Non-communicating Rudimentary Horn Mimicking a Degenerated Subserous Leiomyoma: A Case Report

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Unicornuate uterus with a rudimentary horn is a rare Müllerian anomaly, with an incidence of 0.06%. Due to this relative rarity, an accurate preoperative diagnosis of unicornuate uterus with rudimentary horn still remains to be a diagnostic challenge. Reported here is a case of a 27-year-old nulligravida who complained of severe debilitating dysmenorrhea 5 months prior to admission, necessitating frequent visits to the emergency department for administration of intravenous pain medications and antiemetics. She was misdiagnosed preoperatively with subserous leiomyoma, mainly based on clinical presentation, and findings on ultrasound and pelvic MRI. On exploratory laparotomy, a definitive diagnosis of unicornuate uterus with an enlarged rudimentary horn was established, and patient underwent resection of rudimentary horn, with enterolysis, due to dense adhesions in the left adnexa and cul de sac. We emphasize the need for early diagnosis and prompt management of cases like this to prevent complications, and optimize fertility and quality of life of affected patients.

Key words: unicornuate uterus, Müllerian anomalies, rudimentary horn

Introduction

The prevalence of congenital uterine malformations is about 0.5% in the general population.[1] The least frequent form of these malformations is the unicornuate uterus with a rudimentary horn, with an estimated worldwide incidence of about 0.06%.[2] This condition arises when the Müllerian ducts fail to develop completely or partially between the 7th and 8th weeks age of gestation.[1] Often, women with unicornuate uterus also have an associated rudimentary uterine horn. These rudimentary horns can either communicate with the main unicornuate uterus or exist as non-communicating structures. Additionally, they may contain functional endometrial tissue, which can contribute to various gynecological complications.[3]

Patients with unicornuate uterus and functional, non-communicating horns, often will present with chronic pelvic pain, usually in the form of secondary dysmenorrhea, or dyspareunia. The severe pelvic pain typically do not respond to standard treatments such as non-steroidal anti-inflammatory drugs (NSAIDs) and contraceptive pills, which necessitates prompt surgical management. With a delay in management, patients may suffer gynecologic, and obstetric complications.[3]

Presented here is a case of a 27-year-old nulligravida who experienced progressive severe dysmenorrhea, and was preoperatively misdiagnosed to have subserous leiomyoma, in degeneration, based on clinical presentation and imaging studies. Intraoperatively, the pelvic pain was determined to be due to the hematometra in the non-communicating rudimentary horn, and complications of retrograde menstruation such as hematosalpinx and deeply infiltrating endometriosis. We emphasize the need for early diagnosis and prompt management of cases like this to prevent complications, and optimize fertility and quality of life of affected patients.

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The Case

This is a case of a 27 year old nulligravida, who started to experience severe debilitating dysmenorrhea 5 months prior to admission. This was associated with nausea and vomiting, and necessitated frequent visits to the emergency department for administration of intravenous pain medications and antiemetics, especially during first few days of menses. A transvaginal ultrasound done during this time revealed a subserous myoma with degeneration, measuring 67.5mm x 54.3mm x 50.5mm, with central dilation of 27.5mm^3 containing low level echoes and echogenic periphery (Figure 1). Within the left ovary was a unilocular endometriotic cyst that measured 3.3mm x 25.5mm x 19.5mm. Magnetic Resonance Imaging was likewise requested, and which revealed a pedunculated mass exhibiting peripheral signals similar to the myometrium arising from the left side of the uterine fundus (Figure 2), thus corroborating the ultrasound findings of a subserous leiomyoma. She was referred to a reproductive endocrinology specialist for laparoscopic myomectomy, but due to financial constraints, patient opted for exploratory laparotomy instead. Antimullerian hormone (AMH) level was 2.5 ng/mL.

