



Rudimentary Horn Adenomyosis in A 20-Year-Old Patient With Unicornuate Uterus: A Case Report

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Abstract

Background: A unicornuate uterus with a rudimentary horn is a rare Mullerian anomaly with few cases reported in the literature. The symptoms vary depending on the functionality of the endometrial cavity and the presence of an obstruction. Adenomyosis is a disease defined as areas of endometrial glands and stroma present within the myometrium. Reports describing adenomyosis in relation to the Mullerian anomalies are scarce.

Case Report: We present a 20-year-old middle eastern woman admitted for chronic pelvic pain and abdominal distension. Pelvic ultrasound revealed a unicornuate uterus with an adjacent mass of 10 x 8 cm. An MRI (Magnetic Resonance Imaging) was performed, and a left unicornuate uterus was demonstrated. On the right side, a large non-communicating rudimentary horn with features suggesting diffuse adenomyosis was found. Laparoscopic removal of the horn was performed successfully, and the extraction of the specimen was made by a power morcellation containment system. The patient was discharged uneventfully the second day after the surgery, and the pathology result confirmed the presence of diffuse adenomyosis.

Conclusion: Non-communicating rudimentary horn is a rare finding manifesting as different symptoms in young women. The enlargement of the horn can have obstructive consequences on an active endometrium, and this would lead to the formation of a hematometra and the distension of the cavity, and sometimes adenomyosis.

Keywords: Adenomyosis, laparoscopy, Mullerian anomalies, rudimentary horn, unicornuate uterus

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Introduction

Mullerian duct anomalies are commonly underdiagnosed mainly due to the absence of clinical suspicion and lack of pathognomonic clinical and radiological features. Hence, they are rarely considered in the differentials of gynecologic or obstetric problems [1]. The exact prevalence of Mullerian duct anomalies is difficult to determine. According to a systematic review done in 2011, the prevalence of Mullerian duct anomalies was 8% in infertile women, 12.3% in women with a history of miscarriage, and 24.5% in women with a history of miscarriage and infertility [2]. It is crucial however for healthcare professionals to be aware of their presence because of their association with wide-ranging gynecological and obstetric complications including preterm birth, fetal growth restriction, malpresentation, and cesarean delivery.

A unicornuate uterus with a rudimentary horn is a rare Mullerian anomaly with few cases reported in the literature. The symptoms vary depending on the functionality of the endometrial cavity, and the presence of complications such as an obstruction with a retrograde accumulation of blood leading to hematocolpos, hematometra, hematosalpinx, and secondary endometriosis [3].

Adenomyosis is classically defined as areas of endometrial glands and stroma located deep within the myometrium leading to uterine enlargement secondary to hyperplasia and hypertrophy of the myometrium. These areas can either be widely present throughout the myometrium or localized, forming a focal nodular collection called adenomyoma [4]. It is usually associated with various symptoms such as menorrhagia, dysmenorrhea, dyschezia, dyspareunia, and possibly infertility. However, in one-third of the case, it can remain asymptomatic [5]. The overall adenomyosis incidence is 1.03% and disproportionately higher in black women [6]. Risk factors include exposure to estrogen, high parity, and a history of uterine surgery.

The onset of symptoms is typically in females between 40 and 50 years of age with MRI being the modality of choice to diagnose adenomyosis [7]. Reports describing adenomyosis in relation to the Mullerian anomalies are scarce.

We hereby describe a case of adenomyosis present within the uterine rudimentary horn in a patient with an unusual but previously described finding of unicornuate uterus with a non-communicating rudimentary horn. Informed consent was obtained from the patient for publication of this case report and any accompanying images.

Case Presentation

We are reporting a case of a 20-year-old middle eastern unmarried female referred to our clinic for chronic pelvic pain and progressive painful distention of the lower abdomen. Her past medical and family history showed regular menstrual cycles and menarche at the age of 12. The clinical exam demonstrated a normal female external genitalia. However, upon palpation of the pelvis, a hard fixed painful mass was felt.

The pelvic ultrasound requested after this finding showed a unicornuate uterus with an adjacent 10x8 cm mass. Subsequently, an MRI was ordered to better characterize the anatomy of the pelvis, and identify the anatomic location of the pelvic mass. Results revealed a left unicornuate uterus. The endometrial cavity appeared normal, the same for the cervix and the vagina. On the right side, it showed a large non-communicating rudimentary horn with features suggesting diffuse adenomyosis (Figure 1).

