary infertility and dysmenorrhea. Laparoscopic excision of the ACUM was performed in both patients.

2. Cases

2.1. Case 1

A 22-year-old, single, virgo intacta (VI) woman presented with a 3-month history of severe pain after menses. She had attained menarche at the age of 15 and had not suffered from dysmenorrhea. Her menstrual flow was regular and normal. Being a VI, we could not perform a transvaginal ultrasound scan (TVUS) and she refused a transrectal ultrasound scan (TRUS). Transabdominal ultrasound scan (TAUS) showed a retroverted normal-sized uterus with normal endometrial cavity. The ovaries were not clearly seen. There was a cystic mass measuring 2.65×3.62 cm on the right side of the uterus, which appeared like a right endometrioma (Fig. 1A).

With a diagnosis of a right endometrioma, we proceeded to perform a laparoscopic cystectomy. Laparoscopy showed a uterus with a

Abbreviation: ACUM, Accessory cavitated uterine mass.

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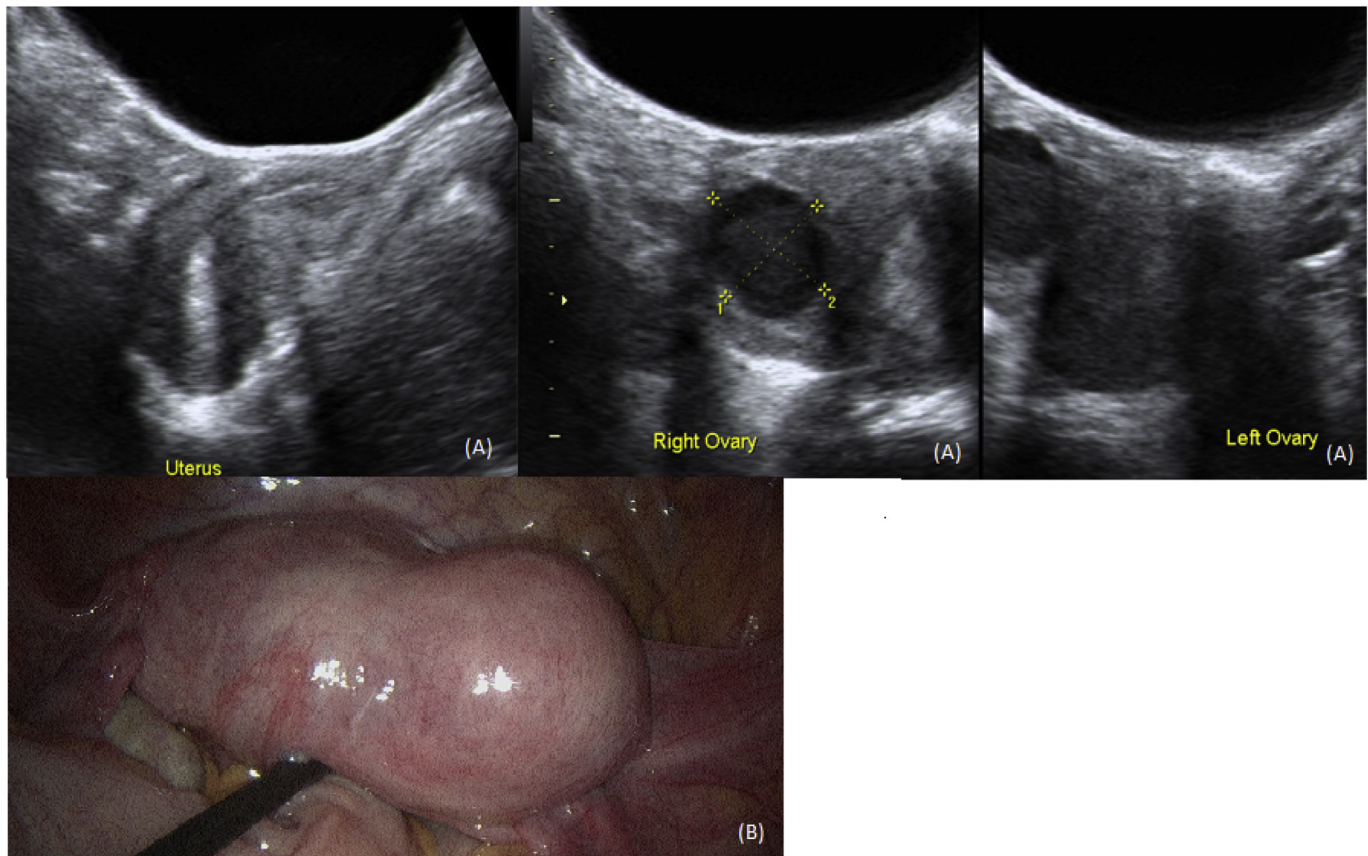


