Complex mullerian duct anomaly in a young female with primary amenorrhoea, infertility, and chronic pelvic pain

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Received: 23.06.12
Review completed: 25.07.12
Accepted: 14.09.12

Access this article online
Quick Response Code:
Website: www.jhrsonline.org
DOI: 10.4103/0974-1208.106345

ABSTRACT

Mullerian duct anomalies, though rare, can be a treatable cause of pelvic pain and infertility. Various complex Mullerian duct anomalies may exist with combination of features of more than one class. Since there are no precise clinical or imaging criteria to enable specific categorisation, there is ambiguous classification of these anomalies by various radiologists and clinicians. A young female presented with complaints of chronic pelvic pain, primary amenorrhoea and infertility. The patient was evaluated by sonography and Magnetic Resonance Imaging and diagnosed as case of complex mullerian duct anomaly, a unicornuate uterus with cervical dysgenesis and cavitated, noncommunicating, rudimentary right horn. The findings were confirmed on laprohysteroscopy and the patient underwent hystertectomy. There should be an integrated clinico-radiological classification scheme and familiarity with rare and complex anomalies for appropriate diagnosis and management of complex Mullerian duct anomalies.

KEY WORDS: Cervical dysgenesis, magnetic resonance imaging, mullerian duct anomaly, rudimentary horn, unicornuate

INTRODUCTION

Mullerian duct anomalies (MDA), though rare, can be a treatable cause of pelvic pain and infertility. The spectrum of mullerian duct anomalies is a continuum rather than distinct entities, and some complex anomalies have features of more than one class. Since there are no precise clinical or imaging criteria to enable specific categorisation of such anomalies, there is ambiguous classification of these anomalies by various radiologists and clinicians. We report a case of a rare complex MDA, diagnosed as unicornuate uterus with cervical dysgenesis and cavitated, noncommunicating rudimentary horn. There has been no similar case reported in the literature till date, adding to the spectrum of complex anomalies.

CASE REPORT

A nulliparous, 30-year-old female, presented to the infertility clinic with complaints of primary amenorrhoea and infertility, with chronic pelvic pain for more than ten years duration. The general physical examination was unremarkable with normal secondary sexual characters. The routine laboratory investigations for amenorrhoea and infertility were normal including normal hormonal profile. Pelvic examination revealed a normally developed vagina and cervix and anteverted nulliparous uterus. The patient had prior been investigated several times, including sonography of pelvis, outside our institution, with all inconclusive reports.

The patient underwent ultrasound in the radiology department of our institute, which demonstrated the presence of a normal sized anteverted uterus, oriented more toward the left side of pelvic cavity. The endometrial cavity was distended with echogenic fluid. The left uterine tube was dilated and filled with fluid of similar echogenecity. The cervix and left ovary were normal. A complex mass comprising of solid and cystic components was found on the right side. There was no separate visualization of right ovary. The patient was advised pelvic magnetic resonance imaging (MRI) to better characterise the ultrasound findings.

MRI of the pelvis showed presence of two asymmetric uterine horns, smaller on the right side with thin myometrium and normal sized left with normal myometrium. Both endometrial cavities were distended with hemorrhagic contents and showed no communication to each other. There was associated hematosalpinx on either side. The cervix and vagina were normally developed, however there was no demonstrable communication between the endometrial cavity and the endocervical canal. Both the ovaries were normal. On the basis of MRI findings, a diagnosis of unicornuate uterus with cervical dysgenesis and a cavitated, noncommunicating rudimentary horn on the right side, was made [Figure 1].

The patient was taken up for laprohysteroscopy. The cervix was well developed but the hysteroscope could not be negotiated beyond the isthmus, consistent with cervical dysgenesis. There were two uterine horns with the right side smaller than the left. Dense adhesions were found in the pelvic cavity, likely due to retrograde menstruation and Stage IV endometriosis. The omentum was adherent to the uterus and the pouch of Douglas was obliterated. The fallopian tubes on both sides were dilated with thick, oedematous walls and were found congested. Bilateral ovaries were normal.

Due to delayed presentation of the patient, advanced endometriosis, dense pelvic adhesions and a failed attempt to recanalize/reconstruct cervix, a decision for hysterectomy was taken after obtaining patient's consent [Figure 2]. The uterus and both fallopian tubes were removed and adhesiolysis was done. There were no intra or postoperative complications. On a one year follow up, the patient is completely relieved of her symptoms of chronic pelvic pain.