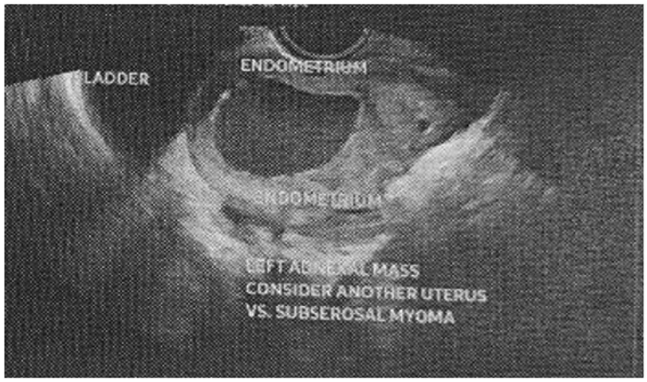

Figure 1. Transvaginal ultrasound showing a pelvic mass, probably a subserosal myoma with degeneration, measuring 67.5mm x 54.3mm x 50.5mm, with central dilation of 27.5 mm^3 containing low level echoes and echogenic periphery.

The past medical history and family history were unremarkable. She was a nulligravid, and had 1 sexual partner. She never had any pap smears done. She was a non-smoker and non-alcoholic beverage drinker. She denied use of any illicit drugs nor any form of contraception. She had regular monthly menses since her menarche at 14 years old, using only 2-3 pads/day, lasting for 3 days, noting only occasional mild dysmenorrhea.

On physical examination, patient was conscious, coherent, and ambulatory, with stable vital signs. She had normal built and stature, with BMI of 22.1 kg/m^2. Cardiorespiratory, gastrointestinal and neuromuscular examination findings were all normal. On pelvic examination, external genitalia was normal-looking, vaginal walls were smooth, there was a single cervix which was firm and smooth, and measured about 2cm x 2cm (Figure 3). There was no cervical motion tenderness. The uterus was asymmetrically enlarged to 12-14 weeks size, with limited mobility. There were no adnexal masses palpated, but slight tenderness was elicited on the left adnexal area. The culdesac was nodular, but non tender. Rectovaginal septum was smooth and left parametrium was nodular.

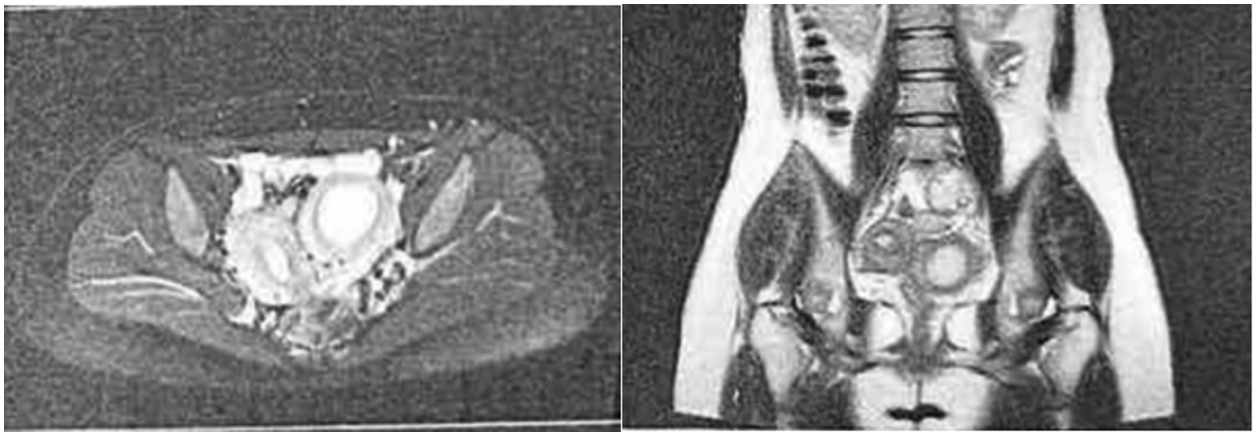

Figure 2. MRI images which revealed a pedunculated pelvic mass exhibiting peripheral signals similar to the myometrium arising from the left side of the uterine fundus.

