Case Report

Accessory Cavitated Uterine Mass: A Rare Cause of Severe Dysmenorrhea in Young Women

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ABSTRACT

We present 3 case reports of a rare Müllerian anomaly called accessory and cavitated uterine mass (ACUM), which is found in young women. They presented with severe dysmenorrhea refractory to medical treatment. The patients were 17, 19, and 25 years old. The patients had the classic Müllerian anomalies. The hysteroscopic examination was normal in all 3 cases, and laparoscopic examination showed a 3- to 4-cm ill-defined mass on the right half of the uterus, without any communication to the uterine cavity. The chocolate-colored material was drained in all of the cases, during excision of the mass. The myometrial defect was sutured laparoscopically. On histological examination, the mass was found to be a cystic cavity, lined by endometrial glands and stroma, which confirmed the diagnosis of ACUM. Journal of Minimally Invasive Gynecology (2015) 22, 1300–1303 © 2015 AAGL. All rights reserved.

Keywords: Accessory and cavitated uterine masses (ACUMs); Dysmenorrhea; Müllerian anomaly

DISCUSS

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Dysmenorrhea is a common condition that occurs in 79.6% of adolescents [1]. Pelvic abnormalities, such as endometriosis or uterine anomalies, may be found in approximately 10% of adolescents and young women with severe dysmenorrhea symptoms [2–4]. Patients are diagnosed by imaging with ultrasonography (USG) and magnetic resonance imaging (MRI). We had 3 cases of severe dysmenorrhea that were preoperatively diagnosed as classic Müllerian anomalies. After hysteroscopy and laparoscopic evaluation, they turned out to be the accessory and cavitated uterine masses (ACUMs). This condition presents a diagnostic challenge and should be considered in the differential diagnosis of severe dysmenorrhea in young women. Acien et al [5,6] showed that the total number of cases of ACUMs reported in the literature by different authors, including their study, was 37 (18 cases reported in 2010 and 19 more cases reported in 2012). We report 3 additional cases of ACUMs.

Informed Consent

All procedures followed the ethical standards of the responsible committee on human experimentation (institutional and national) and the 1975 Declaration of Helsinki, and as revised in 2000. Informed consent was obtained from all patients.

Case 1: 2014

A 19-year-old celibate woman presented with a complaint of chronic lower abdominal pain and severe dysmenorrhea of 4 years’ duration. The pain was severe, intermittent, and colicky and associated with menstruation. The pain was not associated with gastrointestinal or urinary symptoms. She was treated with oral contraceptive pills (OCPs) and nonsteroidal anti-inflammatory drugs without relief.

On transabdominal USG (TAS), she was diagnosed with uterine bicornis with hypoplastic right cornua. MRI findings were suggestive of uterine bicornis with a right horn hemotoma of 2 cm. Because of these findings, the patient was...
referred to our laparoscopy center for further treatment. Diagnostic hysteroscopy revealed a normal cavity and normal bilateral ostia (Fig. 1). A primary infra-umbilical 10-mm port and 2 ancillary 5-mm trocars, lateral to the right and left epigastric vessels, and a median suprapubic trocar were inserted. On laparoscopy, the uterus was found to be enlarged asymmetrically, with marked globular enlargement on the right anterior wall, near the attachment of the round ligament. No other uterine horn could be seen. Both the fallopian tubes and ovaries were normal in appearance. Vasopressin 20 IU diluted in 80 mL of saline was injected into the myometrium to reduce the bleeding. A transverse incision was made on the anterior uterine surface over the swelling with the Harmonic Ace scalpel (Ethicon Endo-surgery, Cincinnati, OH). Approximately 5 mL of a thick, chocolate-colored material was drained on application of a myoma screw (Fig. 2), and the mass was dissected with the Harmonic Ace scalpel and excised (Fig. 3). The uterine cavity was not opened during dissection. The broad ligament defect was closed with the 1-0 braided Lactomer (Polysorb, Tyco Healthcare, Mansfield, MA) suture in 2 layers (Fig. 4). The specimen was retrieved from the peritoneal cavity through the 10-mm trocar (Fig. 5). There were no intraoperative or postoperative complications. The patient was discharged the next day after surgery. The patient did not experience dysmenorrhea in her next cycle when she came for follow-up.

Macroscopic pathologic examination of the lesion revealed a 3-cm irregular grayish white nodular mass with a blood-filled cyst of 2 cm diameter. On histological examination, the mass was found to be a cystic cavity, lined by endometrial glands and stroma, which confirmed the diagnosis of ACUM. There were no adenomyotic foci surrounding the myometrial tissue in the excised specimen.

Case 2: 2010

A 17-year-old adolescent female presented with dysmenorrhea that was refractory to medical treatment for 2 years. Ultrasound revealed a 4- × 3-cm hypoechoic mass in the posterior wall of the uterus. Diagnostic hysteroscopy revealed a normal cavity and normal bilateral ostia. Laparoscopic examination showed an asymmetrically enlarged uterus, with globular enlargement on the posterior surface of the uterus that extended to the right broad ligament. An oblique incision was made in the mass on the posterior wall of the uterus (Fig. 6). Thick, chocolate-colored material was drained with application of a myoma screw on the mass. The mass was dissected with a Harmonic Ace scalpel after safeguarding the right uterine vessels. The myometrium was closed with Quill SRS suture (Surgical Specialties Corporation, Wyomissing, PA) in 2 layers. The specimen was retrieved from the peritoneal cavity. There were no intraoperative or postoperative complications. On histological examination, the mass was found to be a cystic cavity, lined by endometrial glands and stroma, which confirmed the diagnosis of ACUM. We followed this patient yearly for 3 years, and the patient had only mild dysmenorrhea.