DISCUSSION

Mullerian duct anomalies (MDA), though rare, may present in different ways from infancy to young adulthood, with mucocolpos, hematocolpos, hematometra, primary amenorrhea, pelvic pain, infertility, or repeated pregnancy loss.

An understanding of the differences between these uterovaginal anomalies is crucial in understanding the respective clinical manifestations, different treatment regimens and prognosis. Many classifications of uterine anomalies exist including the Buttram and Gibbons[1] and the American Fertility Society (AFS) classification.[2] Though these classifications are important in the treatment of infertility and symptoms arising from obstruction or deformity, it is important to realise that these classification

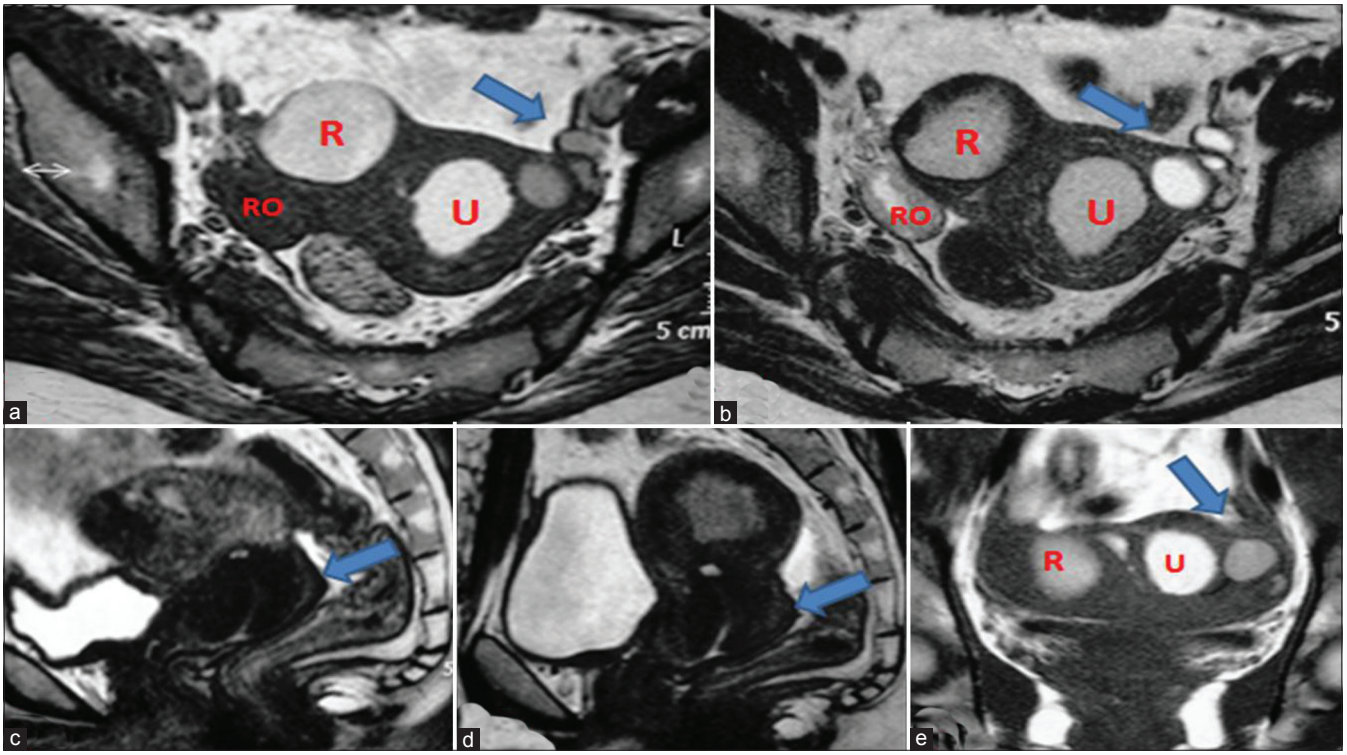

Figure 1: Magnetic resonance imaging of pelvis. Axial T1W (a) and T2W (b) Images showing unicornuate uterus (U) with rudimentary right horn (R). Both cavities are filled with blood (hyperintense on T1W and T2W images). There is left hematosalpinx with dilated, blood filled fallopian tube (arrow). Normal right ovary (RO) seen. Sagittal T2W images (c, d) Showing well formed cervix (arrow) with no communication between endometrial cavity and endocervical canal. Coronal T1W image (e) Showing hematometra in unicornuate uterus (U) and rudimentary horn (R) with left hematosalpinx (arrow)

