Accessory and cavitated uterine mass versus juvenile cystic adenomyoma

We have read with interest the writing of Martin and Koninckx (1) as well as the publication of Arya and Burks (2) in *F&S Reports*. In another letter to the editor of *Obstetrics and Gynecology* in response to our publication on the accessory and cavitated uterine mass (ACUM) (3), Batt and Yeh (4) suggested that functional accessory uterine masses are examples of Müllerianosis, that is, of poorly located (dislocated) Müllerian tissue in the development or Müllerian choristomas. We suggested that it was Müllerian tissue displaced by ectopia or by the duplication and persistence of ductal Müllerian tissue in a critical area at the attachment level of the round ligament, possibly caused by dysfunction of the female gubernaculum and naturally congenital. In any case, ACUM would be a congenital Müllerian choristoma, and we included ACUMs in the embryological–clinical classification of female genitourinary malformations (5–9).

Subsequently, we do not believe that the term juvenile cystic adenomyoma (JCA) is correctly used, and we believe that most of the cases that are published under this terminology are in fact ACUMs, which also require exeresis, and that the disquisition of the presence of adenomyosis around it or not is secondary, because we believe that this is because of the intracystic menstrual pressure. Cavitated adenomyomas and cystic adenomyosis are something different, another pathology (effectively Müllerian) with a different pathophysiology. They have no relationship, either histologic or in location, with ACUM normally located under the insertion of the round ligament in the uterus, which, on the other hand, usually has a normal endometrial cavity that is communicated with the fallopian tubes. However, several ACUMs have been, and continue to be, published as JCA (which is not adenomyosis), although we agree with Martin and Koninckx (1) that the clinical response does not depend on theory or pathogenesis because in any case, it is necessary to eliminate these symptomatic ACUMs or JCAs.

Unfortunately, when one reviews the article by Arya and Burks (2) on JCA, and the other articles mentioned by these investigators, after the literature review, one is surprised that conceptual, diagnostic, and managing doubts persist. Arya and Burks (2) present two cases that we believe are typical of ACUMs. The first case is in "the left lateral uterine wall extending toward the left broad ligament," and the second case is "in right cornual region" but that also had another supposed ACUM "in the left cornual region, concerning to be a remnant of JCA" that later continued to cause symptoms similar to the previous ones. Arya and Burks (2) say that they have reviewed the literature, but of course, they have not reviewed ACUM cases in previous publications (3, 10). However, Arya and Burks (2) in their review reference other recent works. The first is by Kiyah et al. (11) for the "Decidualized Juvenile Cystic Adenomyoma Mimicking a Cornual Pregnancy" associated with ectopic pregnancy in the contralateral tube. Indeed, it could be a JCA, according to us an ACUM, but the patient had a previous cesarean section so it could also be iatrogenic endometriosis resulting from the previous hysterorrhaphy or developed in the diverticulum of the cesarean scar. The decidualized endometrium is due to ectopic pregnancy in the left tube, and the peritumoral adenomyosis is due to intracystic menstrual pressure. The second is by Protopapas et al. (12) who entitled their article "Juvenile Cystic Adenomyoma vs Blind Uterine Horn: Challenges in the Diagnosis and Surgical Management," but after reviewing it, we believe that the case is not well studied. From the images, it appears to be a bicornuate uterus with a rudimentary cavitated and not communicated left uterine horn, but the "histology was suggestive of a JCA." However, we did not know if the entire left uterine horn was excised ("A left neocornu was constructed") because in such a case, the most relevant data for a correct diagnosis would have been the presence of one or two cavities lined by the endometrium in the surgical specimen, and mainly, where did the excised left tube come from? The third is by Wilcox et al. (13) who published in *Obstetrics and Gynecology* two typical cases of ACUM as "Juvenile Cystic Adenomyoma" without mentioning the previous publications on ACUM also in *Obstetrics and Gynecology* (3).

In conclusion, ACUM is a less rare pathology than previously believed, which has frequently been published as "uterus-like mass" or as JCA, and its consideration is essential in clinically suspicious cases for appropriate surgical treatment. Transvaginal ultrasound and magnetic resonance imaging (and if necessary, hysterosalpingography showing a normal uterine cavity) facilitate a correct diagnosis. Furthermore, the diagnostic criteria suggested by Chun et al. (14) for the JCA are also equally applicable to ACUM. The differential diagnosis includes the rudimentary and cavitated uterine horns, such as those found in other uterine malformations (unicornuate and bicornuate uterus and Müllerian segmentary atresias), adenomyosis with cystic or degenerated areas, and degenerated leiomyomas. Early surgical treatment that includes proper removal of the mass by laparoscopy or laparotomy can prevent the typical recurrent cyclical distress of these young women. The pathogenesis of this entity is controversial; however, in our opinion, ACUM is a Müllerian anomaly that is probably related to a dysfunction of the female gubernaculum. Occasionally, more than one ACUM with functional endometrium can be found below the insertion of the uterine round ligament (or also on the contralateral side, as suggested by case two of Arya and Burks (2), and then, the symptoms recur if left in situ). Other cases may have a tubal rudiment adjacent to ACUM (which would speak in favor of detached Müllerian choristoma due to female gubernaculum traction). Moreover, although it is not confirmed, it could be that some cases are associated with a uterine malformation (a bicornuate uterus, but the rudimentary cavitated and noncommunicated uterine horn must be ruled out). Likewise, there may be adenomyosis in the myometrial wall surrounding the ACUM, probably related to an increase in menstrual intracystic pressure. Finally, for ACUM treatment, we recommend that the mass is always removed through

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the face or anterior wall of the uterus to achieve the best results and optimize safety.

Pedro Acién, M.D., Ph.D.[a,*]
Maribel Acién, M.D., Ph.D.[a,b]
[a] Department/Division of Gynecology, Miguel Hernández University; and [b] Obstetrics and Gynecology, San Juan University Hospital, San Juan Campus, San Juan, Alicante, Spain
*Reprint requests: Pedro Acién, M.D., Ph.D., School of Medicine, Miguel Hernández University, San Juan Campus, 03550 San Juan, Alicante, Spain.
E-mail address: acien@umh.es

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