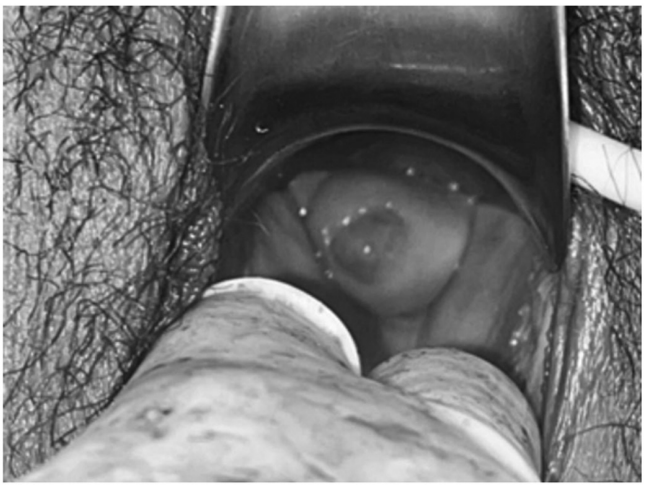

Figure 3. Speculum exam showing a single cervix, measuring approximately 2cm x 2cm.

Given the findings on imaging studies, the surgical plan on admission was exploratory laparotomy with myomectomy and cystectomy. Intraoperatively, there was a unicornuate uterus that measured approximately 5cm x 4 cm, with normal right fallopian tube and right ovary. No leiomyoma was seen. Instead, there was a left dilated, non-communicating, rudimentary uterine horn, which measured 7cm x 6cm, and contained approximately 10cc of chocolate-like fluid on cut section (Figure 4). There were dense adhesions noted between the sigmoid colon and the posterior wall of the uterus, uterosacral ligaments and left adnexa, that partially obliterated the culdesac. The left fallopian tube attached to the rudimentary horn was dilated and converted to a cystic mass that measured 5cm x 5cm, containing chocolate-like fluid on cut section. Intraoperative referral to colorectal surgery service was done to help with enterolysis. Once the operative site was adequately cleared of adhesions, surgeons proceeded to resection of the rudimentary horn. First, the bilateral round ligaments were identified and the uterovesical pouch was opened and the bladder was pushed down. The left round ligament attached to the rudimentary horn, as well as the left uteroovarian ligement were doubly clamped and ligated. The attaching pedicle of the left non-communicating horn was doubly clamped, transfixed-ligated and excised together with the left hematosalpinx. Cystectomy of the left endometriotic cyst was done. Endometriotic implants were fulgurated. Bilateral kidneys were palpated and both ureters were visualized. Intraoperative blood loss was 800ml, which necessitated blood transfusion of 1 unit pRBC. The patient had an unremarkable post-operative course, and was discharged on third day post-op. Histopathologic examination of the specimens confirmed Endometriosis (Figures 5 & 6).

KUB ultrasound done during her outpatient follow-up showed normal kidneys, ureters and bladder. Patient was also given 3 monthly doses of GnRH agonist 3.75mg IM, to manage her endometriosis.

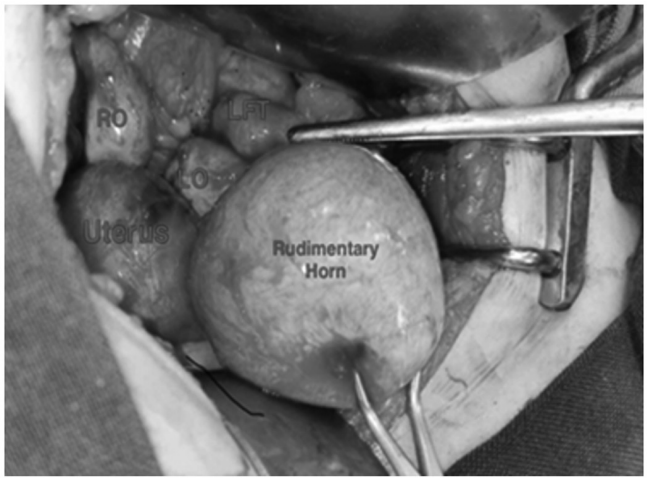

Figure 4. Intraoperative findings which showed a unicornuate uterus that measured approximately 5cm x 4cm, with normal right fallopian tube and right ovary. No leiomyoma was seen. Instead, there was a left non-communicating, rudimentary uterine horn, which measured 7cm x 6cm, and contained approximately 10cc of chocolate-like fluid on cut section.

