Case report

Unicornuate uterus with a functional, non-communicating rudimentary cavity associated with ovarian endometriosis and ipsilateral renal agenesis

H D Manamendra\textsuperscript{a}, W Abeykoon\textsuperscript{b}, R M D B Ranatunga\textsuperscript{c}

Abstract

Introduction: Müllerian anomalies are rare developmental anomalies of the female reproductive tract, that are usually associated with renal abnormalities.

Case Presentation: A 16-year-old Sri Lankan schoolgirl, presented with progressive dysmenorrhea and inter-menstrual lower abdominal pain for 2 years. There was recent lower abdominal distention over 6 months. Examination revealed a non-tender mass in the suprapubic area.

A subsequent CT scan, followed by an ultrasound abdomen, revealed features of a right-sided unicornuate uterus with a non-communicating left-sided horn containing endometrium and a homogenous large cystic lesion between the non-communicating left horn and the uterine body, associated with ipsilateral renal agenesis.

She underwent a laparotomy and resection of the left-side rudimentary horn of the uterus with a left-side ovarian cystectomy, as intra-operative findings suggested an endometrioma.

Conclusion: Clinical presentations of Müllerian anomalies are largely non-specific. Early diagnosis and prompt treatment are useful in avoiding complications that arise as a consequence of Müllerian anomalies.

Keywords: Müllerian anomalies, ovarian endometriosis, renal agenesis, unicornuate uterus

Introduction

Müllerian anomalies are rare developmental anomalies of the female reproductive tract. Uterine anomalies have been classified according to the American Society for Reproductive Medicine, which divides uterine malformations into nine main groups: Müllerian agenesis, cervical agenesis, unicornuate uterus, uterus didelphys, bicornuate uterus, septate uterus, longitudinal vaginal septum, transverse vaginal septum, and complex anomalies. Unicornuate uterus develops as a result of unilateral failure of the normal Müllerian system development. In most instances, the abnormal Müllerian duct has partially developed into a rudimentary uterine horn\textsuperscript{1}.


DOI: https://doi.org/10.4038/sljog.v46i2.8151

\textsuperscript{a} Undergraduate, Faculty of Medicine, University of Peradeniya, Sri Lanka
\textsuperscript{b} Consultant in Obstetrics and Gynaecology, National Hospital Kandy, Sri Lanka
\textsuperscript{c} Senior Registrar in Obstetrics and Gynaecology, National Hospital Kandy, Sri Lanka

Correspondence: HDM, e-mail: hesharamanamendra99@gmail.com

https://orcid.org/0009-0003-9399-7486

Received 3\textsuperscript{rd} March 2024
Accepted 13\textsuperscript{th} June 2024

This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License, which permits unrestricted use, distribution and reproduction in any medium provided the original author and source are credited.
In this article, we describe a rare case of unicornuate uterus with a functional non-communicating rudimentary horn, associated with ovarian endometriosis and ipsilateral renal agenesis to improve our understanding of this rare Müllerian anomaly, along with the proper diagnosis methods and treatment options.

**Case presentation**

A 16-year-old Sri Lankan schoolgirl, presented with progressive dysmenorrhea and inter-menstrual lower abdominal pain that developed after one year of menarche, affecting her quality of life. She attained menarche at the age of 13 and had regular periods. There is no heavy menstrual bleeding, intermenstrual bleeding, vaginal discharge, or urinary symptoms. Lower abdominal distention developed gradually over the last 6 months with mild discomfort. Past medical, surgical, and family histories were not significant.

On examination, there was a visible mass in the suprapubic area. It was a rounded, firm mass of around 10 × 9 cm with regular margins and a smooth surface, which was pelvic in origin and extended into the abdominal cavity.

A transabdominal ultrasound scan suggested a right-side ovarian cyst measuring 13×15 cm. Subsequent CT scan revealed features of a right-sided unicornuate uterus with a non-communicating left-sided horn containing endometrium categorized in Class II Müllerian duct anomaly, type alb (American Fertility society classification), a distended left endometrial cavity with high-density fluid collection within (5×3.5×3.5 cm), and a homogenous large cystic lesion between the non-communicating left horn and the uterine body, which very much represent endometriotic hematometra secondary to repetitive shedding of the rudimentary horn in size of 9 (A)×13 (Trans) ×15 (CC) cm. Left-side renal agenesis was identified with a hypertrophied right kidney and BOSNIAK type 1 renal cyst in the interpolar region of the right kidney.

