New cases of accessory and cavitated uterine masses (ACUM): a significant cause of severe dysmenorrhea and recurrent pelvic pain in young women

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Introduction

In a recent publication (Acién et al., 2010), we suggested that most of the published cases of non-communicating accessory uterine cavities and juvenile or isolated cystic adenomyomas, as well as some cases of uterus-like masses, were actually the same pathology: an accessory and cavitated uterine masses (ACUM) with a functional endometrium. In that article, we reviewed the literature and found 18 cases (including 4 cases reported by us) that we considered to be ACUMs but had been described as accessory uterine cavities, isolated or juvenile cystic adenomyomas (JCAs), or as uterus-like masses. We found another 11 cases that were questionable. It has been suggested that ACUMs represent a new variety of Müllerian anomaly that is generally located at the level of the insertion of the round ligament and is possibly related to a dysfunction of the female gubernaculum (Acién et al., 2011). However, ACUMs have been generally observed in women with an otherwise normal uterus and they are single, but we will see that probably this is not always so.

An ACUM is a rare pathology, observed in young women, that has significant clinical manifestations, particularly severe dysmenorrhea and recurrent pelvic pain. ACUMs are problematic because of the broad differential diagnosis, which includes rudimentary and cavitated
tics (Acien et al., 2010) because they present similar clinical and histopathological characteristics and localization of the ACUM for quickly choosing the best surgical treatment.

The objectives of this report are as follows: (i) to raise awareness about this pathology and about the appropriate term, ACUM, as a different entity from adult adenomyosis; (ii) to review the literature beginning in 2010 and to present four new clinical cases attended by us within the first 7 months of 2011; (iii) to review the methods and criteria for the diagnosis of ACUM; and (iv) to provide information about this interesting pathology that is not as infrequent as previously believed, emphasizing the importance of gathering clinical evidence for making a correct diagnosis and the importance of determining the characteristics and localization of the ACUM for quickly choosing the best surgical treatment.

**Cases and Methods**

**Our new cases**

**Case 1. 305689**

A 36-year-old female immigrant from South-America with the antecedents of two deliveries (in 1994 and 1995) and a termination of pregnancy in August 2010 presented at our hospital with retained tissue, and a curettage was performed in September 2010. During the physical examination, we detected a uterine mass via transvaginal ultrasonography (TVU) that appeared as a cavitated tumor on the left anterior side of the uterus and was compatible with a degenerated leiomyoma or cystic adenomyoma.

The patient had been treated in other clinics since the beginning of 2010 for painful episodes in the left iliac fossae, which increased during menstruation despite her 15-year use of oral contraceptives (OC). After discovering a simple cyst in the patient’s right ovary and a degenerated leiomyoma in her left side, her physicians recommended discontinuing OC use and taking analgesics. While the patient was being studied, she became pregnant. Following the termination and curettage, she came to one of our outpatient clinics in November 2010. She complained of abdominal pain that was more intense in the left iliac fossae and increased during menstruation. She reported that the pain had become stronger since she had discontinued OC 8 months earlier. A new TVU examination revealed an intramural-subserosal mass of 3.5 × 5 cm in the left front side of the uterus that appeared to be cavitated with homogeneous contents, similar to an endometrioma and consistent with an ACUM.

**Case 2. 219598**

A 20-year-old nulligravida woman presented with an 8-month history of left iliac fossae pain and progressive dysmenorrhea that were not relieved by analgesic drugs.

The patient underwent menarche at 15 years of age and shortly thereafter began experiencing severe dysmenorrhea that was treated with non-steroidal anti-inflammatory drugs (NSAIDs) and OC. She repeatedly sought treatment at other hospitals for severe dysmenorrhea and pelvic cramping during and after menstruation. During the preceding months, the pelvic pain (in the left iliac fossae) and the recurrent dysmenorrhea had worsened and sometimes necessitated hospitalization for intravenous analgesia. A TVU examination revealed a nodule in the left front side of the patient’s uterus that suggested a rudimentary uterine horn occupied by a 35-mm hematometra. However, a magnetic resonance image (MRI) revealed the presence of an accessory endometrial cavity in the anterior left side of the uterus, as shown in Fig. 2. In addition, the radiologist identified an adjacent tubular structure that was interpreted to be an accessory and rudimentary Fallopian tube that ended in a cul-de-sac. The remainder of the uterus, Fallopian tubes and ovaries were anatomically normal. Thus, the patient was referred to our hospital for evaluation and to consider possible surgical treatment for a uterine malformation.