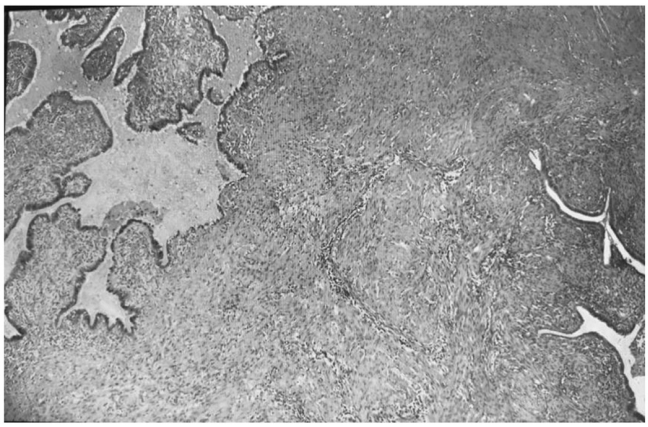

Figure 5. Microscopic image (LPO) of left fallopian tube showing presence of endometrial glands within the fallopian tube wall, suggestive of endometriosis.

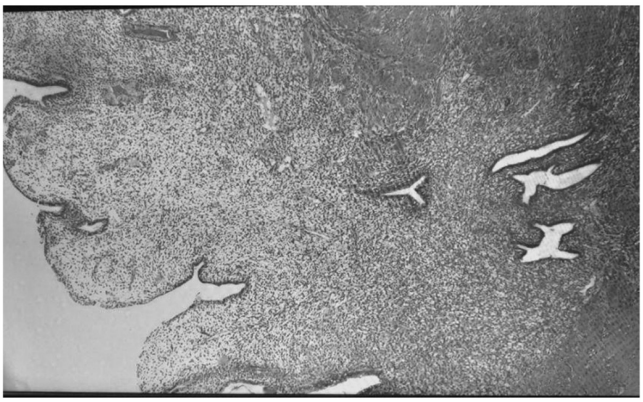

Figure 6. Microscopic image (LPO) of rudimentary horn showing presence of abnormal endometrial glands in the surrounding myometrial stroma, suggestive of adenomyosis.

Discussion

The female reproductive organs develop from bilateral mullerian ducts beginning at the 6th week age of gestation, which fuse and cavitate to properly form the uterus, cervix, and upper two-thirds of the vagina. Congenital uterine anomalies mostly result from lack of development, failure of midline unification, or incomplete resorption, of these müllerian ducts.[4] Failure of Müllerian tube formation causes aplasia or atresia of one side, resulting in a unicornuate uterus.[5] Incomplete atresia of a Müllerian duct leads to a rudimentary horn which may be broadly connected, or connected through streak tissue with the unicornuate uterus. Usual causes of congenital uterine anomalies may include infections, ionizing radiation, or teratogenic drugs (e.g., thalidomide and diethylstilbestrol) during early pregnancy.[4]

In 1988, the American Fertility Society (AFS) published what is now known as the iconic 7-classification system for congenital uterine anomalies. But since there remains a wide range of müllerian anomalies that is still largely unknown, the American Society for Reproductive Medicine (formerly AFS) Task Force on Mullerian Anomalies Classification designed a new classification system to expand and update the old one, to include cervical and vaginal anomalies. This new system, called the ASRM Müllerian Anomalies Classification 2021 (MAC2021) classifies the müllerian anomalies into 9 categories (Figure 7). Since most müllerian

