Case 16271

Eurorad

Role of MRI in Bilateral rudimentary uterine horns with endometriosis – a rarest of rare anomaly explained by latest ESHRE/ESGE classification

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Section: Genital (female) imaging
Area of Interest: Genital / Reproductive system female
Procedure: Diagnostic procedure
Imaging Technique: MR
Special Focus: Congenital Case Type: Anatomy and Functional Imaging
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Patient: 22 years, female

Clinical History:

A 22-years-old unmarried female patient presented to our department for evaluation of primary amenorrhoea. She also complained of cyclical pain in the lower abdomen. On examination, secondary sexual characters were normal. A previous ultrasound revealed absence of uterus and ovaries suggestive of mullerian agenesis. Both kidneys were seen normally.

Imaging Findings:

The patient underwent non contrast MRI of the pelvis which revealed absence of definite uterus and cervix in the retro-vesical region in the mid line. Note was made of two small structures of uterine morphology placed laterally near the iliac vessels suggestive of rudimentary uterine horns. Both of them were of similar size with visualisation of the endometrial lining which measured approximately 4-5 mm in thickness on either side. Two dysplastic cervices were seen with a band of tissue seen to unite them, posterior to the urinary bladder at the level of pubic symphysis. The vagina also appeared dysplastic. Both ovaries were visualised. The left ovary was enlarged and polycystic with a small endometriotic cyst within.

Discussion:

Union and resorption of paired müllerian ducts in the embryo gives rise to the uterus, fallopian tubes, cervix and upper two-thirds of vagina. Mullerian Duct Anomalies (MDA) are a complex group of disorders of female genital development which present clinically with primary amenorrhoea, infertility, obstetric complications, and endometriosis. Examination of the kidneys in these patients is extremely important because of the association of MDAs with renal anomalies. [1]

The classification proposed by Buttram and Gibbons in 1979 and modified by American Society for Reproductive Medicine (ASRM) in 1988 has been widely used for designation of the type of genital malformation. [1][2] The lack of categorisation of vaginal anomalies and other anomalies which cannot be classified into one of the categories has been the major limitation of this classification system. The VCUAM (Vagina Cervix Uterus Adnex-associated

Malformation) classification was put forward by Oppelt et al in 2005 which had the potential to classify and describe even complex malformations specifically and precisely. [3] However, the only drawbacks of this classification were that the classification of vaginal anomalies was highly complicated and comments on the fallopian tube were needed too, which was difficult to visualise on MRI.
A new classification was introduced in 2013 by the European Society of Human Reproduction and Embryology (ESHRE) and the European Society for Gynaecological Endoscopy (ESGE) which was mainly based on anatomical features. They categorised the uterine anomalies with gradually increasing anatomical digression and also classified cervical and/or vaginal anomalies independently. The advantage of this classification is that it is reproducible, simplified, precise and helpful in treatment planning. [4]
Our case could not be categorised into any one specific category by the ASRM classification, but could be easily categorised using ESHRE/ESGE classification. To the best of our knowledge, the occurrence of bicornuate unfused rudimentary uterine horns with endometrium and complete cervical and vaginal dysgenesis and associated endometriosis is a rarest of rare case and only one case could be found in literature by Goluda et al [5]. According to the ESHRE/ESGE classification, our case will be described as U5a/C4/V4.
In conclusion, our case in addition to being extremely unique also highlights the superiority of ESHRE/ESGE classification over ASRM in describing and characterising complex female genital tract anomalies.
Written informed patient consent for publication has been obtained.
Differential Diagnosis List: Bicornuate uterus with bilateral rudimentary horns, cervico-vaginal dysgenesis and endometriosis., Unicornuate uterus with contralateral rudimentary horn, Bicornuate uterus