Fig. 1. (A): TAUS shows a cystic lesion on the right side of the uterus. (B): Laparoscopy shows a nodular lesion on the right side of the uterine fundus.

globular swelling in the right fundal area, next to the right fallopian tube insertion (Fig. 1B). Minimal endometriotic nodules were found on the right uterosacral ligament and the posterior aspect of the uterus. Both ovaries and Fallopian tubes were normal. A needle was used to aspirate the mass. Brownish fluid resembling endometriosis was obtained. The mass was thought to be a non-communicating uterine horn. Since no consent was obtained to excise the lesion, this was not performed.

The findings were discussed with the patient. We also advised her to undergo a TRUS and hysteroscopy. 3D TRUS showed a cystic lesion in the right side of the uterus measuring 2.65×3.62 cm (Fig. 2A). MRI scan showed a unicornuate uterus with a non-communicating rudimentary horn (Fig. 2B).

Just before the second laparoscopy, a hysteroscopy was performed (without injuring the hymen). The right tubal ostium was directly cannulated and chromopertubation was performed. Dye was seen coming out of the right fallopian tube (Fig. 2C). This indicated that the right fallopian tube arose directly from the uterus and not from the cystic lesion, thus eliminating the diagnosis of a rudimentary horn. Diluted vasopressin was injected into the myometrium of the nodule and excision of the uterine mass was performed with monopolar diathermy. This was done easily without entering the uterine cavity. The defect was sutured with interrupted polyglactin 1 sutures. The specimen was removed in an *endo*-bag after cutting it into smaller pieces.

Post-operatively, the patient had an uneventful recovery and was discharged 48 h later. During her follow-up, she was asymptomatic and an ultrasound scan showed no abnormal findings.

Histopathological examination confirmed a cystic hemorrhagic mass. The cavity was lined with endometrial tissue with glandular atrophy. These results along with the operative findings confirmed the diagnosis of ACUM.

2.2. Case 2

A 36-year-old married nulligravida woman presented with a history of chronic pelvic pain and severe dysmenorrhea. She had attained menarche at 12 years of age and had been having dysmenorrhea since then. She experienced intermittent severe colicky pain during the first 3 days of menses every month. For the last 4 years she had been trying to conceive but had been unsuccessful. Her husband's semen analysis was normal.

No abnormalities were detected on abdominal and pelvic examination. TVUS and TAUS revealed a normal-sized uterus with a cavitated right-sided intramural mass measuring 3.29×3.28 cm (Fig. 3). The cavitated mass contained echogenic homogenous material resembling an endometrioma measuring 1.01×1.77 cm. The uterus, endometrial layer and ovaries were normal. The antral follicle count was 2 in the right ovary and 1 in the left ovary. The mass was thought to be either an ACUM or a degenerating fibroid. Anti-Mullerian hormone (AMH) was 0.91 ng/ml.

Since her AMH level was low, we discussed several treatment options. Her main concern was preservation of fertility. Since the cystic lesion did not involve the endometrial cavity, we discussed the option of performing in-vitro fertilization (IVF) first, with embryo transfer and surgery later. Due to time constraints, she decided to do a cycle of IVF, freeze the embryos and then undergo a laparoscopic excision of the ACUM. We thought that frozen embryo transfer after the surgery may give her a better chance of implantation. She was started on an antagonist protocol. Unfortunately, we only collected 2 oocytes, which resulted in a single embryo, which was frozen. We suggested a second IVF cycle to collect more embryos before surgery but she chose to undergo the surgery without this.