She underwent a laparotomy and resection of the left-side rudimentary horn of the uterus with a left-side ovarian cystectomy. Intra-operative findings were a large homogeneous cyst, which was suggestive of endometrioma of the left side ovary of 15×10×8 cm in size, and a functional left side rudimentary horn, which was 5×8×6 cm in size with hematometra. The left-side fallopian tube was not visible. The right-side functional uterine horn was normal in size, and the right-side fallopian tube and right-side ovary appeared to be normal as well. An excised sample of the rudimentary horn and ovary was sent for histopathological diagnosis. Histology report findings were an endometriotic cyst in the left ovary and the rudimentary horn with late secretory phase endometrium. We plan to follow up on her until 10-15 years from now, to assess her pregnancy outcomes as well.

![Figure 1. Endometrioma of the left ovary (A) Altered blood filled ovary (B) After the evacuation of filled blood.](image-url)
Case report

Discussion

The overall prevalence of Müllerian anomalies was 9.8% of the general population, with a 1.6% prevalence of unicornuate uteri. Müllerian ducts (Paramesonephric ducts), whose development is preceded by Wolffian (mesonephric) ducts within the paired urogenital ridges, provide critical paracrine growth factors for Müllerian duct growth. Therefore, Müllerian anomalies are usually associated with urinary tract anomalies. Statistically, 40.5% of the patients with a unicornuate uterus presented with urinary tract anomalies such as renal agenesis, ectopic kidney, horseshoe kidney, double renal pelvis, and/or unilateral medullary sponge kidney. In our case, the patient had a unicornuate uterus with congenital renal agenesis on the side of the rudimentary horn, along with ovarian endometriosis.

The incidence of a unicornuate uterus with endometriosis on the side of the rudimentary horn is 20-40%. The pathogenesis of endometriosis remains controversial. According to the theory of retrograde menstruation, endometrial cells and tissues derived from menstruation implanted retrogradely into the abdominal cavity invade and induce a local inflammatory response that is accompanied by angiogenesis, adhesion, fibrosis, scarring, and anatomical distortion, which is the most applicable theory in our patient as she had collected blood in the functional rudimentary cavity of the uterus, which probably led to the development of endometriosis of the left ovary.

A similar case study, which was regarding an 18-year-old girl who presented with progressive dysmenorrhea and was diagnosed to have a unicornuate uterus with a left-sided cavitary rudimentary horn with left hematometra and hematosalpinx and left renal agenesis, mentioned that she underwent a successful laparotomy and excision of the left-sided cornu along with left salpingo-oophorectomy. Another study of a 15-year-old girl with progressive dysmenorrhea, who was diagnosed to have a left unicornuate uterus with a non-communicating right cavitary horn and hematometra, had laparoscopic removal of the right fallopian tube and rudimentary horn. In both cases, sonographic examination was the first-line imaging modality used, followed by MRI in diagnosing.

MRI has proved effective in preoperative evaluation of Müllerian duct anomalies. Combined hysteroscopy and laparoscopy is considered to be the gold standard in diagnosis. Our patient underwent a CT scan before surgery due to the unavailability of an MRI.

It has been proven that a unicornuate uterus is associated with an increased risk of adverse pregnancy outcomes, specifically functional rudimentary horns, which are at particularly high risk of both pelvic endometriosis and rudimentary-horn pregnancy. Therefore, surgical excision of the rudimentary horn was done to prevent potentially serious complications in pregnancy, and it is recommended to reduce the risk of recurrent or de novo endometriosis. Removal of the left ovary, which consists of endometriosis, was necessary for our patient to alleviate dysmenorrhea. Laparoscopy proves to be an effective surgical approach for the removal of the cavitated, non-communicating rudimentary horn in patients with a unicornuate uterus. If facilities are available, laparoscopic excision should be offered, especially to young girls, as it is less morbid and cosmetically more acceptable. Our patient underwent a CT scan before surgery, which has poorer soft tissue resolution than an MRI and does not give adequate information about the condition to proceed with laparoscopic surgery. Furthermore, the lack of infrastructure led us to proceed with laparotomy and resection of the rudimentary horn and the ovarian cyst.
Case report

Conclusion
The clinical presentations of rudimentary uterine horn with ovarian endometriosis, including dysmenorrhea and non-menstrual pelvic pain, are largely non-specific. Therefore, when evaluating dysmenorrhea, especially in adolescents, it is important to consider the rare possibility of mullerian anomalies. Early diagnosis and prompt treatment are useful to avoid future gynecological and obstetrical complications. Laparoscopic surgery is the preferred mode of management if the facilities are available.

Conflicts of interest
The authors declare that they have no conflicts of interest.

References