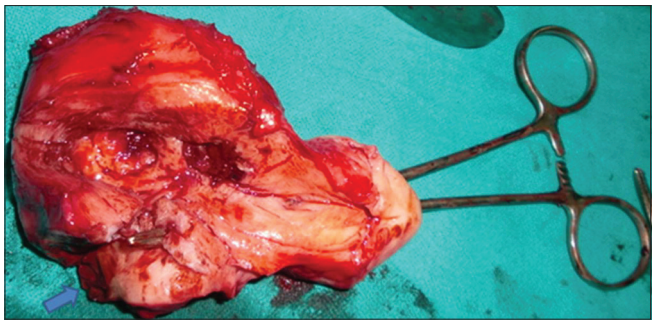

Figure 2: Gross hysterectomy specimen showing unicornuate uterus with rudimentary right horn (arrow). The forceps could not be advanced beyond the cervix consistent with obstruction at isthmus

systems serve merely as a framework and not all anomalies will fit completely into one of these categories. It is more important to accurately describe the different components of the complex anomalies so that appropriate management can be planned.

The modified AFS classification by Rock and Adam[3] embraces a broader collection of uterine and vaginal anomalies without conflicting observations or over simplicity encountered in other classifications. This classification correlates anatomic anomalies with embryologic arrests, classifying uterovaginal anomalies as dysgenesis disorders or vertical or lateral fusion defects. Anomalies are further subcategorized into obstructive or nonobstructive forms, since their treatment differs. Class IV of this classification is a useful addition, embracing any possible unusual configurations or combination of defects, since genital tract aberrations do not necessarily follow any defined and consistent pattern.

A unicornuate uterus with a rudimentary horn is the most uncommon uterine anomaly, representing approximately 20% of all MDA. A cavitated, non-communicating rudimentary horn in a unicornuate uterus represents approximately 4.4% of all MDA.[4] The treatment of this anomaly is surgical resection of the rudimentary horn.

Congenital cervical anomalies, including agenesis and dysgenesis are even rarer, with less than 200 cases of cervical agenesis being reported till date.[5] Many authors have recommended hysterectomy as an initial procedure for a patient with cervical dysgenesis/agenesis with a functioning uterine corpus. This eliminates needless suffering from associated problems of endometriosis, sepsis and multiple surgeries. Considering the small potential for pregnancy, some authors advocate procedures of recanalisation or reconstruction of cervix, especially in cases of cervical dysgenesis.[5,6]

Imaging plays an important role in detection and classification of MDA, thereby guiding appropriate management. Although many of these anomalies may be initially diagnosed at hysterosalpingography or sonography, further imaging by MRI is often required for definite diagnosis and elaboration of secondary findings. MRI has a reported accuracy of up to 100% in evaluating MDA.[7] MR imaging provides clear delineation of internal and external uterine anatomy. Complex anomalies and secondary findings of endometriosis along with renal anomalies are optimally characterised noninvasively.

The present case is unique, comprising of combination of two rare anomalies, a unicornuate uterus with cervical dysgenesis and a cavitated noncommunicating rudimentary horn. This anomaly can be assigned to class IV of modified AFS classification by Rock and Adam. The review of literature did not reveal a similar case, though many authors have emphasized the possibility of complex anomalies in a patient with features of more than one class. In the present case, the diagnosis was delayed due to complex nature of the anomaly and absence of an integrated clinico-radiological classification scheme for specific diagnosis of such anomalies.

To conclude, various complex MDA may exist with combined features of more than one class. An integrated clinical and radiological classification and familiarity with rare and complex MDA is essential. MRI remains the modality of choice for preoperative diagnosis. Early surgery offered to the patient may reduce patients suffering, help restore a patent outflow tract and may preserve fertility in some cases.

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How to cite this article: Kumar S, Satija B, Wadhwa L. Complex mullerian duct anomaly in a young female with primary amenorrhoea, infertility, and chronic pelvic pain. J Hum Reprod Sci 2012;5:295-7.
Source of Support: Nil, **Conflict of Interest:** None declared.