Our gynecologic examination was normal, but the TVU confirmed the presence of an accessory cavitated mass with the appearance of an endometrioma in the front of the left uterine horn. Hysterosalpingography (HSG) showed a normal uterine cavity with a slight concavity on the left side, which was probably related to the observed parauterine mass. Both the Fallopian tubes and the peritoneal spilling were normal. We performed a laparotomy with a tumorectomy in February 2011 by completely removing the ACUM located below the uterine insertion of the left round ligament.

The tumor exeresis was not technically easy because of the ambiguous limits of the mass and the presence of fibrotic tissue near the normal endometrium. We did not observe the accessory Fallopian tube that had been detected in the MRI, but we removed adjacent, undefined remnant tissue. We reconstructed the uterine wall, and the final result was a normal uterus and normal adnexae (Fig. 3). An accessory uterine mass of 40 × 30 mm with a cystic hemorrhagic cavity lined with atrophic endometrial mucosa associated with fibrosis was confirmed histologically. The adjacent remnant tissue morphologically resembled tubal fimbriae with vascular dilatation (Fig. 4).

In a posterior examination, the patient was recovered and has remained completely asymptomatic with normal ultrasounds.

The remainder of the uterine anatomy and ovaries was normal. The results of tumor marker assays and other blood analyses were also normal.
Case 3. 312903

An 18-year-old nulligravida woman with no previous sexual history presented with a history of left iliac fossae pain, hypogastric pain and progressive dysmenorrhea that were not relieved by analgesics dating from menarche at age 14.

The patient repeatedly sought evaluations at other hospitals because of severe dysmenorrhea and pelvic cramping. She had been examined at her reference hospital for a possible bicornuate uterus with hematometra. An MRI indicated the presence of a focal lesion of $2.4 \times 2.6$ cm located in the myometrial wall on the left side of the uterine body; this lesion was rounded, smooth and hyperintense on T1-weighted sequences. The use of contrast dye did not change the signal intensity, and the lesion was surrounded by a hypointense ring on T2-weighted sequences that did not enhance with i.v. contrast. These findings are characteristic of supposed cystic adenomyosis for the radiologist. The endometrial cavity appeared normal and showed no connection to the cystic mass. The rest of the myometrium was normal and free of adenomyosis, and the ovaries were normal (Fig. 5).

The patient was referred to our hospital for evaluation and for consideration of surgical treatment. Transrectal ultrasonography revealed a cavitated uterine mass of $23 \times 25$ mm in the anterior aspect of the left horn that was cavitated, contained echogenic material consistent with blood and resembled an ACUM. The uterus and adnexae were normal.

A laparotomy was performed (May, 2011) with a tumorectomy of an intramural uterine mass located under the left round ligament insertion. The mass was removed, and a section of the nodule spilled the retained chocolate-colored substance (Fig. 6). A histopathological examination of the specimen showed the endometrial lining of the ACUM. Two months after surgery, the patient is asymptomatic and has had a normal evolution.

Case 4. 313384

A 19-year-old nulligravida woman presented with a history of pelvic pain and progressive dysmenorrhea with increased pain following menstruation despite using NSAIDs and OC for 1.5 years. The pain was more intense in the left iliac fossae, and she had suffered from dysmenorrhea since menarche at 10 years of age.

The patient repeatedly sought evaluations at other hospitals for severe abdominal pain and colicky lumbar pain accompanied by frequent urination, suprapubic pain and some episodes of vomiting. She also presented at the emergency room of our hospital with severe dysmenorrhea despite the use of OC.

A physical examination indicated an anterior intramural fibroid uterus that was otherwise normal. A TVU showed that the uterus was basically normal, but a 2-cm myoma-like mass with a small cavitation was evident in the anterior surface of the left horn. The adnexae were normal. HSG showed a normal uterus and Fallopian tubes. Laparotomy was indicated, and a tumorectomy was performed.
Under the left tube and round ligament, there was a small intramural mass that was removed. Below and behind, there was another mass that was also removed, extracting a fragmented 2-cm tumor from the left uterine side. The histopathological examinations of both tumors revealed 2 ACUMs with functional endometria and adenomyosis within the adjacent myometrium (Fig. 8). The post-operative course was normal. The patient is currently asymptomatic, and a recent physical examination was also normal.