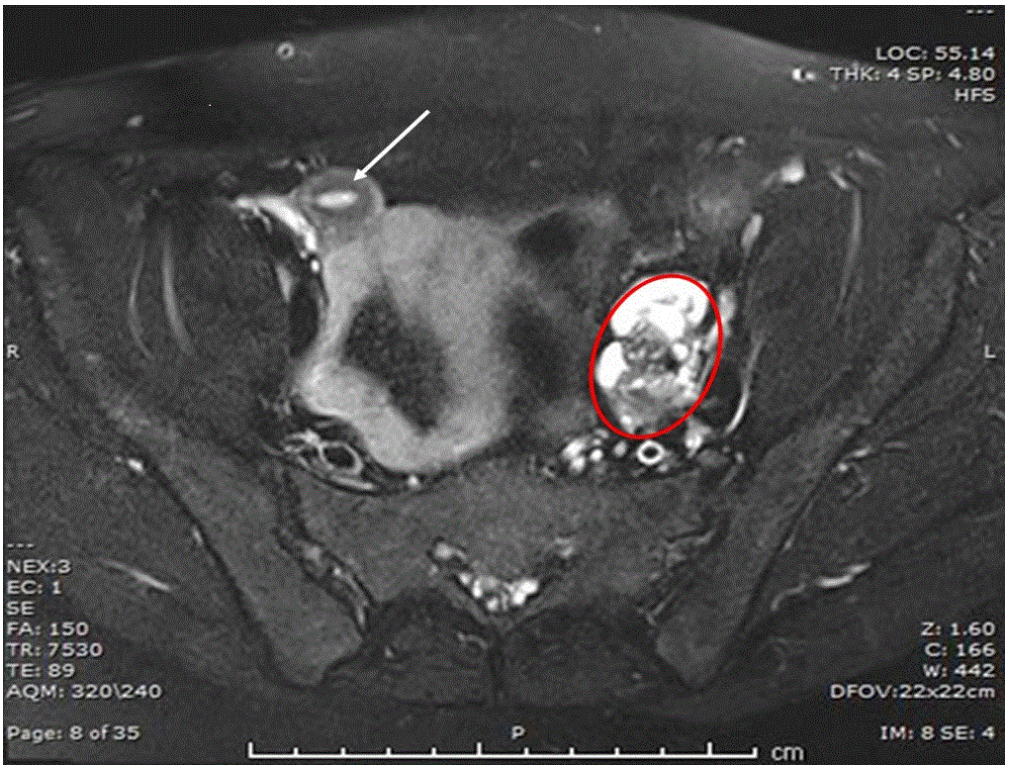
Final Diagnosis: Bicornuate uterus with bilateral rudimentary horns, cervico-vaginal dysgenesis and endometriosis.

References:

Behr SC, Courtier JL, Qayyum A (2012) Imaging of müllerian duct anomalies. Radiographics 32(6):E233-250 (PMID: 23065173)
Buttram VC Jr, Gibbons WE (1979) Müllerian anomalies: a proposed classification. (An analysis of 144 cases.). Fertil Steril 32:40–46 (PMID: 456629)
Oppelt P, Renner SP, Brucker S, Strissel PL, Strick R, Oppelt PG, Doerr HG, Schott GE, Hucke J, Wallwiener D, Beckmann MW (2005) The VCUAM (Vagina Cervix Uterus Adnex–associated Malformation) Classification: a new classification for genital malformations. Fertil Steril 84:1493-1497 (PMID: 16275249)
Grimbizis GF, Gordts S, Di Spiezio Sardo A, Brucker S, De Angelis C, Gergolet M, Li TC, Tanos V, Brölmann H, Gianaroli L, Campo R (2013) The ESHRE/ESGE consensus on the classification of female genital tract congenital anomalies. Hum Reprod 28:2032-2044 (PMID: 23771171)
Goluda M, Gabry? MS, Ujec M, Jedryka M, Goluda C (2006) Bicornuate rudimentary uterine horns with functioning endometrium and complete cervical-vaginal agenesis coexisting with ovarian endometriosis: a case report. Fertil Steril 86:462-e9-11 (PMID: 16806208)

