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Case Report

Unicornuate uterus with an obstructed rudimentary horn: A report of two cases with an imaging perspective

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Introduction

Congenital anomalies of the uterus and vagina (caused by alterations in development or fusion of the müllerian ducts) are associated with a high incidence of infertility and multiple obstetric problems. MR evaluation allows accurate morphologic demonstration and classification of these anomalies and helps in planning an accurate treatment and also permits assessment of obstructive anomalies and associated pelvic and renal anomalies.¹

Cases

Case 1

A 17-Yr old unmarried girl who achieved her menarche at the age of 12 years with initial cycles of 50–60 days and menstrual

bleeding lasting 1–2 days presented with history of severe postmenstrual abdominal pain. The initial sonography of the pelvis revealed a fluid containing lesion abutting and deviating the uterine body and the fundus to the left. The patient underwent a diagnostic laparoscopy in the civil and the diagnosis of a bicornuate uterus was made with a normal left horn and hematometra in the right horn. No HSG or transvaginal scan study was undertaken as the orthodox parents declined the procedure. A detailed trans-abdominal USG was done and revealed a 4 cm sized fluid-containing structure lying behind the urinary bladder and adjacent and on the right of a normal uterine body and fundus which appeared to be continuous with one cervix. Both the ovaries were identified separately and a provisional diagnosis of a congenital anomaly of the uterus (unicornuate) with an obstruction and hematometra of the rudimentary horn was given (Fig. 1). A detailed MR scan for the patient was performed on a 1.5 T MRI equipment. The MR study (Fig. 2(a)–(c)) revealed continuity of the uterus with one cervix. A globular structure measuring $4.5 \times 5 \times 5.4$ cm was seen lying to the right side of the uterus. The MR morphology of this structure was suggestive of uterine in nature with a preserved zonal anatomy. It had an internal content which was hyperintense on T2WI, T1WI and on fat suppressed (FS) images. There was no continuity with the uterine lumen. Both the ovaries showed multiple small subcentimeter-sized cysts that were hyperintense on T2WI and hypointense on T1WI. A diagnosis of congenital anomaly

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Fig. 1 – (a) USG pelvis image shows an obstructed, dilated, fluid/blood filled rudimentary uterine horn (short arrow) with a developed unicornuate uterus (long arrow) lying adjacent to it. (b) Better sonographic demonstration of the unicornuate uterine horn (long arrow) in the same patient.

of the uterus – unicornuate uterus with an obstructed functioning rudimentary horn with a hematometra in the obstructed element was made.

Case 2

A 36-Yr old lady who has been treated for primary infertility presented for MR imaging during her workup. She attained menarche at the age of 12 years and had initial cycles of 20–25 days with the menstrual bleeding lasting 3 days. There was a history of abdominal pain which was severe in the post-menstrual period. There was an associated lump that was observed by the patient after her marriage at the age of 18 years. She underwent extensive treatment that included pelvic surgery the details of which are not available. Presently during the course of her management of the primary infertility she underwent imaging evaluation for the uterus and pelvis. The HSG (Fig. 3) study revealed a unicornuate uterus with a partly visualized tube with no peritoneal spill. The USG scan of the pelvis including the uterus (Fig. 4) revealed fluid containing structure lying behind the urinary bladder adjacent to a normal uterine body and fundus which appeared to be continuous with one cervix. A provisional diagnosis of a congenital anomaly of the uterus (unicornuate) with an obstruction and hematometra of the rudimentary horn was given. In addition a fluid collection was seen in the pelvis adjacent to the uterus. The MRI scan (Fig 5(a)–(c)) revealed one cervix leading to a uterus which is seen to be adjacent to a rounded structure with walls resembling uterine in morphology with internal collection – hematometra (hyperintense on T1WI T2WI and T1WI FS). This was surrounded by a fluid collection. A diagnosis of congenital anomaly of the uterus – unicornuate uterus with an obstructed functioning rudimentary horn with a hematometra in the obstructed element was made.