Review of the literature

We reviewed the literature beginning in 2010 by collecting and analyzing all the published cases of JCA, uterus-like masses and uterine accessory masses that had not been included in our previous publication (Acién et al., 2010). Table I lists these cases and publications beginning with a tabulation of the cases included in our previous report and ending with the cases included in this study.

We have also analyzed the published case by Persson et al. (2010) reporting a robot-assisted laparoscopic surgery for a rudimentary uterine horn with two non-communicating cavities. Considering the communicated facts and images of MRI and TVU that we know, we think that the cavity no.3 could actually be an ACUM.

Diagnostic methods

The primary fact for diagnosis is clinical suspicion in young women who present with severe dysmenorrhea that has been progressive since menarche and pelvic pain that persists during the pre- and post-menstrual periods and does not respond to common analgesics. It is necessary for such patients to repeatedly seek emergency services. Occasionally, the discomfort associated with adolescent dysmenorrhea becomes more intense and unbearable during adulthood, sometimes after childbirth; however, severe dysmenorrhea generally occurs before age 30.

A bimanual clinical examination usually shows a nodule on the right or left uterine horn that is suggestive of a fibroid and that is painful.

TVU (or transrectal in young women with no sexual history) is a decisive diagnostic method for ACUMs because it can detect a nodule, such as a myoma, in the anterior right or left wall of the uterine horn. In the center of this presumed fibroid, a small cavitation with content resembling an endometrioma can be seen. The remainder of the uterine endometrium and myometrium is normal with no other associated uterine anomalies. The ovaries also tend to be normal (Fig. 9). A diagnosis can be made based on these findings, followed by the decision to perform laparoscopic surgery or a laparotomy.

However, to rule out the possibility of a non-communicated cavi
tated rudimentary uterine horn, it is desirable to perform HSG to verify the presence of a normal uterus and patent Fallopian tubes (Fig. 10). It is also highly valuable to ensure diagnostic accuracy with an MRI that shows an accessory endometrial cavitation in the myome-
trium of the right or left wall of the uterus. The cavitation does not communicate with the normal uterine cavity and is observed to be
as superintense on T1 as a hematometra, which suggests the presence of an endometrioma-like mass in the anterior and right or left side of the uterine body (see Figs 2 and 5).

Results

As shown in Table I, there have been four papers on cases diagnosed as JCA within the last year, excluding ours, making a total of 15 such cases, which we suggest were actually ACUMs. The greatest number of cases \( n = 9 \) were communicated by Takeuchi et al. (2010), although they limited the inclusion criteria to women under 30 years of age. Kriplani et al. (2011) reported on four cases, and the other two were from Akar et al. (2010) and Chun et al. (2011). The ages of these patients were between 15 and 30 years (mean: 22.3 years). All of these patients’ nodules were located on the left antero-lateral surface of the uterus (3 cases), the right anterior surface of the uterus (10 cases), the anterior myometrium (1 case) and the left posterior aspect of the uterus (1 case).

There have also been three other publications (Tijani et al., 2010; Seki et al., 2011; Shin et al., 2011) that each communicated a case involving a uterus-like mass, but none of these appeared to be an ACUM. In the case of a 44-year-old woman from Seki et al. (2011), we believe that the uterus-like mass observed was a rudimentary left uterine horn that was cavitated, non-communicated and included in the left inguinal mass; therefore, the patient’s uterus, which was not adequately examined in the study, must be unicorneuate.

However, as previously mentioned, considering the facts and images from the Persson et al. (2010) case report, probably the cavity no.3 is actually an ACUM associated with a cavitated and non-communicated uterine horn. In this case, this would be the first reported case of an ACUM associated with a malformed uterus.

The four cases on which we report in this article underwent surgery between January and July 2011. The ages of the patients were 36, 20, 18 and 19 years and, interestingly, all the ACUMs were located on the left anterior side of the uterus. Of particular interest, there was a single nodule in three cases and a double nodule in one case (the first communicated case). Both nodules of the double nodule were attached back-to-back and located more posterior than usual under the left tube and round ligament. Another case also had a rudimentary fallopian tube adjacent to the ACUM.