Figure 1

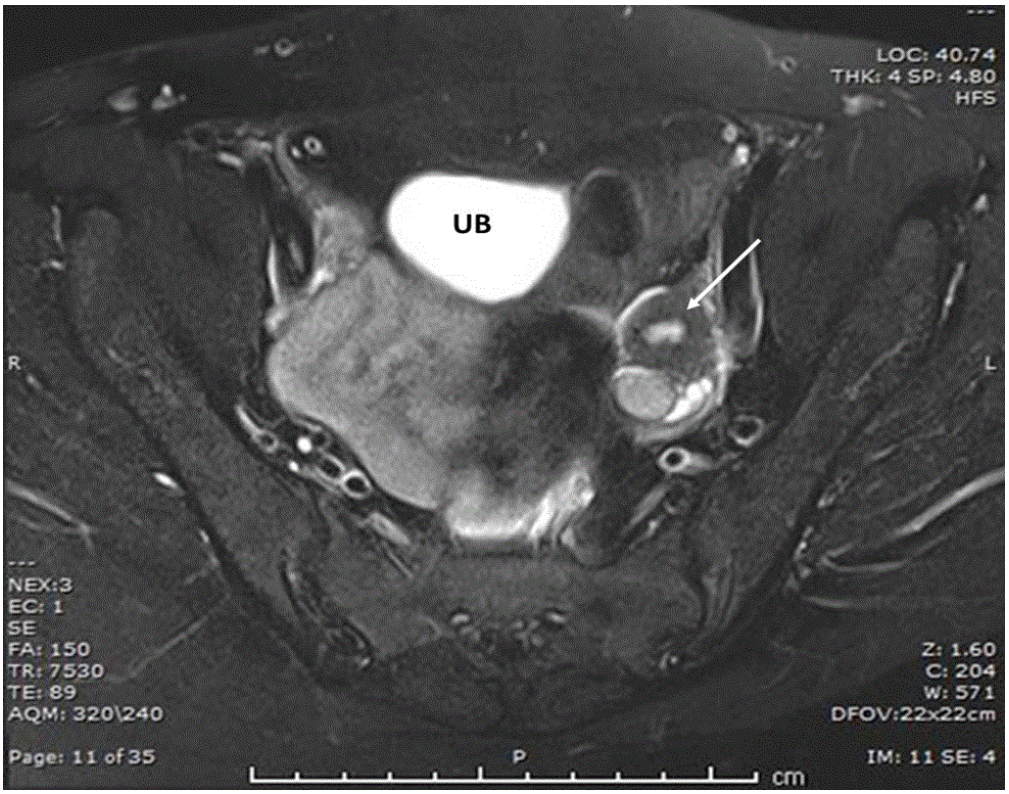
a



Description: Axial T2W fat-sat image of the pelvis showing right uterine horn (white arrow) and bulky and polycystic left ovary (red circle). **Origin:** Department of Radiodiagnosis, TMMC & RC, Teerthanker Mahaveer University, Moradabad- INDIA

