Mini-Review

Uterine Cystic Adenomyosis: A Disease of Younger Women

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A B S T R A C T

Purpose of Review: We adopted a life-cycle approach to further our understanding of the natural history of the cystic forms of uterine adenomyosis first described by Cullen in 1908.
Search Strategy: Scopus and PubMed were searched for all terms referring to cystic variant of adenomyosis or adenomyoma. References found in major publications were also included in the review.
Main Findings: With the introduction of non-invasive imaging techniques, a number of cases of cystic variants of adenomyosis have been reported. Progressive, severe, medication-resistant dysmenorrhea is the main clinical feature but delay in diagnosis remains problematic. The life-cycle approach demonstrates that cystic adenomyosis is more relevant to adolescent and young adulthood. Congenital müllerian and wolffian cysts and the uterus-like masses are more frequent in women >30 years of age. The latter is frequently located outside the uterus and may represent a form of endometriosis rather than adenomyosis. Differential diagnosis includes ovarian cysts and congenital uterine anomalies. Menstruation suppression with continuous oral contraceptive pill with ultrasonographic monitoring of cyst regression may prove successful in the young woman, but surgical excision using minimally invasive endoscopy is highly effective. The various terms used in literature to describe these lesions cause considerable confusion. Here we propose a classification into 3 subtypes and standardized reporting criteria to enable comparison.
Conclusions: Endometrial lined myometrial cysts are almost specific to adolescent and young women. We propose a new classification system.
Key Words: Cystic adenomyosis, Myometrial cyst, Dysmenorrhea, Adolescent

Introduction

Adenomyosis involves the presence of nests of ectopic non-neoplastic, endometrial glands, and stroma within the myometrium surrounded by a hypertrophic and hyperplastic myometrium.[1] The use of modern imaging techniques enables its identification as diffuse or focal thickening of the myometrial junctional zone.[2,3] Less common forms of adenomyosis such as adenomyoma, adenomyomatous polyps, and cystic adenomyoma have also been described. Although adenomyosis seems very rare before age 20, a cystic form has mainly been reported in young women.[4] This cystic variant can be identified by magnetic resonance imaging (MRI), since its fluid content exhibits high signal intensity on T1-weighted images and the surrounding myometrial wall exhibiting distinct low signal intensity on T2-weighted images.[5]

Kishi et al[6] described 4 subtypes of adenomyosis based on lesion location and their extension in the uterus as seen by MRI: subtype (1) is confined to the inner uterine wall; subtype (2) is confined to the outer uterine wall; subtype (3) includes solitary lesions; and subtype (4) includes all other lesions. But the applicability of this classification to cystic forms is unclear.

Submucous adenomyomas with cyst formation were first described by Cullen in 1908[7] in 5 hysterectomy specimens from women aged 34, 35, 36, 39, and 50 years. The cysts (1 case had 3 cysts) measured around 10 mm. The cavities were lined by normal endometrial mucosa and were filled by chocolate-colored contents. Cullen demonstrated that in advanced cases submucous adenomyoma with cyst formation can be part of a more complex structure with subperitoneal and submucous extension. He speculated that in contrast to subperitoneal adenomyomas, submucous adenomyomas do not show much cystic dilatation because they are continuously subjected to uterine pressure from all sides.

Cystic adenomyosis is certainly not very common, but the use of modern imaging techniques enabled the identification of different types of adenomyotic cysts; some of these were in the younger age groups. The juvenile cystic variant of adenomyosis or adenomyoma is of particular interest as it may have implications for the pathophysiology of the disease. We undertook a literature review and analyzed reported cases using a life cycle approach.

Strategy of Search

We used Scopus and PubMed search for the terms: circumscribed, cavitated, or cystic adenomyoma or adenomyosis, juvenile adenomyotic cyst, adenomyotic cyst,

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juvenile cystic adenomyoma, intramyometrial or congenital uterine cyst, accessory cavitated uterine mass, accessory uterine cavity and uterus or uterine-like mass. In addition, references found in identified publications were included in the review. Knowledge of these rare lesions is largely based on individual case reports and a few small series.

Review of the Literature

It is to be noted that the various terms used to describe uterine cysts cause confusion. Strict histopathologic or imaging criteria are needed to distinguish uterine cystic adenomyosis from other congenital or acquired uterine cysts. In line with the initial description by Cullen,[7] criteria for the diagnosis of an adenomyotic cyst include a cavity filled with hemorrhagic fluid that has no communication with the uterine cavity and that is lined by endometrium and surrounded by myometrium. Progress in imaging techniques such as transvaginal (TVU) or transabdominal (TAU) ultrasonography (US) and especially MRI allows accurate evaluation of the content of the cyst and of the surrounding endometrial and myometrial tissue, as well as in the myometrial junctional zone.