Discussion

Congenital uterine anomalies or müllerian duct anomalies are an uncommon but often treatable cause of infertility amongst women. They are estimated to occur in 0.1–0.5% of women, while the prevalence of these anomalies in females with a history of multiple pregnancy loss is about 3%. 25% of women with müllerian duct anomalies have reproductive problems like higher incidence of infertility, frequent first-

Fig. 2 – MR images show an obstructed dilated blood filled rudimentary horn of a bicornuate uterus (short arrow) lying adjacent to its normally developed horn (long arrow). These MR findings are demonstrated well in (a) coronal (b) sagittal and (c) axial plane.

Fig. 3 – HSG study shows a contrast outlined horn of the unicornuate uterus which is located on the right side of the pelvis with an indentation (by the dilated obstructed rudimentary horn of the uterus not appreciated in this contrast study) on its left side.

trimester spontaneous abortions, fetal malposition, preterm labor, etc. The role of imaging is to help diagnose and identify the anomalies amenable to surgical correction and the surgical approach may be altered based on the imaging findings.^{1–3}

Increased morbidity is seen in some types of anomalies in patients with obstructed or partially obstructed müllerian systems who can present with hematosalpinx, hematocolpos and hematometra. An association exists between müllerian duct anomalies and renal anomalies also.⁴

The firstline weapon in the present era is sonographic examination (abdominopelvic and trans-vaginal imaging if possible). Müllerian duct anomalies cannot be excluded on the basis of negative US findings. Newer 3-dimensional (3D) sonographic techniques can offer an added advantage. However it is operator dependent and image resolution can be a limiting factor. Hysterosalpingography (HSG), permits visualization of the uterine cavity and tubal patency. Anomalies may be suggested but positive findings can be nonspecific for an accurate diagnosis hence characterization of müllerian anomalies can be difficult. Similar looking entities on HSG can have differing treatment. MRI is considered the modality of choice for imaging uterine anomalies. MRI provides high-resolution images of the uterine body, fundus, cervix and can assess the urinary tract for concomitant anomalies (IVU was used in the past for assessing the urinary tract). The usual contraindications to MRI have to be kept in mind.^{1–4}

Classification of anomalies^{1,5–7}

Many classifications of uterine anomalies exist; for instance, the American Fertility Society (AFS) classification,⁵ the modified AFS classification by Rock and Adam⁶ and the Buttram and Gibbons classification.⁷

Fig. 4 – (a) USG pelvis image shows an obstructed, dilated, fluid filled rudimentary uterine horn (short arrow) of a developed unicornuate uterus (long arrow) lying adjacent to it. (b) Fluid debris level well demonstrated in the rudimentary horn.

Müllerian duct anomalies are categorized most commonly into 7 classes according to the American Fertility Society (AFS). The classification Scheme in an abbreviated form is as follows:

Class I (hypoplasia/agenesis): This class includes uterine/cervical agenesis or hypoplasia.

Class II (unicornuate uterus): The unicornuate uterus results complete/almost complete, arrest of development of the müllerian duct. If the arrest is incomplete, a rudimentary horn with/without functioning endometrium is present. If the rudimentary horn is obstructed, it may present as an enlarging pelvic mass. If the contralateral horn is healthy and well developed, a full-term pregnancy can occur.

Class III (didelphys uterus): This anomaly results from complete nonfusion of both müllerian ducts. The individual horns are fully developed and cervixes are inevitably present. **Class IV (bicornuate uterus):** A bicornuate uterus results from partial nonfusion of the müllerian ducts. The central

Fig. 5 – (a) Serial sagittal MRI images show an obstructed dilated blood filled rudimentary horn (short arrow) lying adjacent to a normally developed unicornuate uterus (long arrow). Fluid is seen surrounding the uterus. These MR findings are demonstrated well in (i) sagittal (ii) coronal and (iii) axial plane. (b) Sagittal MRI image shows hyperintense content on T1 weighted image confirming blood content in the obstructed dilated rudimentary horn.

myometrium may extend to the level of the internal os (bicornuate unicollis) or external os (bicornuate bicollis).