Figure 2

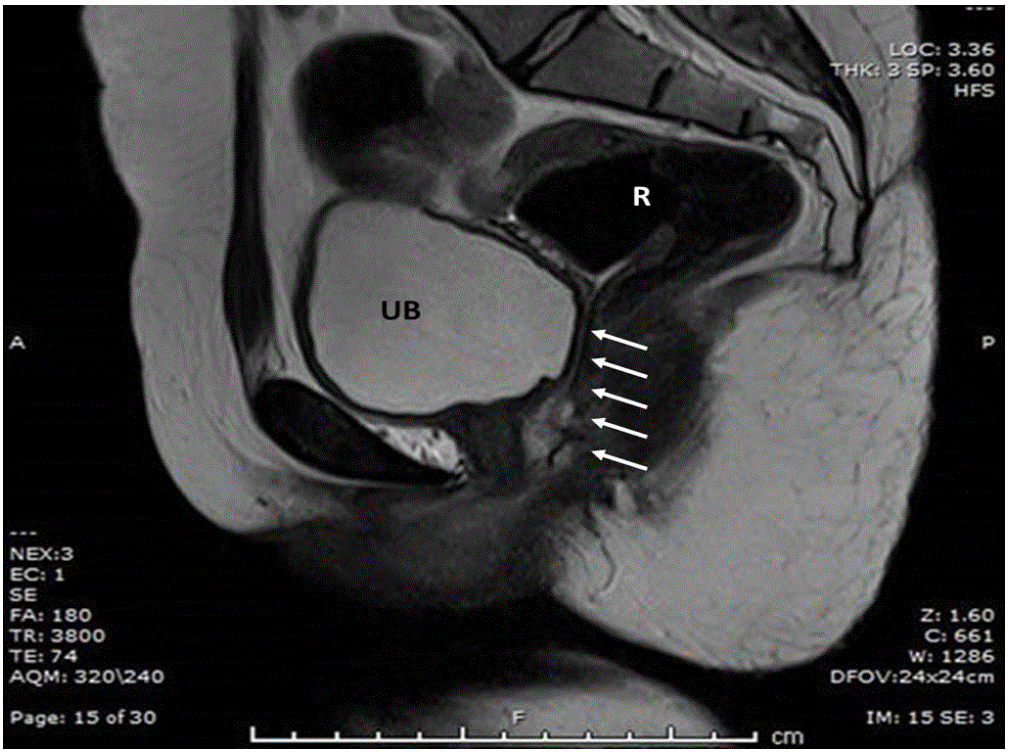
a



Description: Axial T2W fat-sat image of the pelvis showing left uterine horn (white arrow), UB-Urinary Bladder. **Origin:** Department of Radiodiagnosis, TMMC & RC, Teerthanker Mahaveer University, Moradabad- INDIA

Figure 3

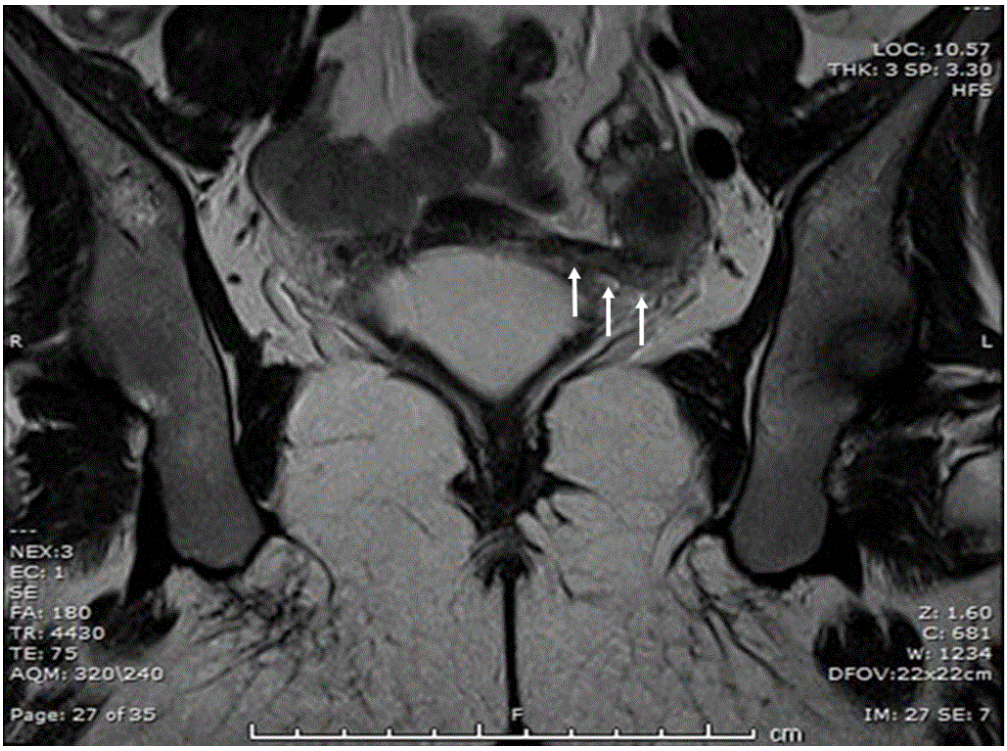
a



Description: Sagittal T2W image of the pelvis showing cervico-vaginal dysgenesis (white arrows), UB-Urinary Bladder, R-Rectum **Origin:** Department of Radiodiagnosis, TMMC & RC, Teerthanker Mahaveer University, Moradabad- INDIA