Although the first mention of a form of adenomyosis called "adenomyomatous polyp" is contained in an article published in 1860 by Carl Rokitansky,[8] Cullen[7] was the first to carefully describe the different appearances of adenomyomas including the solid and cystic variants. Cullen observed that cystic structures are frequently present and vary in diameter from a few millimeters to several centimeters. Some of them represented dilated glands or a part of a polycystic adenomyotic structure; others were isolated cysts within the uterine wall. Cullen described a case where a subserosal adenomyoma was linked to the uterine cavity by a duct a few millimeters wide. Similar subserous cysts connected to the uterine cavity have been described by Keating[9] and Dobashi.[10] Buerger and Petzing[11] pointed to the difference between epithelium-lined intramural cysts of the uterus, which they thought were of congenital origin (müllerian or wolffian duct cysts) and acquired cysts, such as cystic degeneration of myomas, cystic adenomyomas and cervical retention cysts. During the second half of the 20th century several cases of congenital cysts of the corpus uteri were described in mature women undergoing hysterectomy.[12,13] Distinguishing Müllerian forms from wolffian cysts remains difficult and mixed forms may exist.

Imaging technology facilitated the detection of "adenomyotic" cysts in young women. Tamura et al[14] described an adenomyotic cyst of the corpus uteri (coined name: juvenile cystic adenomyoma, JCA) in a 16-year-old girl who presented with severe dysmenorrhea that started 3 years after menarche. The cyst measured 15 mm in diameter and did not communicate with the uterine cavity. There were no other foci of adenomyosis in the uterus. Troiano et al reported a well-circumscribed cyst within the myometrium. The hemorrhagic contents were at different stages of organization.[15] According to Takeuchi et al,[16] a total of 30 young patients with "cystic adenomyoma" had been reported in Japanese literature and were included in their review. Takeuchi et al reported 9 new cases of JCA based on the following criteria: (1) age ≤30 years; (2) presence of a cystic lesion of ≥10 mm in diameter that did not communicate with the uterine lumen and that was surrounded by hypertrophic myometrium on diagnostic imaging; (3) an association with severe dysmenorrhea. Cucinella et al[17] argued against the use of the term 'adenomyoma' in connection with these lesions, stating that they are more correctly described as adenomyotic cysts or, depending on age at onset—as suggested by Takeda et al[18]—as "juvenile adenomyotic cyst" or "juvenile cystic adenomyoma" (JCA). Acién et al[19] reviewed published images on the subject. Their assessment was that the majority were in fact instances of what they termed an "accessory cavitated uterine mass" (ACUM). Lesions classified under this category were mostly isolated cysts located in the right or left insertion of the round ligament. The uterine cavity was otherwise normal. Cysts similar to those described as JCA but identified in adults have been termed "adult cystic adenomyosis". Still these represent the same pathologic lesions.

Acién et al[20] and Bedaiwy et al[21] suggested that most published cases of non-communicating accessory uterine cavities or isolated cystic adenomyomas, as well as some cases of uterus-like masses, are actually the same pathology: an ACUM which contains a functional endometrium. They also argued that these need to be differentiated from cases of müllerian anomaly with a rudimentary cavitated uterine horn. Acién et al[19,22] suggested that these lesions may arise from duplication and persistence of ductal müllerian tissue in a critical area related to the attachment of the round ligament and proposed an association between this anomaly and a 'dysfunction' of the development of the gubernaculum.

An early description is that by Oliver, who in 1912[23] reported an "accessory uterus distended with menstrual fluid" which he enucleated from the right broad ligament. In modern literature, a "uterus-like mass" affecting the ovary was first reported in 1981 by Cozzuto.[24] Fukunaga et al[25] described a 25-mm mass growing into the endocervical canal in a 47-year-old woman. This uterus-like mass had both superficial cervical endometriosis and florid smooth muscle metaplasia.

There are a few descriptions of a uterus-like mass related to the lower uterine segment. Jung et al[26] described 2 cases of uterine adenomyoma with uterus-like features. The first was a 43-year-old woman who had a 3-cm submucous mass attached to the posterior wall of the lower uterine segment. The cyst was lined by well-developed secretory endometrium and was surrounded by smooth muscle. The second was a 52-year-old woman who had an enlarged uterus with an 8-cm mass protruding from the left side of the lower uterine segment. The cyst was lined by thin basalis-type endometrial glands and stroma. Menn et al[27] reported a case of a 37-year-old woman in whom a 5-cm mass was demonstrated by MRI in the right uterine wall. The diagnosis of a uterus-like mass was based on histology. They speculated that the structure may have resulted from metaplastic changes, a congenital anomaly, or heterotopia. Tijani et al[28] described the case of a 35-year-old woman who had a large,