Class V (septate uterus): A septate uterus results from failure of resorption of the septum between the 2 uterine horns (septum can be partial or complete). Differentiation between a septate and a bicornuate uterus is important because the management differs.

Class VI (arcuate uterus): An arcuate uterus has a single uterine cavity with a convex or flat uterine fundus, the endometrial cavity, which demonstrates a small fundal cleft or impression with a flat outer.

Class VII (diethylstilbestrol-related anomaly): The uterine anomaly is seen in the female offspring of as many as 15% of women exposed to DES during pregnancy. Female fetuses who are affected have a variety of abnormal findings that include uterine hypoplasia and a T-shaped uterine cavity.

The modified AFS classification by Rock and Adam: This classification correlates anatomic anomalies with embryologic arrests. Thus, uterovaginal anomalies are categorized as dysgenesis disorders or vertical or lateral fusion defects. There is a further subdivision into obstructive or non-obstructive forms. Immediate treatment is not needed for nonobstructive forms, but obstructive uterovaginal anomalies require immediate treatment because of retrograde flow of trapped fluids with increasing pressure on surrounding organs.

Class 1—Dysgenesis of müllerian ducts. This class includes agenesis or hypoplasia of the müllerian duct derivatives (the uterus and upper two-thirds of the vagina).

Class 2—Disorders of vertical fusion. There is failure of fusion of the müllerian system with the sinovaginal bulb (cervical dysgenesis and obstructive and nonobstructive transverse vaginal septa).

Class 3—Disorders of lateral fusion. This class of anomalies comprises of a duplicated or partially duplicated reproductive tract. The disorders are due to impaired fusion and/or septal resorption of fusing müllerian ducts attempting to form the uterus, cervix, and upper vagina. It includes anomalies due to failure of fusion of the paired müllerian ducts (as in didelphic and bicornuate uteri) and failure of midline septum resorption after fusion (as in septate uterus). Disorders due to lateral fusion defects are further subclassified into (a) the symmetric nonobstructive form (unicornuate, bicornuate, didelphic, septate, and DES-related uteri) (b) the asymmetric obstructive form (unicornuate uterus with obstructed horn, double uterus with unilaterally obstructed horn, and double uterus with unilaterally obstructed vagina).

Class 4—Unusual configurations and combinations of defects.

Both our cases fall into Class II of the American Fertility Society (AFS) Classification Scheme or Class 3 of the modified AFS classification by Rock and Adam. Both these cases have showed evidence of unicornuate uteri with an obstructed rudimentary contralateral horn. One case was unmarried and presented with severe abdominal pain warranting surgery. She was not yet married to consider infertility as a problem. The second case presented with abdominal pain related to the menses, later on with an appreciable pelvic lump and associated infertility.

It is important to understand that it may not be possible to fit all anomalies into categories. Hence it is then imperative to be able to describe the components of the anomaly rather than to attempt a fit. Class 4 of the modification of the AFS classification by Rock and Adam thus assumes significance.

Conflicts of interest

All authors have none to declare.

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Book Review

Oxford Textbook of Public Health, R. Detels, R. Beaglehole, M.A. Lansang, M. Gulliford, 5th ed. Oxford University Press, Great Clarendon Street, Oxford OX26DP (2011). 1769 pp., Soft Bound, Paperback, ISBN 978-0-19-969347-4.

This edition of the famous Textbook captures the creative challenges for Community Medicine specialists in the current millennium. A wide galaxy of public health luminaries has contributed to the rich resource material provided in this concise textbook. New chapters on contemporary topics like management of public health disasters, application of genomics, gene environmental interactions and clinical epidemiology have greatly enhanced the scope of this evergreen

publication. The book is a “must read” for all postgraduate residents of community medicine and those interested in enhancing their skills in research methodology.

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