In our cases, as in all of the others reported in the literature, the main symptoms were severe dysmenorrhea and pelvic pain that increased during menses in young women, even with OC use, and responded poorly to common analgesics. These patients were treated at the emergency departments on multiple occasions. Clinical suspicion and transrectal or TVU clearly pointed to a diagnosis, and MRI showed a cavitation of the antero-lateral uterine myometrium that was superintense on T1 and clearly independent from the...
**Figure 4** Case 2. Histological images of the little specimen adjacent to the accessory uterine mass (ACUM) extracted during tumor exeresis. 
(A) Rudimentary fallopian tube adjacent to the ACUM; H-E, x2. (B) Tubal fimbriae; H-E, x4. (C) Tubal epithelium and stroma; H-E, x10. (D) Tubal epithelium and stroma; H-E, x20.

**Figure 5** Case 3. Pelvic MRI showing an accessory endometrial cavity into the left myometrial wall, hyperintense in T1, suggesting hematometra. The lesion is rounded by a hypointense ring in potenciated sequences of T2 suggesting deposits of hemosiderine. (A) T1, transversal cut. (B) T1, coronal cut. (C) T1, sagital cut. (D and E) T1, sagital cuts with contrast. (F and G) T2, transversal cuts. (H) T2, coronal cuts. (I) T2, central sagital cut. (J) T2, left lateral sagital cut. (K and L) T2, transversal cuts.
normal endometrial cavity on T2. These findings, together with a normal HSG, confirmed a diagnosis of an ACUM.

The best therapeutic approach is tumorectomy, which can be performed laparoscopically; however, we prefer laparotomy because it is difficult to completely enucleate the tumor, especially when it is located inside the myometrium at the lateral uterine parametrium/broad ligament.

Figure 6 Case 3. (A) Extracted accessory and cavitated uterine mass. (B and C) Section of the uterine mass spilling chocolate fluid.

The tumor is commonly single and isolated, and it is always located at the anterior left or right side of the uterus below the insertion of the round ligament. In one of the cases from Kriplani et al. (2011), it is mentioned that the mass was located in the anterior myometrium. In addition, the case from Chun et al. (2011) is confusing because the authors indicate the left posterior aspect of the uterus, but according to the laparoscopic images in their article, the location seems to be

Figure 7 Case 4. (A) Internal genitalia of the patient showing the tumor on the left side of the uterus under the tube and round ligament. There were two ACUMs in the same location. (B) Exeresis of the accessory mass number 1. (C) Exeresis of the accessory mass number 2. (D–E) Both ACUMs after extraction.
left uterine parametrium/broad ligament. Moreover, the laparoscopic images from the tumoral excisions by Akar et al. (2010), Kriplani et al. (2011) and Takeuchi et al. (2010) do not appear to direct the reader to the correct locations of the masses.

Discussion

The 4 new ACUM cases reported in this article and the 15 cases of JCA identified in the review of the literature beginning in 2010 add up to more cases than all those previously published and collected in our previous publication (Acién et al., 2010). Therefore, it appears that this pathology is not as rare as previously believed. If ACUMs were more readily diagnosed, this condition might be more frequently identified and properly treated, which would prevent the constant suffering of these young women. However, the implementation of the proper diagnosis, treatment and surgical approach requires that the physician clearly understands the nature of this pathology and the systematic localization of the accessory and cavitates masses on the anterior left or right side of the uterus, below the insertion of the round ligament on the uterine horn.

Although Oliver described a case in 1912 with an ‘accessory uterus distended with menstrual fluid’ in a 34-year-old woman, Tamura et al. (1996) first described a 16-year-old girl with a ‘juvenile adenomyotic cyst of the corpus uteri’ that manifested as a cystic lesion measuring 15 mm in diameter, located on the left side of the anterior wall of the uterus and independent of the normal uterine lumen. Since then, the cases collected in our previous publication (Acién et al., 2010) and in Table 1 of this study have been reported. Most of these cases have been labeled as ‘isolated cystic adenomyomas’ or JCA, and most have been published by Japanese authors. Takeuchi et al. (2010) noted that such cases are often presented at Japanese academic meetings, and the pathologic features of this type of cystic lesion have become widely recognized in Japan as characteristic of JCA, but this pathology has also been reported in other countries.