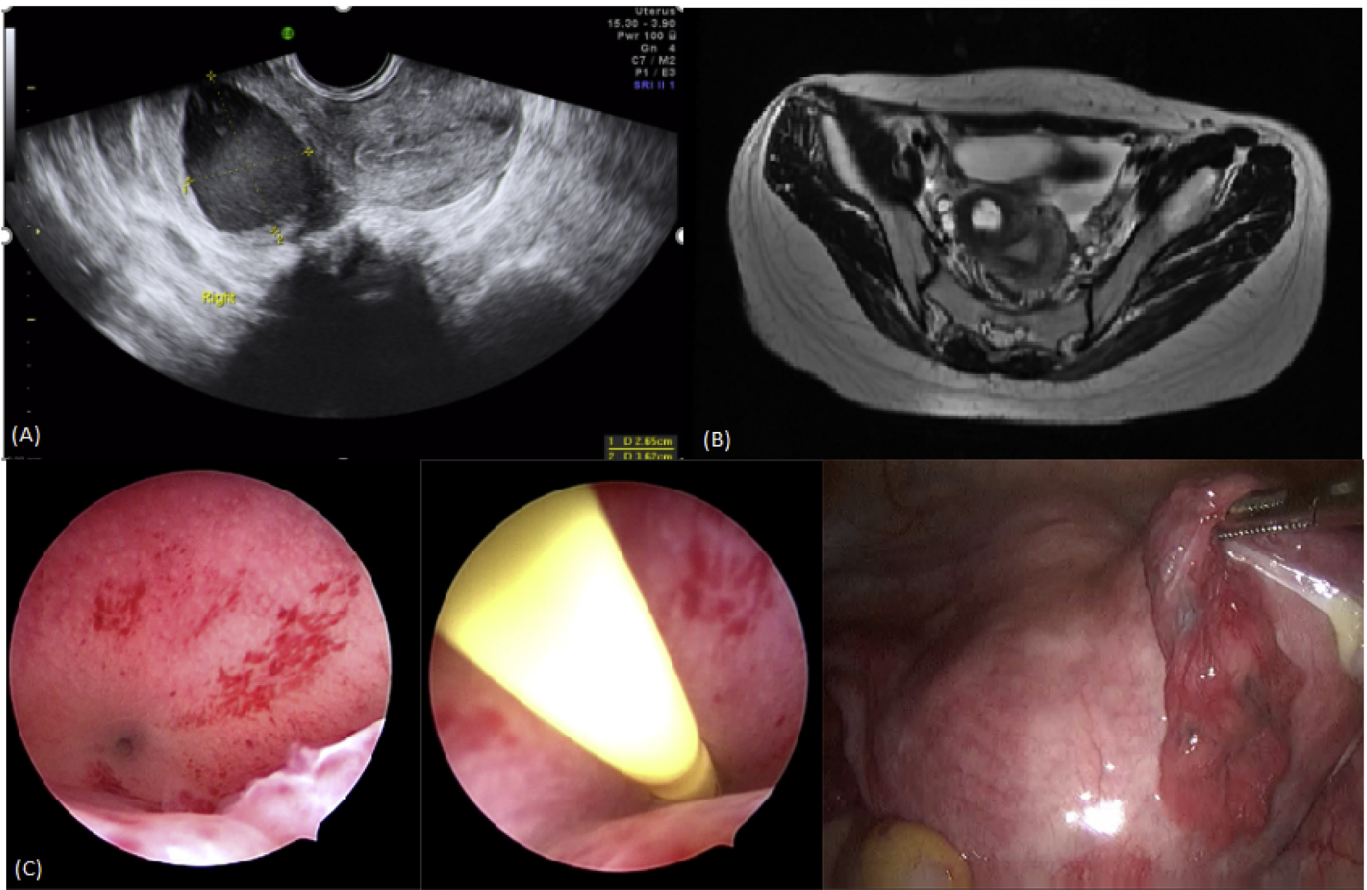


Fig. 2. (A): TRUS shows a cystic lesion on the right cornual part of the uterus. (B) MRI shows a cystic lesion on the right side of the uterus. There was no communication between the endometrial cavity and the cystic lesion. (C) Showing hysteroscopic cannulation of the right tubal ostium and chromopertubation showing that the right tube is patent.

Hysteroscopy revealed a normal uterine cavity and both ostia were seen. On laparoscopy, a nodular lesion attached to the right anterior uterine wall close to the round ligament insertion was seen (Fig. 4A).

