



Large uterine juvenile cystic adenomyoma in an adolescent

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ABSTRACT

Juvenile cystic adenomyoma (JCA) is a rare uterine pathology with < 40 cases reported in the current literature since 1996 when Tamura described it. We report a 13-year-old girl with a history of chronic pelvic pain and dysmenorrhea for 12 months. After diagnostic evaluation and identification of the cystic structure (~6 cm) within the myometrium, fertility-sparing surgery was successfully performed. Histopathological examination of the uterine cyst was consistent with the diagnosis of JCA. The postoperative course was uneventful, and the patient was disease-free three years after surgery. JCA is a rare condition but should be considered in the differential diagnosis in adolescents with moderate to severe dysmenorrhea. Despite diagnostic advances, the awareness of the disorder remains low. Thus, our case report aims to increase awareness of this rare pathology with unclear etiology.

1. Introduction

The presence of ectopic endometrial glands within the uterine myometrium, with adjacent smooth muscle hypertrophy, is a benign disorder known as adenomyosis [1]. Diffuse adenomyosis is the most common form, while focal or nodular adenomyosis, particularly the cystic variant, is extremely rare, especially in the pediatric population [2]. Small hemorrhagic foci (≤ 5 mm) within the myometrium, caused by secretory changes and menstrual bleeding within this heterotopic endometrial tissue, are typically found in adult parous women of > 30 years of age with diffuse adenomyosis [3]. On the contrary, large cysts, termed as an adenomyotic cyst, cystic adenomyosis, juvenile cystic adenomyosis/adenomyoma (JCA), or adenomyoma, can be found within the uterine myometrium, with adjacent smooth muscle hypertrophy, of adolescents and women < 30 years. Although the first published report on JCA comes from Tamura et al., in 1996 [4], Takeuchi et al., in 2009 defined the diagnostic criteria of JCA based on age (< 30 years), presence of cystic lesion ≥ 1 cm in diameter independent of the uterine lumen and covered by hypertrophic myometrium on diagnostic images, and associated with severe dysmenorrhea [5].

Although some women may be asymptomatic, most women present with severe dysmenorrhea, pelvic pain, menorrhagia, uterine enlargement, and infertility [6]. Because of its rarity, JCA presents a diagnostic challenge and is often misdiagnosed as a congenital anomaly with hematometra in a non-communicating horn, fibroid with hemorrhagic or fatty degeneration, congenital uterine cysts, and intramyometrial hydrosalpinx [7]. These diagnoses can be distinguished by magnetic resonance imaging (MRI), the imaging

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techniques of choice for assessing cystic adenomyosis and making the correct diagnosis in adolescent girls [8]. Due to the evidence that drug therapy may temporarily be effective, surgical treatment remains necessary for a definite improvement in pain [9].

Herein we report a very rare case of a 13-year-old female who presented with dysmenorrhea and pelvic pain that progressively increased over 12 months. After diagnostic evaluation and identification of the cystic structure within the myometrium, fertility-sparing surgery was successfully performed. We also present a brief literature review on JCA.

2. Case report

A 13-year-old adolescent girl presented to the emergency department for acute abdominal pain with a previous history of chronic pelvic pain and dysmenorrhea for 12 months. The pain symptoms started one year after menarche at age 11 and progressively worsened with regular but increasingly heavy and painful menstrual cycles. The patient was not sexually active. She was taking no medications and had no allergies. Her medical history was unremarkable. Physical examination revealed no significant findings. The laboratory studies and tumor markers were normal. The abdominal ultrasound examination demonstrated a large, complex cystic lesion (6.1×6.3 cm) filled with a thick liquid content in the right top of the uterus (Fig. 1). A paratubal cyst (3.0×3.4 cm) was also observed on the left side. Pelvic MRI showed the presence of a large, round-shaped cystic structure (6.6×6.0 cm) within the myometrium of the right uterine fundus with an internal fluid-fluid level (Fig. 2A and B). Fluid within the cyst was hyperintense on T1 and showed intermediate intensity on T2-weighted images, indicating a hemorrhagic and/or proteinaceous fluid (Fig. 2). Previously ultrasound visualized the left paratubal cyst measured 3.0×3.1 cm on MRI. No other urogenital anomalies were noted. The patient underwent an exploratory laparotomy that revealed an asymmetrically enlarged uterus with a cystic mass arising from the right uterine fundus.