After counseling the patient, a decision of laparoscopic removal of the rudimentary horn was taken. During the surgery, an adhesiolysis was needed to remove extensive adhesions between the omentum and the enlarged horn (Figure 2). Laparoscopic removal of the horn was then performed successfully, with the extraction of the specimen by a power morcellation containment system (Figure 3). The patient

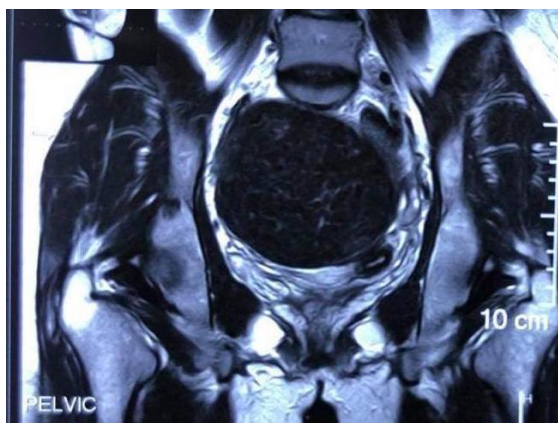


Figure 1: Pelvic MRI showing enlarged rudimentary horn (Arrow).

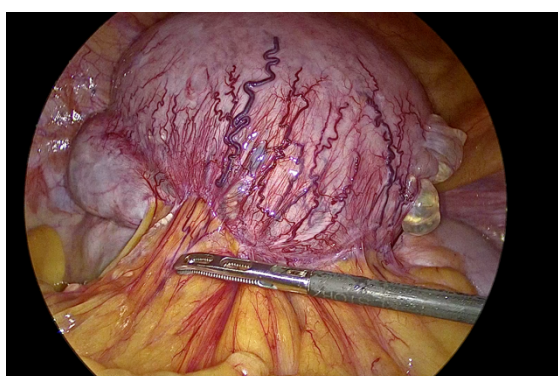


Figure 2: The first laparoscopic view showing the adhesions between the omentum and the rudimentary horn.

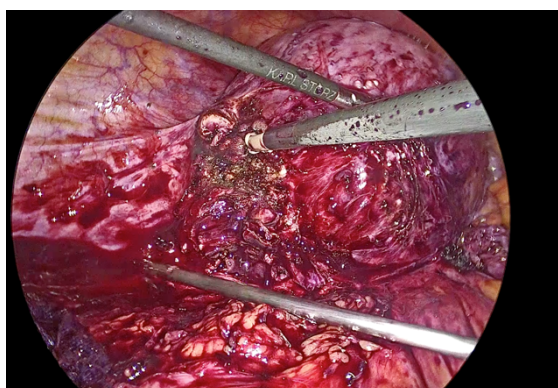


Figure 3: Laparoscopic separation of the rudimentary horn.

The patient was then discharged uneventfully the second day after the surgery. The pathology result confirmed the presence of diffuse adenomyosis.

Discussion

A unicornuate uterus is a rare Mullerian anomaly with a fluctuating prevalence of 2.4% to 13.7% of all uterine anomalies,

although its true incidence rate is unknown [8]. Buttram and Gibbons classify a unicornuate uterus into different subtypes based on the anatomical findings: unicornuate uterus either with a rudimentary horn or without a rudimentary horn [9]. Those with a rudimentary horn are further divided as with an endometrial cavity or without an endometrial cavity; Furthermore, those with an endometrial cavity may be communicating or noncommunicating with the opposite horn. Partial development of the Mullerian duct may result in different degrees of rudimentary horn connected to the opposite horn. The isolated unicornuate uterus is the most common presentation, with an estimated reported frequency of 35%. Whenever a rudimentary horn is present, it is found to be noncavitary in 33% of cases, cavitary noncommunicating in 22%, and cavitary communicating in 10% [10].

In general, Mullerian anomalies are not routinely considered risk factors for the development of uterine adenomyosis, although they can occur [11]. Reports describing adenomyosis in relation to the Mullerian anomalies are rare. To the best of our knowledge, this is the first case that describes a unicornuate uterus with a non-communicating adenomyotic rudimentary horn.

Our case is different from previously described cases of adenomyosis in Mullerian anomalies which reported adenomyosis present in a rudimentary horn of a patient with the Mayer-Rokitansky-Kuster-Hauser Syndrome, in a unilateral horn of a patient with a bicornuate uterus, or within a uterine septum in a patient with a septate uterus [12-14].

The unicornuate uterus is classified as U4 in the new ESHRE/ESGE classification (Figure 4), however, even if this classification is user-friendly, the presence of a rudimentary horn can add some confusion [15]. Even though the rudimentary horn was not communicating in our case, we couldn't confirm if there was a cavity or not, thus, we couldn't classify it as U4a (with cavity) or U4b