Case 3: 2007

We had a 25-year-old woman with a complaint of dysmenorrhea that was nonresponsive to medical treatment
for 7 years. TAS revealed a right-sided mass with mixed echoes of 3.1 cm suggestive of an obstructed rudimentary horn. Diagnostic hysteroscopy revealed a normal cavity and normal bilateral ostia. Laparoscopic examination showed that the uterus was enlarged asymmetrically, with a globular enlargement of 3 cm near the right cornu of the uterus (Fig. 7). A vertical incision was made on the globular mass. Thick, chocolate-colored material was drained with application of a myoma screw on the cystic lesion. The mass was excised with the Harmonic Ace scalpel. The myometrium was closed with the Quills SRS suture in 2 layers. The specimen was retrieved from the peritoneal cavity. There were no intraoperative or postoperative complications. Histological examination showed the mass to be a cystic cavity, lined by endometrial glands and stroma, which confirmed the diagnosis of ACUM. We followed this patient yearly for the last 7 years, and the patient currently has only mild dysmenorrhea.

Discussion

Management of severe dysmenorrhea is a common clinical problem. Rarely, this can be due to a recently described clinical entity called ACUM. We reported 3 cases of ACUM; 1 was an adolescents and 2 were a young women who were provisionally diagnosed with a noncommunicating rudimentary horn. This terminology was first described first by Acien et al In 2007, Takeda et al [7] described this condition as juvenile cystic adenomyosis.

The pathognomonic clinical feature of juvenile cystic adenomyoma or ACUM is its early onset of severe dysmenorrhea, which usually starts soon after menarche. This symptom could be attributed to intracystic bleeding and stretching of the cystic cavity [5,6]. The criteria used to diagnose ACUM were (1) an isolated accessory cavitated mass; (2) normal uterus endometrial cavity, tubes, and ovaries; (3) a surgical case with an 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 the uterus was removed), but there could be small foci of adenomyosis in the myometrium adjacent to the accessory cavity, as described by Acien et al [5,6]. This entity is unlikely to be adenomyosis in young women. An adenomyoma lacks the typical endometrial lining and is associated with
adenomyotic changes in the rest of the uterus, unlike ACUM [5,6]. Formation of a blind horn from these endometrium-lined cavities is very unlikely, because the uterine cavity, both ostia, and the tubes are normal. Therefore, ACUM is probably a separate clinical entity different from other classic Müllerian anomalies. Although ACUM is diagnosed more frequently in women <30 years of age and nulliparous women, some cases in women >30 years and multiparous women have also been reported [5,6]. However, our 3 patients were 17, 19, and 25 years old, and they satisfied the criteria for ACUM.

Early occurrence of the symptoms soon after menarche suggests that juvenile cystic adenomyoma is a congenital malformation, which is caused by a defect in the developmental process of Müllerian ducts. The most common location of this lesion is the anterior wall of the uterus at the level of insertion of the round ligament [5]. An accessory uterine mass could be caused by duplication and persistence of ductal Müllerian tissue in a critical area at the attachment level of the round ligament, which is possibly related to a gubernaculum dysfunction [5,6]. Female gubernaculum (which later forms a round ligament) formed by muscle fibers that are not mesonephric or paramesonephric in origin, and their attachment to Müllerian ducts, allows or induces the fusion and adequate development of the uterus [8]. In 2 of our patients, the location of the ACUM was on the anterior surface of the uterus near the cornual end on the right side at the level of insertion of the round ligament. In the second case, it was more posterior near the right cornu.

Diagnosis of ACUM is difficult, and is done by the exclusion of known causes of dysmenorrhea-like diffuse adenomyosis, endometriosis and Müllerian duct anomalies. USG and MRI are useful in diagnosing these conditions. Conventional hysterosalpingography and hysteroscopy can exclude a noncommunicating rudimentary horn. In all 3 of our patients, a normal cavity and both normal ostia were seen on hysteroscopy, and the actual diagnosis was made during laparoscopic exploration of the mass when the chocolate-colored material was drained from the localized cavity. Regarding management, these 3 patients were already treated for severe dysmenorrhea with OCP and progestin before surgery for 2, 4, and 7 years, respectively, without relief.

Acien et al mentions treatment with OCP in their publications, but does not mention progestins or gonadotropin-releasing hormone. All of their patients were managed by surgical resection by laparotomy, laparoscopy, or robot-assisted laparoscopy [5,6,9]. Surgical excision provided a significant improvement in dysmenorrhea and pelvic pain. We managed all our cases by laparoscopic excision. For complete symptom relief, complete excision of the mass is required. Due consideration has to be given to the myometrial integrity of the uterus for a safe obstetric outcome. Our patients were young and had not started their obstetric life, and after surgery they had significant pain relief.

**Conclusion**

ACUM or juvenile cystic adenomyoma is a rare Müllerian anomaly in women with an otherwise normal-appearing uterus on imaging and hysteroscopy. It is a rare and often misdiagnosed cause of refractory dysmenorrhea and chronic pelvic pain in young women. The true incidence of this anomaly at present is not available. We should be aware of this Müllerian anomaly while managing these young patients with severe dysmenorrhea. More reporting of such cases is needed. Laparoscopic surgery in expert hands is a good diagnostic and therapeutic option for this condition.

**References**