Endometriotic lesions were found on the posterior peritoneum of the broad ligaments, uterosacral ligaments, rectum and over the

ureters. The rectum was adherent to the uterus posteriorly. The uterus, both fallopian tubes and ovaries were otherwise normal. Chromopertubation with methylene blue injection confirmed that both tubes were patent. Laparoscopic adhesiolysis was performed. The ureters were dissected out. The rectum was released from the adhesions in the rectovaginal septum. Nodules on the uterosacral

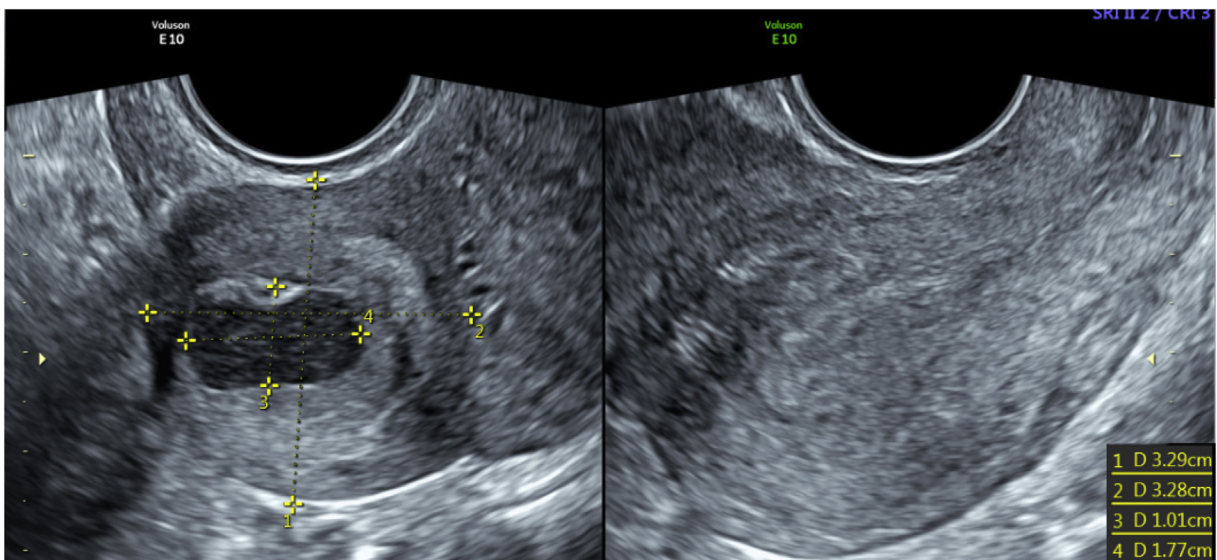


Fig. 3. TVUS shows a cystic mass on the right side of the uterine fundus.