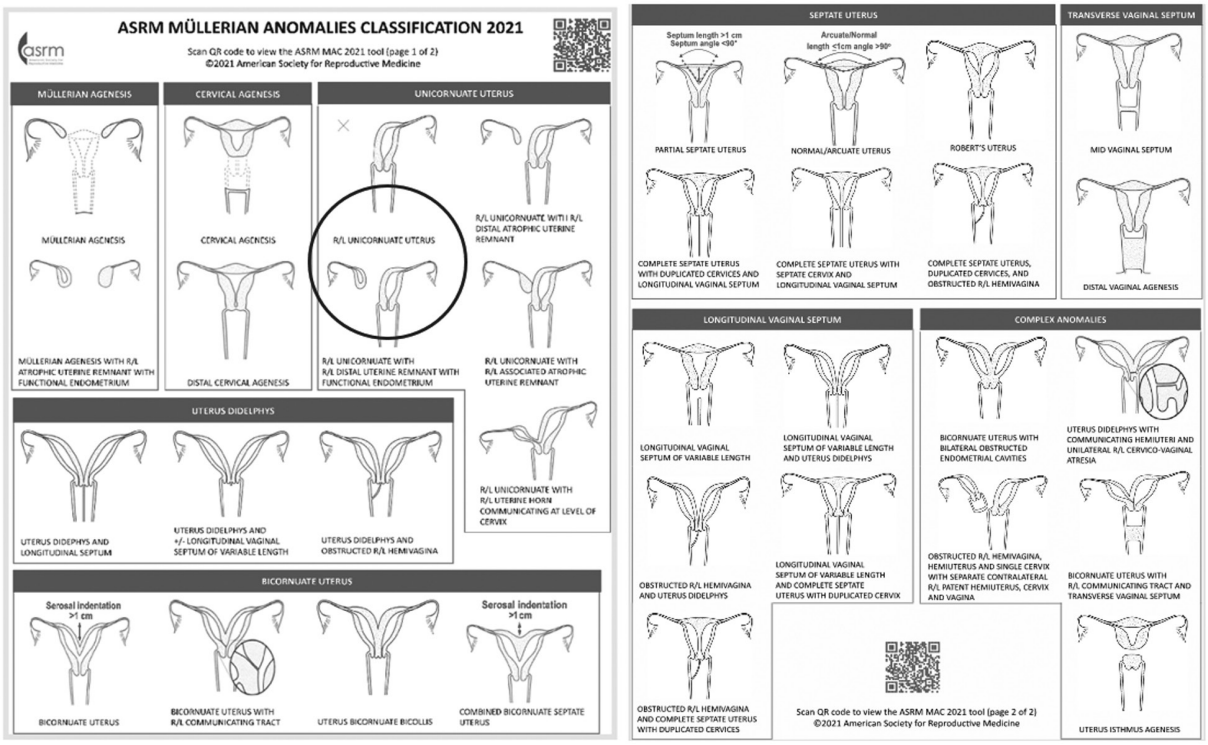

Figure 7. The new ASRM classification for müllerian anomalies, published in 2021 (MAC2021).[3] This new system expanded and updated the 1988 AFS 7-classification system, to include cervical and vaginal anomalies. Our index patient falls under the general classification of unicornuate uterus, with the subtype as indicated above with a circle.

abnormalities have combined elements, some anomalies appear in more than 1 category, thereby allowing cross-referencing and comparison between categories to help determine diagnosis and treatment options. Because of this, the categories are not numbered, but instead are identified by descriptive terminology, unlike the original AFS system.[6]

Unicornuate uterus with a rudimentary horn is a rare Müllerian anomaly, with an incidence of 0.06%.[2] Seventy-five to ninety percent of unicornuate uteri with rudimentary horns are non-communicating.[7] Non-communicating cavitated or functioning rudimentary horns are the most clinically significant subtype, as they are more likely to be associated with severe pelvic pain from hematometra or from endometriosis due to retrograde menstruation, as seen in our index case.