The cyst was punctured, and viscous chocolate-like material was noted coming out of the cyst. After that, the cyst was enucleated and completely resected after careful blunt and sharp dissection (Fig. 3A). The posterior cyst wall was highly adherent and entirely separated from the endometrial cavity. Finally, the myometrial defect was sutured in two layers with a 3-0 vicryl suture following meticulous hemostasis. In addition, dissection of the left paratubal cyst was performed. The postoperative course was uneventful, and the patient is free of symptoms three years after surgery. Histopathological examination of the uterine cyst was consistent with the diagnosis of JCA (Fig. 3B).

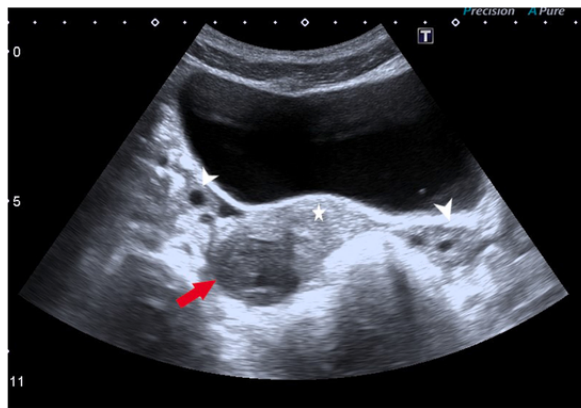


Fig. 1. Ultrasound image showing hypoechoic, round lesion in uterine wall/myometrium (red arrow) with normal ultrasound presentation of both ovaries (white pointed arrows) and uterine corpus (white star). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

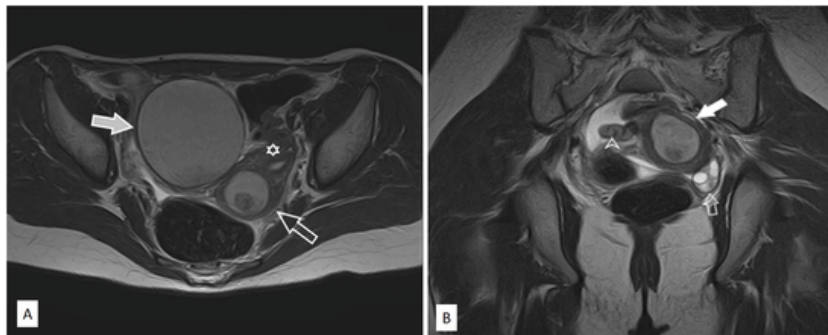


Fig. 2. A-B: MRI scans. (A): T2 weighted axial image showing a complex cystic mass in the myometrium (white hollow arrow), a large, round-shaped periuterine cyst (white arrow); a normal uterine corpus (white star-maker); (B): T2 weighted coronal image showing a normal left ovary (white hollow arrow), complex myometrium cystic mass (white arrow), and partially presented twisted cord from a periuterine cyst (pointed arrow).

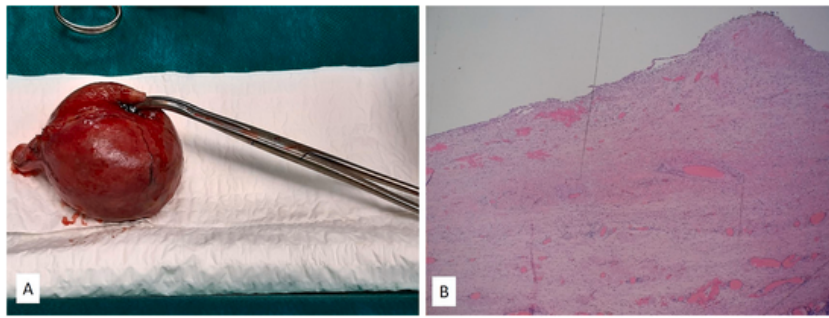


Fig. 3. A-B. (A): The gross appearance of the resected cystic mass was histopathologically confirmed as a juvenile cystic adenomyoma (B).

Table 1

Review of the publications reporting juvenile cystic adenomyoma in adolescents ≤ 16 years (PubMed/MEDLINE, Web of Science and Scopus were comprehensively surveyed).