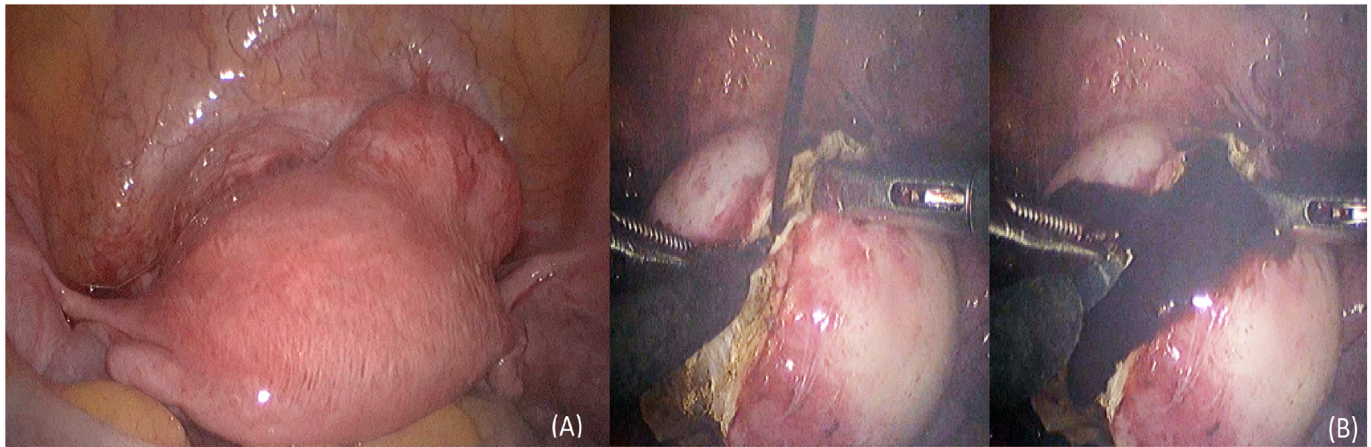


Fig. 4. (A): Nodule seen on the right side of the anterior wall of the uterus. (B): As the nodule was incised, a chocolate-colored material spilled out.

Table 1
Characteristics of ACUM and its differential diagnoses.

	Accessory cavitated uterine mass	Non-communicating rudimentary uterine horn	True cavitated adenomyoma	Degenerating fibroid
Pathophysiology	Mullerian anomaly without uterine malformation caused by duplication and persistence of the ductal Mullerian tissue at the insertion of the round ligament, believed to be due to the gubernaculum dysfunction.	Mullerian anomaly with uterine malformation due to the failure of one of the Mullerian duct to elongate towards the urogenital sinus while the contralateral Mullerian duct develops normally [9].	Invagination of the endometrial basal layer into the myometrium [7]. Misplaced pluripotent Mullerian remnants [8].	Benign monoclonal tumor mainly composed of smooth muscle cells and fibrous connective tissue.
Definition	Isolated cavitated mass consistent with the normal myometrium resembling the normal uterus. It is found at the level of round ligament insertion. It is associated with a normal shaped and functional true uterus.	Usually associated with uterine malformation (unicornuate uterus, bicornuate uterus). 74–90% of unicornuate uterus associates with rudimentary uterine horn [5,6]. An isolated cavity with a horn and fallopian tube attaching on it without communicating with the true cavity.	A focal adenomyosis which is not in direct continuity with the junctional zone. Has central degeneration and endometriotic fluid accumulation. Common among elderly women. Often associated with fibroids.	Benign tumor of myometrial origin with cystic lesions in the middle of the tumor. Fibroids can undergo various forms of degeneration like hyaline, cystic, myxoid and red degeneration.
HPE findings	Endometrioid epithelium line the cavity. Epithelial glands and stroma lined the cavity, surrounded by smooth muscle cells. Myometrium adjacent to the ACUM may develop adenomyosis but do not present in the rest of the uterus (small foci).	Thick myometrial wall. Cavity lined by endometrial epithelium. No evidence of adenomyosis.	Absence of internal epithelial lining of the cystic cavity. Lack of uterus like smooth muscle organization. Diffusely spread adenomyotic foci in the uterus corpus.	It is well circumscribed with solid rubbery firm texture [10]. The color and texture may be different based on the types of degeneration. Composed of fascicles of elongated smooth muscle cells with eosinophilic cytoplasm and centrally located cigar-shaped nucleus. It is rich in vasculature of various calibers and types including muscle-rich arteries, arterioles and veins.
Fallopian tubes	Normal ostia. Normal fallopian tubes. The uterine horn is attached to the true uterus cavity.	The fallopian tube arises from the accessory uterine horn. The true uterine cavity only has one fallopian tube. Hysteroscopy shows a single tubal ostium.	Fallopian tubes arise from the uterus.	Fallopian tubes arise from the uterus.
MRI findings	A nodular uterine lesion with central cavity containing cystic component and hemorrhagic content within it. Hyperintense signal on T1-weighted images. Hypointense signal on T2-weighted images and clearly independent from the normal endometrial cavity.	The unicornuate uterus is displaced off the midline with normal myometrial zonal anatomy and normal endometrial-to-myometrial width and ratio [4]. If the non-communicating rudimentary horn contains endometrial tissue, the zonal anatomy is preserved while the rudimentary horn may become distended with blood products. In an empty non-communicating rudimentary horn, the zonal anatomy is absent with a diffuse low signal intensity shown on the uterine horn.	Hypointense mass on T2-weighted images. Ill-defined borders, minimal mass effect, multiple bright foci.	Cystic degeneration shows isointense relative to myometrium on T1; hyperintensity with lack of contrast enhancement of the internal areas on T2 [10]. Hemorrhagic degeneration shows hyperintensity on T1 and moderate-to-high intensity on T2. Hyaline degeneration appears isointense on T1, hypointense on T2; similar to non-degenerated fibroids. Fatty degeneration appears consistent with fat on MRI.