Takeuchi et al. have reported nine cases, with a particular emphasis on the relevance of the laparoscopic enucleation. Furthermore, these authors have noted that the pathogenesis of the disease remains unclear; however, after analyzing the terms of adenomyosis and adenomyoma, they have proposed that JCA is a ‘cystic adenomyosis’ or a ‘variation of cystic adenomyosis’ rather than a congenital abnormality. In addition, we have previously noted that the cases published as non-communicating accessory uterine cavities and isolated cystic adenomyomas, as well as some cases of uterus-like masses, correspond to the same pathology: an ACUM. This ACUM could be caused by ectopia or by the duplication and persistence of the ductal Mullerian tissue (displaced tissue) in a critical area at the attachment level of the round ligament and might be related to a gubernacular dysfunction (Acién et al., 2011). It was noted in the Results section that although a few cases point to another location for this pathology, the images in the publications appear to refer to the same location as that implicated in the majority of cases.

Our previously reported cases (Acién et al., 2010) also demonstrated that ACUMs are clearly different from true cavitates adenomyomas and cystic adenomyosis in terms of clinical presentation, ultrasonography, surgical findings and histopathology. Adenomyomas and cystic adenomyosis are more characteristic of older women, who develop adenomyosis spread anywhere inside the uterine corpus, and the cysts frequently exhibit the absence of an internal
Table I  Published cases of accessory uterine masses, JCAs and uterus-like masses beginning in 2010.

<table>
<thead>
<tr>
<th>Reference</th>
<th>No. of cases and period</th>
<th>Age (year)</th>
<th>Parity</th>
<th>Presenting symptoms</th>
<th>Age at onset of symptoms (year)</th>
<th>Lesion size (cm)</th>
<th>Lesion location</th>
<th>Previous medical treatment</th>
<th>Diagnosis from authors</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acién et al. (2010)</td>
<td>4, 2001–2009</td>
<td>15, 21, 32, 33</td>
<td>0, 0, 4, 0</td>
<td>Severe dysmenorrhea and pelvic pain</td>
<td>14, 18, 30, 32</td>
<td>2.25, 5, 3.5</td>
<td>Anterior wall of the uterus. Right side in 3. Left side in 1 case</td>
<td>NSAIDs Accessory and cavitated uterine mass</td>
<td>JCAs</td>
<td>This paper included other 14 cases from literature considered ACUMs</td>
</tr>
<tr>
<td>Takeuchi et al. (2010)</td>
<td>9, 2000–2007</td>
<td>Only ≤30 years, 20–30 (mean 25.2)</td>
<td>0, 0, 2, 0, 2, 0, 0, 0</td>
<td>Severe dysmenorrhea and pelvic pain</td>
<td>1–13 years (mean = 6.6) from menarche</td>
<td>2.5–4.2</td>
<td>Lateral wall near the uterine round ligament attachment site. Right side in 6. Left side in 3</td>
<td>OC in 3. GnRH analogs in 3</td>
<td>JCA</td>
<td>All nine cases are ACUMs. Diagnostic criteria included age ≤30 years and cystic lesion &gt;1 cm</td>
</tr>
<tr>
<td>Akar et al. (2010)</td>
<td>1, –</td>
<td>15 years</td>
<td>0</td>
<td>Severe dysmenorrhea and intermittent episodic pain</td>
<td>12 years</td>
<td>4 cm</td>
<td>Right anterior fundal portion of the uterus</td>
<td>Aromatase inhibitors, progestins, OC, GnRH analogs</td>
<td>JCA</td>
<td>We think this tumor was an ACUM with quickly and early growth</td>
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</tr>
<tr>
<td>Tijani et al. (2010)</td>
<td>1, –</td>
<td>35</td>
<td>0</td>
<td>Intermittent bouts of intense pain in the left iliac fossae and pelvis</td>
<td>34</td>
<td>21 cm</td>
<td>Posterior fundus of the uterus and posterior face of the bladder, attached to these organs</td>
<td>OC and NSAIDs in 3</td>
<td>JCAs</td>
<td>We think that three cases are ACUMs, but Case 3 is doubtful</td>
</tr>
<tr>
<td>Kriplani et al. (2011)</td>
<td>4, 2008–2009</td>
<td>16, 18, 16, 24</td>
<td>0, 0, 0, 0</td>
<td>Severe dysmenorrhea</td>
<td>14.5, 17, 15, 15</td>
<td>3.5, 4.2, 2.8, 2.9</td>
<td>Right uterine wall near fundus in 3. Anterior myometrium in the case 3</td>
<td>OC and NSAIDs in 3</td>
<td>JCAs</td>
<td>We think that three cases are ACUMs, but Case 3 is doubtful</td>
</tr>
<tr>
<td>Chun et al. (2011)</td>
<td>1, –</td>
<td>19</td>
<td>0</td>
<td>Severe dysmenorrhea and pelvic cramping</td>
<td>Menarche, 12 years</td>
<td>3</td>
<td>Nodular lesion on the left posterior aspect of the uterus</td>
<td>OC, NSAIDs</td>
<td>JCA</td>
<td>Figures from MRI and laparoscopy show that tumor is located on the left side of the uterus under tube and round ligament</td>
</tr>
<tr>
<td>Seki et al. (2011)</td>
<td>1, –</td>
<td>44</td>
<td>0</td>
<td>Mass in a left inguinal surgical scar, larger and more painful during menstruation</td>
<td>Operated of a sliding hernia of the left ovary at 10 m, and endometriosis in ISS at 26 years</td>
<td>3.8</td>
<td>In the left inguinal subserosal tissue</td>
<td>Subserosal uterus-like mass</td>
<td>Subserosal uterus-like mass</td>
<td>We think it was a cavitated non-communicating rudimentary left uterine horn. Present uterus would be unicornuate</td>
</tr>
<tr>
<td>Shin et al. (2011)</td>
<td>1, –</td>
<td>31</td>
<td>0</td>
<td>Low abdominal pain</td>
<td>30 years</td>
<td>3</td>
<td>In the sigmoid mesocolon</td>
<td>Uterus-like mass</td>
<td>Subserosal uterus-like mass</td>
<td>We think it was a cavitated non-communicating rudimentary left uterine horn. Present uterus would be unicornuate</td>
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<tr>
<td>Present study</td>
<td>4, January–July 2011</td>
<td>36, 20, 18, 19</td>
<td>2, 0, 0, 0</td>
<td>Severe dysmenorrhea and recurrent pelvic pain</td>
<td>30, 15, 14, 14</td>
<td>3.5, 3.5, 2, (1 + 1.5)</td>
<td>Anterior wall of the uterus. Left side in the four cases</td>
<td>OC, NSAIDs Accessory and cavitated uterine masses. One of them also had a rudimentary fallopian tube</td>
<td>Accessory and cavitated uterine masses</td>
<td>The last case had two nodules, both inside broad ligament under tube and RL</td>
</tr>
</tbody>
</table>