Perhaps because of this relative rarity, and hence the limited exposure of sonologists, radiologists and clinicians to cases such as this, an accurate preoperative identification of unicornuate uterus with rudimentary horn still remains to be a diagnostic challenge. Consequently, individuals may endure a delay in diagnosis for weeks, months, and even years. Some cases may undergo inappropriate or inadequate surgical interventions, and may have persistent issues including chronic pain and/or loss of reproductive function as a result. Early diagnosis is crucial in order to avoid consecutive damage of the reproductive system and further painful complications due to endometriosis.[8]

Our index case was preoperatively misdiagnosed as having a subserous uterine leiomyoma with degeneration as the acute cause of her severe pelvic pain. To obtain a correct preoperative diagnosis, the patient's history is crucial: women with unilateral obstructive anomalies like a non-communicating, functioning rudimentary horn can experience cyclic or noncyclic pelvic pain and severe chronic dysmenorrhea, with pain symptoms usually starting at or near the age of menarche, and often worsening over time.[9] For women with partners, dyspareunia and infertility are common complaints.[10] However, our patient only reported severe dysmenorrhea less than 6 months prior to consult, which may have deceived the clinicians to think that the root cause was something that could acutely happen, like a degeneration of a large leiomyoma.

Imaging studies are crucial in establishing a correct preoperative diagnosis. Diagnosing such anomalies can be challenging with a single imaging modality. The usual first-line diagnostic method is a transvaginal grey-scale sonography. The use of either three-dimensional ultrasound and pelvic magnetic resonance imaging (MRI) has increased the accuracy of diagnosis by providing greater tissue details.[11] Pelvic MRI is the preferred imaging, or even the gold standard technique for identifying, classifying, and guiding the surgical treatment of müllerian anomalies. However, in our case, the anomaly was misidentified as a pedunculated uterine leiomyoma. A retrospective study by Kim, et al (2021)[12] found an 8.4% discrepancy rate between the final diagnosis after surgery and the initial MRI findings, among patients with müllerian anomalies. Interpretational errors can occur in MRI due to factors such as the presence of long-standing hematoma, which can affect signal quality, resolution, and visualization.[12] Additionally, interobserver bias and the experience of the reader can impact interpretation. Often, surgery provides a definitive diagnosis of uterine malformations.[13]

Intraoperatively, as what usually happens in most cases of müllerian anomalies, a unicornuate uterus with an enlarged rudimentary horn was established, instead of a subserous myoma. In this patient, the severe debilitating dysmenorrhea was caused mainly by the hematometra in the noncommunicating horn, the hematosalpinx, and the deep infiltrating endometriosis that developed by retrograde menstruation due to the outlet obstruction.

Some of the surgeries to correct these müllerian anomalies require specific expertise, and therefore should only be performed with the help of an experienced surgeon. The surgical resection of non-communicating rudimentary uterine horns aims to alleviate pain symptoms and optimize fertility. If not diagnosed and managed early, patients with non-communicating rudimentaruy horns that contain functional endometrium are at increased risk for complications such as ectopic pregnancy, hematosalpinx, endometriosis, chronic pelvic pain, intraoperative adhesions, and infertility, secondary to retrograde menstruation. Therefore, consideration of prophylactic resection of a non-communicating functional uterine horn should be considered in an asymptomatic, reproductive age patient once

the diagnosis is made. The successful management of these anomalies requires careful diagnostic evaluation and appropriate surgical intervention to prevent these potential complications and improve patient outcomes.[3,14]

An integral part of the wholistic management of cases like this include an investigation of the renal or urinary tract. There is a high association (30–40%) of müllerian anomalies with urinary tract anomalies, and therefore all women who are diagnosed with a uterine anomaly should have an intra-venous urogram, or a KUB ultrasound.[15] Our index case fortunately, had normal kidneys, ureters and urinary bladder on further investigation.

Conclusion

This case underscores the importance of considering obstructive uterine anomalies in the differential diagnosis of patients presenting with severe debilitating pelvic pain and dysmenorrhea. Due to the relative rarity of this case, obtaining an accurate preoperative diagnosis is oftentimes challenging, even with thorough history and physical exam and use of imaging modalities, such as MRI and ultrasound. Nevertheless, we emphasize the need for prompt management of cases like this to prevent complications such as ectopic pregnancy, hematosalpinx, endometriosis, chronic pelvic pain, intraoperative adhesions, and infertility, secondary to retrograde menstruation. This approach not only alleviates symptoms but also optimizes fertility and quality of life for affected patients.

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