ligaments, rectum, posterior wall of the uterus and peritoneum were excised. Vasopressin 20 IU diluted in 200 ml of saline was injected into the myometrium at the junction of the uterus and the ACUM nodule to achieve hemostasis. An oblique incision was made on the mass. Approximately 20 ml of thick, chocolate-colored material spilled out of the mass (Fig. 4B).

Circumferential incision of the mass was done using monopolar diathermy. The nodule was excised without entering the true endometrial cavity. The defect was then sutured with interrupted polyglactin 1 sutures. The specimen was placed in an *endo*-bag, cut into smaller pieces and retrieved from the peritoneal cavity through a 10 mm trocar site. Anti-adhesion barrier agent was applied to all areas where the surgery was performed. Post-operatively, the patient did well. She was discharged the following day.

The endometriotic nodules and the uterine nodule (ACUM) were sent for histopathological analysis. The first specimen was consistent with endometriotic nodules. The histopathology of the second specimen showed the presence of tubular and mildly dilated endometrial glands with stroma between bundles of smooth muscle fibers. Areas of myxohyaline changes were found adjacent to the stroma with no adenomyotic foci. A significant cellular atypia surrounding the myometrial tissue in the specimen was noted. The histopathological findings were consistent with the diagnosis of ACUM.

After surgery, the patient had no further dysmenorrhea. She was advised not to conceive for 6 months and to undergo frozen embryo transfer after that. However, she conceived successfully on her first attempt. At the time of writing she was 33 weeks pregnant without any maternal or fetal problems.

3. Discussion

ACUM has been documented in the literature, but using different terminologies, such as juvenile cystic adenomyosis, non-communicating accessory uterine cavities and isolated cystic adenomyoma. It can be confused with several other gynecological diseases. Table 1 summarizes the characteristics of ACUM and its differential diagnoses.

Based on the criteria [2,3], ACUM diagnosis can only be suspected preoperatively and confirmed only after histopathological examination. However, accurate preoperative diagnosis is important to determine the need for and type of surgery, as this will differ in different conditions. The 2 cases reported here demonstrate that ACUM is readily managed with surgery, as the endometrial cavity is not entered.

There are several lessons that can be learnt from these 2 cases. In the first case, a diagnosis of right endometrioma was made only with a TAUS. Endometrioma can usually be diagnosed confidently with only a TVUS, with no need for MRI. Since this patient was VI, TVUS could not be performed. TRUS will give an equally clear picture of the pelvis and could have distinguished an endometrioma from a uterine cystic lesion. So, if a VI patient refuses TRUS, then MRI is mandatory to exclude ACUM. An office hysteroscopy and/or hysterosalpingography is another way of differentiating an ACUM from a rudimentary uterine horn (Table 1). These tests were not done in this patient, as she was VI.

In the second case, ACUM was the provisional diagnosis from the beginning. Excision of the ACUM was done after 1 cycle of IVF. Fortunately, the patient conceived spontaneously after the excision of the ACUM and endometriotic nodules in the pelvis. It is difficult to postulate whether the excision of the ACUM and/or the endometriotic nodules in the pelvis was the reason she conceived spontaneously.

4. Conclusion

ACUM is a difficult condition to diagnose. One needs a high index of suspicion. Since most patients with ACUM suffer from dysmenorrhea, surgical excision is necessary and can be easily done by laparoscopy. When a cystic lesion seen in the myometrium is asymptomatic, it is difficult to distinguish ACUM from its differential diagnoses (Table 1) and a correct diagnosis can be made only after excision and histopathological evaluation.

Contributors

Sevellaraja Supermaniam is the corresponding author and was the surgeon in charge.

Wei Lin Thye was in charge of collecting and assembling the data.

Both authors participated in the writing and critical revision of the manuscript and both read and approved the final version.

Conflict of Interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

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Patient Consent

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Provenance and Peer Review

This case report was peer reviewed.

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