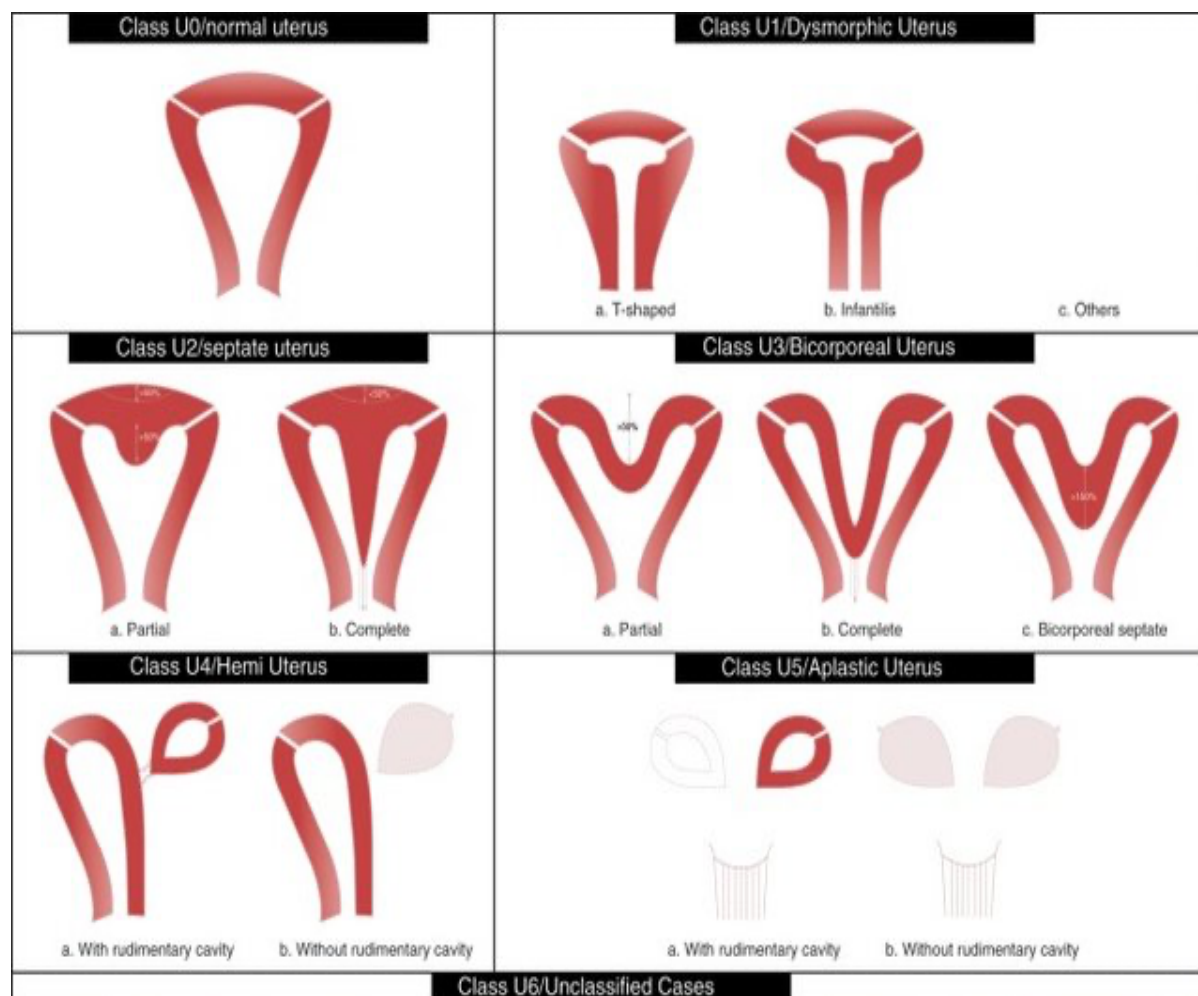


Figure 4: ESHRE/ESGE classification of uterine anomalies

(without cavity). This brought the whole discussion about whether our case was primary adenomyosis in the uterine wall of the horn or secondary adenomyosis due to an active endometrial cavity where the endometrial cells invaded the muscle.

The specific pathogenesis of adenomyosis remains a challenge with two major prevailing hypotheses: invagination and metaplasia. In the invagination hypothesis, adenomyosis was largely the result of repeated tissue injury, and repair leading to the invagination of the endometrial layer into the myometrium thus resulting in adenomyosis. This theory is backed up by the fact that adenomyosis is observed more frequently in cases of high parity, and a history of uterine surgery as previously mentioned. In the metaplasia theory, however, adenomyosis was postulated to originate from the metaplasia of fragmented and displaced pluripotent epithelial of the Mullerian ducts or remnants during their

fusion, which might be the case in this patient [16]. Adenomyosis in a patient with no functional endometrium (Mayer-Rokitansky-Kuster-Hauser Syndrome) also challenges the theory that adenomyosis arises from an invasion of the uterine mucosa into the musculature and supports the theory of metaplasia of Mullerian remnants. Laparoscopic removal of the rudimentary horn has become the standard of care after its first documentation in 1990 because of its short operative time, less blood loss, and postoperative pain. Studies have reported reproductive outcome improvement after removing the rudimentary horn, but little information on the topic is present in the literature [17].

In our case, the major difficulties faced during the surgery were the large size of the rudimentary horn and the omentum adhesions over it despite the absence of any past surgical history.

Conclusion

Non-communicating rudimentary horn is a rare entity that can lead to different symptoms in young women. The enlargement of the horn can have obstructive consequences on an active endometrium, and this would lead to the formation of a hematometra and the distension of the cavity but also could lead to adenomyosis. Additional reports of cases and a larger systematic review of the literature are needed to prove the theory that obstructive congenital anomalies are a major cause of secondary endometriosis and adenomyosis.

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