Figure 4

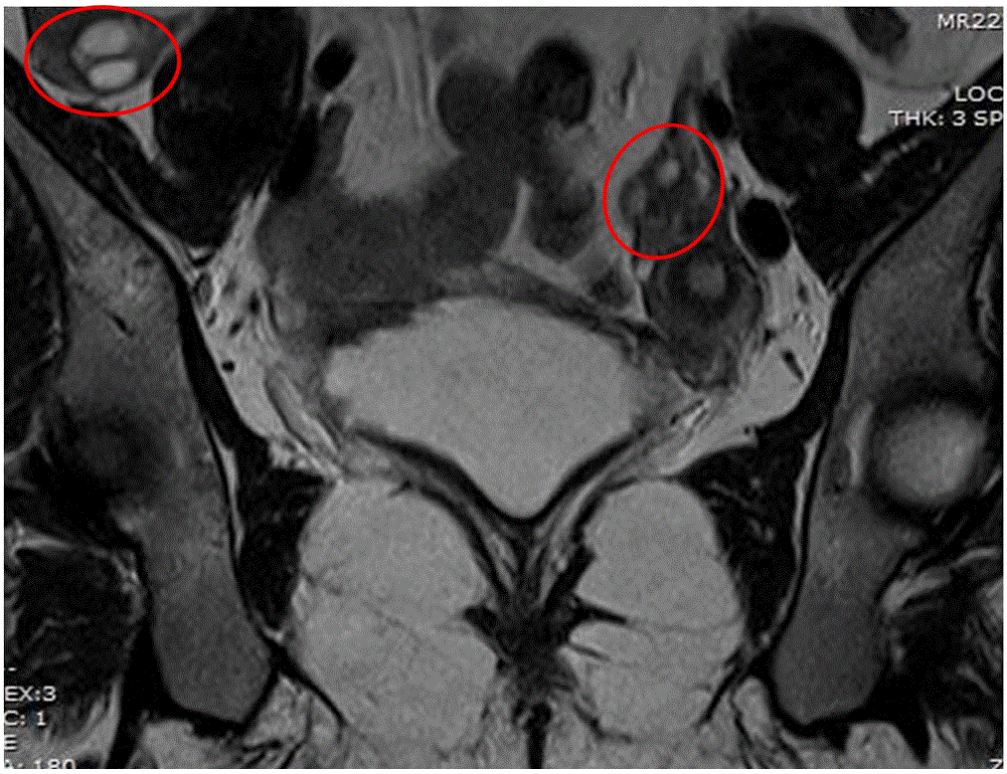
a



Description: Oblique-coronal T2W image of the pelvis showing dysgenetic band of cervical tissue extending from left uterine horn (white arrows). **Origin:** Department of Radiodiagnosis, TMMC & RC, Teerthanker Mahaveer University, Moradabad- INDIA