OC, oral contraceptives; NSAIDs, non-steroidal anti-inflammatory drugs; ISS, inguinal surgical scar; RL, round ligament.
epithelial lining, as indicated by histopathology. However, the myometrium adjacent to the ACUM may develop adenomyosis that is not present in the rest of the uterus and is probably related to the increased intracystic pressure present during menstruation. The ACUMs seem to be bigger when the woman is older.

In a letter to the editor of Obstetrics and Gynecology in response to our publication on ACUMs (Acién et al., 2010), Batt and Yeh (2011) suggested that the functioning accessory uterine masses described by Oliver (1912), Ahmed et al. (1997) and Liang et al. (2010) are examples of müllerianosis, developmentally misplaced Müllerian tissue or Müllerian choristomas, which have a different pathogenesis from other cases of ACUMs. However, in our reply, we asked the question, ‘Where does this ectopic tissue come from?’ and we suggested that it was displaced tissue caused by the dysfunction of the female gubernaculum (Acién et al., 2011). In subsequent publications, we have insisted on this idea and have included ACUMs in Group 4 of the updated embryological-clinical classification for female genito-urinary malformations that was recently published in Human Reproduction Update (Acién and Acién, 2011).

Dietrich (2010) recently published an update on adenomyosis in adolescents, and the cases included in a table were mostly those previously referred to as ‘adenomyotic cysts’. Some of the cases, however, were actually uterine malformations with a cavitated rudimentary horn (a ‘unicornuate uterus with an occult adenomyotic rudimentary horn’, Frontino et al., 2009), which we believe is the first pathology to have a differential diagnosis of ACUMs.