Author, year	No of patients	Age (years)	Size (cm)	Laterality	Imaging	Medical management	Surgery	Outcome
Tamura et al., 1996 [4]	1	16	3	Left	US MRI	NA	LT	Complete
Potter and Schenken, 1998 [14]	1	15	4 × 3	Left	US	Incomplete	LT	Complete
Fisseha et al., 2006 [15]	1	13	2.1 × 2	Left	US MRI	Appropriate	Not attempted	Stable on meds
Ho et al., 2008 [2]	1	16	NA	Right	US	Incomplete	LT	NA
Akar et al., 2010 [16]	1	15	4.8 × 3.4	Right		Incomplete	RAL	NA
Dadhwal et al., 2017 [17]	1	16	4 × 3	Left	US MRI	NA	LPS	Complete
Debleare et al., 2019 [12]	1	16	2 × 1.9	Right	US MRI	Incomplete	LPS	Complete
Protopapas et al., 2020 [18]	1	14	3.8 × 3.4	Left	MRI	Incomplete	LPS	Complete
Park et al., 2021 [19]	1	14	3	Right	US MRI	NA	LPS	Complete
Arya et al., 2021 [3]	1	16	5	Right	US CT	Incomplete	LPS	Stable on meds
Zvizdic et al., 2022 ^a	1	13	6.6 × 6	Right	US MRI	NA	LT	Complete

US = Ultrasonography; MRI = Magnetic resonance imaging; CT = Computed tomography; LT = Laparotomy; LPS = Laparoscopy; RAL = Robotic-assisted laparoscopy; NA = Not available.

^a The current study.

3. Discussion

This case illustrates that JCA should be considered in the differential diagnosis in adolescents with moderate to severe dysmenorrhea and chronic pelvic pain, especially adolescents with identified uterine cystic structures. However, this condition is exceedingly rare. Less than forty cases have been reported in the scientific literature since JCA was first described by Tamura et al., in 1996 [4]. The cases in adolescents < 16 years are particularly rare; our literature search revealed only ten reported cases in this category (summarized in Table 1).

Although several investigators have proposed diverse theories to determine the origin of these unusual uterine abnormalities, the etiology of JCA is not yet fully understood. There has long been a debate over whether JCA is an acquired or congenital Müllerian defect. Takeuchi et al. believe that JCA is a rare variant of adenomyosis rather than a congenital abnormality [5]. In contrast, Acien et al. suggested that the JCA represented a miniature partial uterus with a functional endometrium, corroborating this view that the JCAs are found in young women with severe dysmenorrhea and pelvic pain before the age of 30 years [10]. Either way, further research is needed to confirm and expand on these findings. However, evidence, not theory, is needed for an accurate diagnosis and suitable treatment decisions.

Uterine cystic structures, especially in adolescents with severe dysmenorrhea, cannot be accurately characterized by ultrasound and usually require MRI. MRI defined diagnostic criteria and made inter-observer variability less significant [8]. On MRI, JCA presents as a well-circumscribed cystic lesion filled with hemorrhagic fluid in various stages of organization within the myometrium [8]. However, the lack of awareness of this condition among health professionals is due to the rarity of the disease and the lack of diagnostic and therapeutic protocols. This might cause significant delays in the management of JCA [11].

Treatment of JCA is determined by the patient's age, the severity of symptoms, and the size and location of the adenomyotic cyst. The main goal of medical treatment is to inhibit endometrial proliferation and shedding using nonsteroidal anti-inflammatory drugs (NSAIDs), continuous oral contraceptive pills, and gonadotropin-releasing hormone (GnRH) agonists [5]. The medical treatment usually provides partial and temporary alleviation of symptoms that tend to recur after the therapy is discontinued [12]. Fertility-sparing surgery (open or laparoscopic) is the preferred treatment modality in young patients with severe symptoms or treatment-refractory disease [12]. The surgical approach involves enucleation and resection of JCA as applied in the presented case. Similar to our case, a cavity filled with hemorrhagic fluid that has no communication with the uterine cavity, is bordered by endometrium, and is surrounded by myometrium are histopathologic criteria for diagnosing JCA [13].

In conclusion, JCA is a rare condition but should be considered in the differential diagnosis in adolescents with moderate to severe dysmenorrhea. Despite diagnostic advances, the awareness of the disorder remains low. Thus, our case report aims to increase awareness of this rare pathology with unclear etiology.

Patient consent

Consent to publish the case report was obtained from the patient's mother.

Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

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Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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