Figure 5

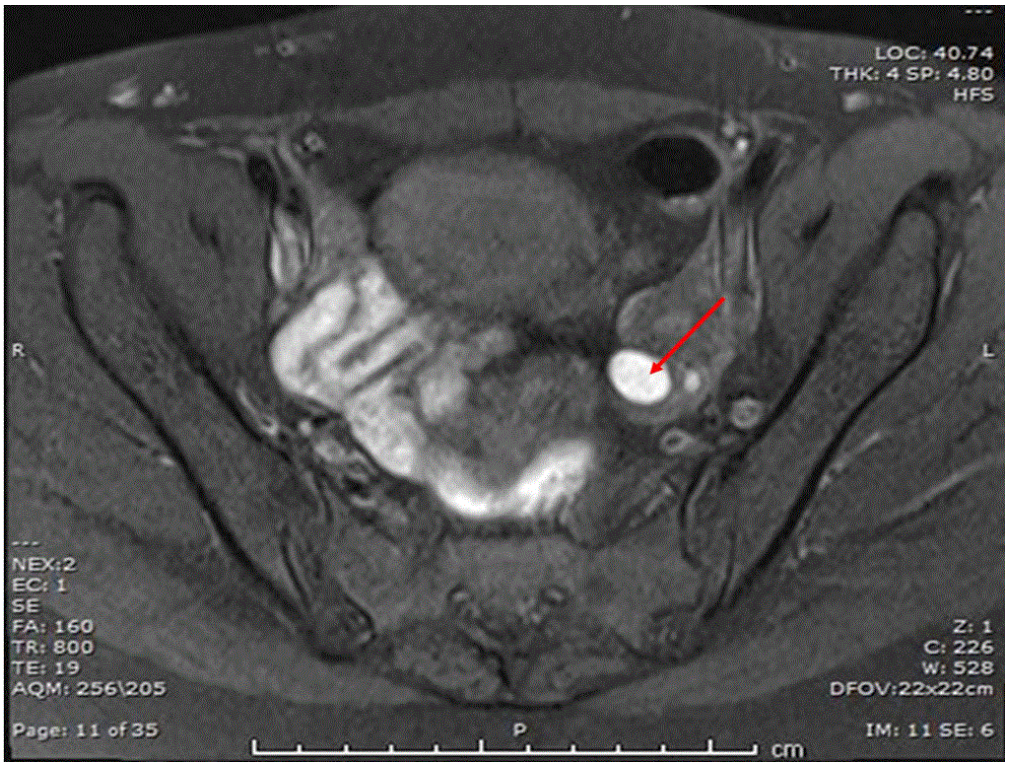
a



Description: Oblique-coronal T2W image of the pelvis showing bilateral ovaries with multiple follicles within (red circles). **Origin:** Department of Radiodiagnosis, TMMC & RC, Teerthanker Mahaveer University, Moradabad- INDIA

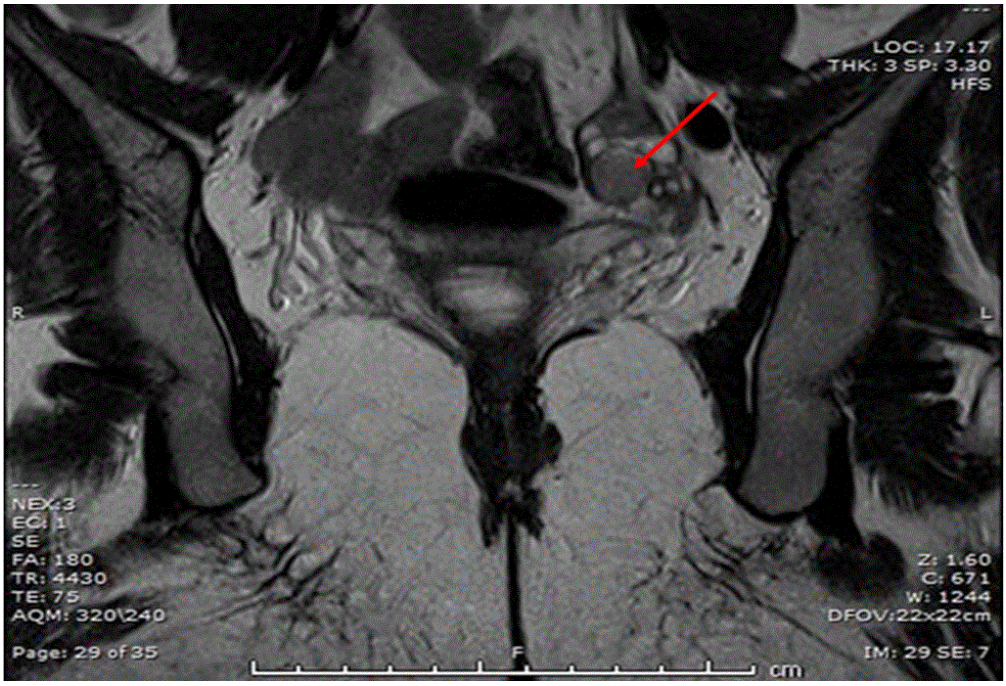
Figure 6

a



Description: Axial T1W fat sat image of the pelvis showing small endometriotic cyst in left ovary (red arrow). **Origin:** Department of Radiodiagnosis, TMMC & RC, Teerthanker Mahaveer University, Moradabad- INDIA

b



Description: Oblique-coronal T2W image showing small endometriotic cyst in left ovary showing T2-shading sign (red arrow). **Origin:** Department of Radiodiagnosis, TMMC & RC, Teerthanker Mahaveer University, Moradabad- INDIA