Regarding diagnostic methods for ACUM, as we have previously mentioned, the key-point is clinical suspicion based on the appearance of symptoms, the recurrence of symptoms, patient age, knowledge of this pathology, and the search for a mass using transrectal or TVU and the clinical data on the common location of ACUMs. Naturally, HSG excludes occlusive uterine malformations, and MRI results are highly valuable in ensuring diagnostic accuracy, if interpreted properly (both the T1 and T2 images provide abundant information).

The current criteria for considering this pathology to be an ACUM are the same as those mentioned in our previous publication (Acién et al., 2010) and in the Background section of this report, but we have found that the first criterion, ‘an isolated accessory cavitated mass’, may not be accurate because some cases have two ACUMs in the same unilateral area of the uterus. Neither would the second criterion (normal uterus) be indispensable if it was correct that the Persson et al. (2010) case had an ACUM associated with a bicornuate uterus with segmentary defects and cavitated rudimentary horn. Otherwise, we continue to employ the same criteria, particularly the absence of adenomyosis or adenomyotic cysts in the remainder of the uterus that is free of ACUM, the localization under the round ligament, the endometrial lining and chocolate content and the
habitual presence of a normal lumen. Chun et al. (2011) have suggested the following new diagnostic criteria for JCA: (i) the age of onset of severe dysmenorrhea is within 5 years after menarche or ≤18 years of age; (ii) no history of suspected endometrial or uterine injuries (delivery, myomectomy or dilatation and curettage); and (iii) the presence of a cystic lesion >5 mm indicated by imaging studies or observed during surgery. These criteria are appropriate, but we do not consider them to be necessary to categorize a JCA as an ACUM. Furthermore, ACUM patients are largely young and nulliparous, but some patients have been more than 30 years of age. In our material, these are frequently immigrant women, with less medical care and who carried the pain for several years.

Regarding therapeutic management, most recent publications have included laparoscopic excision of the tumor (Takeuchi et al., 2010; Kriplani et al., 2011) or robot-assisted laparoscopy (Akar et al., 2010), always considering that tumor to be a JCA. We agree with performing laparoscopic resection, but the systematic anterior and lateral localization of this pathology should be considered when choosing the appropriate site for the uterine incision and the excision of the lesion, particularly when contemplating the absence of net limits of the tumor and the depth of the penetration of the fibrotic and adenomyotic tissue into the myometrium. In the reported images within the mentioned publications (Akar et al., 2010; Takeuchi et al., 2010; Chun et al., 2011; Kriplani et al., 2011), the incision and excision of the tumor were not always performed in the most appropriate way. In the new cases reported here, we performed laparotomy for a better documentation of ACUMs instead, but laparoscopic excisions can be performed with proper knowledge and care.

Conclusions

ACUM is a pathology that is less rare than previously believed, making the consideration of an ACUM diagnosis essential in clinically suspicious cases. TVU and MRI facilitate a correct diagnosis. Early surgical treatment that includes the adequate removal of the mass via laparoscopy or laparotomy can prevent the typical relentless suffering of these young women. The pathogenesis of this entity is controversial; however, in our opinion, ACUM is a new type of Mullerian anomaly that is probably related to a dysfunction of the female gubernaculum. Occasionally, more than one ACUM with functional endometria can be found below the uterine round ligament insertion, although some ACUMs are located in a more lateral site under the Fallopian tube and the round ligament. Other cases can have a tubal rudiment adjacent to the ACUM and, although not confirmed, it could also be that some cases can be associated with uterine malformation. For the treatment of ACUM, we recommend that the mass is always removed through the anterior surface of the uterus to achieve the best results and to optimize safety.

Details of Ethics Approval

Comité Etico de Investigación Clínica (CEIC), San Juan University Hospital, but not required for case studies in Spain. Written informed consent was obtained from each patient for publication of case and accompanying images. A copy of the written consents are available for review by the Editor-in-Chief of this Journal.

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Authors’ roles

P.A. studied and operated the reported patients, designed the study, reviewed the literature, made the table and figures and wrote the paper. A.B., F.F., M.A. and J.M.R. reviewed the reported cases, participated in their study and surgical operations and reviewed the manuscript. M.J.M. performed the histopathological studies. P.A. had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

Conflict of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of this paper. The authors declare that they have no competing interests.

References


