# Juvenile cystic adenomyomas: acquired adenomyosis variant or congenital Müllerian defects?

Arya and Burks (1) continue the long debate about acquired and congenital forms of Müllerian diseases. They contrast the studies by Takeuchi et al. (2) and Acién et al. (3), summarize the differences, and agree with Takeuchi et al. (2) who concluded that juvenile cystic adenomyomas (JCA) are a rare variant of adenomyosis rather than a congenital abnormality. This is contrasted with Acién et al. (3) who concluded that accessory and cavitated uterine masses (ACUM) with a functional endometrium and JCA were the same pathology. Each has the appearance of a miniature partial uterus, and both are found in young women with severe dysmenorrhea and pelvic pain before the age of 30 years. The mean age in the study by Acién et al. (3) excluding Takeuchi et al. (2) was 20 years (range, 15–44 years), and that of Takeuchi et al. (2) was 25 years (range, 20–30 years). Batt and Yeh (4) would have agreed with Acién et al. (3) and added that these are organoid choristomas, masses of histologically normal tissue that are not normally found in the organ or structure in which they are located. The organoid choristoma-like appearance of an accessory uterus is demonstrated in the coronal plane images in the study by Acién et al. (3). Other suggested distinguishing criteria include the presence of a denser area of adenomyosis surrounding the cystic area lined with the endometrium in JCA but not in ACUM and a junctional zone in ACUM but not in JCA. Takeuchi et al. (2) also differentiated adenomyosis from JCA when the basal layer of the endometrium-like tissue covering the cystic lumen was absent. Similar uterine organoid-like appearances have also been seen in ovarian endomyometrioma, uterine-like masses, and deep, retroperitoneal, retrocervical endometriosis.

A junctional zone can be important in surgery where it facilitates the removal of the mass like it does when removing myomata. Without a junctional zone, the removal of only the cyst wall while sparing as much of the myometrium as possible is used similar to a partial removal of adenomyosis.

The studies by Arya and Burks (1), Takeuchi et al. (2), and Acién et al. (3), and Cullen (5) are useful in comparing the age distribution. In his 1908 book, Cullen (5) illustrated cystic adenomyomas in six patients and miniature cavities in 15 patients. The mean age with cystic adenomyomas was 40 years (range, 25–53 years), and with miniature cavities was 43 years (range, 23–59 years). Although there is an overlap, the later ages in Cullen (5) suggest an acquired adult-onset form or observation bias, while Arya and Burks (1), Takeuchi et al. (2), and Acién et al. (3) discuss congenital or early-onset anomalies. Based on the age distribution, it may be concluded that both ACUM and JCA fit Batt and Yeh's (4) criteria as representing congenital forms of adenomyosis. Congenital adenomyosis is one of the four congenital versions of Müllerian disease in Batt and Yeh's (4) theory of Müllerianosis, the presence of congenital adenomyosis, endometriosis, endocervicosis, and endosalpingiosis. Those four congenital diseases complement the four acquired diseases of the same name.

Batt and Yeh (4) reviewed almost 150 years of congenital anomalies including discoveries of the endometrium in the uterine wall of a 9-month-old fetus and ectopic endocervical-like tissue in newborns at autopsy. They included the more recent observations of primitive nests of adenomyosis within the myometrium of a fetus at autopsy, and organoid structures resembling "primitive endometrium" outside the uterus. Immunohistochemical staining for CD10, cytokeratin, CA 125, and estrogen receptors was used to confirm identification. Batt and Yeh (4) focus on the Müllerian origin and localization and not the methods of dissemination, activation, transition, or inactivation. Their concepts of acquired Müllerian diseases parallel Sampson's decision to use the terms endometriosis and endosalpingiosis rather than Müllerianosis because they considered most forms of endometriosis and adenomyosis to be acquired. Batt and Yeh (4) would likely consider that some, if not all, publications on JCA and ACUM are on variations of congenital adenomyosis.

Although these theories may help orient research projects, education, and discussing why treatment may work, treatments should be judged by their efficacy. Evidence, not theory, is needed for treatment decisions. Moreover, theory does not clarify the risks, benefits, cost, availability, insurance coverage, preauthorization, in-network providers, or any of the major concerns of most patients. Whatever the theory or pathogenesis of ACUM and JCA is, the clinician will remove them when symptomatic.

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