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Radiodiagnosis

A Rare Case of Bilateral Rudimentary Uterine Horns with Cervico-Vaginal Aplasia- A Case Report

Dr. Manish Bhagat[1], Dr. Shilpi Gupta[2*]

[1]Professor & HOD, Department of Radiodiagnosis, Sri Aurobindo Medical College & PG Institute, Indore, M.P, India
[2]Resident, Department of Radiodiagnosis, Sri Aurobindo Medical College & PG Institute, Indore, M.P, India

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***Corresponding author:** Dr. Shilpi Gupta
Resident, Department of Radiodiagnosis, Sri Aurobindo Medical College & PG Institute, Indore, M.P, India

Abstract **Case Report**

The purpose of this study was to review the embryology, classification, imaging features and treatment options of Müllerian duct anomalies. MDAs are a broad and complex spectrum of abnormalities that are often associated with primary amenorrhea, infertility, obstetric complications, and endometriosis. MDAs are commonly associated with renal and other anomalies; thus, identification of both kidneys is important. However, MDAs are not associated with ovarian anomalies. The role of imaging is to help detect, classify and guide surgical management. At this time, MRI is the modality of choice because of its high accuracy in detecting and accurately characterising Müllerian duct anomalies. In conclusion, radiologists should be familiar with the imaging features of the seven classes of Müllerian duct anomalies, as the appropriate course of treatment relies upon the correct diagnosis and categorisation of each anomaly.

Keyword: Müllerian duct anomalies, cervico-vaginal agenesis, rudimentary uterine horns, primary amenorrhea.

INTRODUCTION

Mullerian duct anomalies (MDA) are rare congenital malformation that affects about 1:4500 female newborns [1, 2]. Its cause is unknown, but the reproductive abnormalities are due to lack of development of the Müllerian ducts between the fifth and the sixth weeks of gestation. This syndrome is characterized by complete or partial vaginal agenesis, tubal, and uterine-cervical abnormalities. In clinical presentation, the disease typically presents itself as primary amenorrhea in an adolescent who has secondary sexual characteristics compatible with their age and may be accompanied by cyclic dysmenorrhea when a rudimentary uterus with functional endometrium is present. Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is the second most common cause of primary amenorrhea, after gonadal dysgenesis [3, 4].

CASE REPORT

A 23 year old married female presented to the Obstetrics and Gynecology OPD with complains of primary amenorrhea & failure to conceive.

During the physical examination, it was observed that the patient had normal secondary sexual characteristics such as breast development and axillary and pubic hair corresponding to her chronological age.

According to the patient's medical history, patient was admitted to the doctor many times and each time oral contraceptive pills have been started or been told that you should wait a little more.

Total blood count and biochemistry laboratory values were within normal limits. Estradiol: 82 pg/mL (20-160) pg/L, FSH: 5.48 mIU/mL (2.8-11.3), LH: 4.69 (1.1-11.6) mIU/mL, prolactin: 21.94 ng/mL (1.9-25) and androgen levels (free testosterone: 1.81 pg/mL (0.06-2.57) and 17-hydroxyprogesterone: 0.3 ng/mL (0.10-1.0); androstenedione: 1.8 ng/mL (0 from 0.21 to 3, 08), DHEA-S: 145 g/dL (65-380)) were normal in hormonal evaluation

External genitalia, urethra and vaginal orifice were normal in gynecological examination. Imperforate hymen could not be ruled out because patient did not accept vaginal examination. Pelvic MRI was advised.

MRI revealed two non-communicating rudimentary uterine horns in both adnexa with non-

visualisation of cervix & upper part of the vagina. Right ovary was bulky with a hemorrhagic cyst. Left ovary was normal. Thus, we concluded bilateral rudimentary uterine horns with cervico-vaginal aplasia.

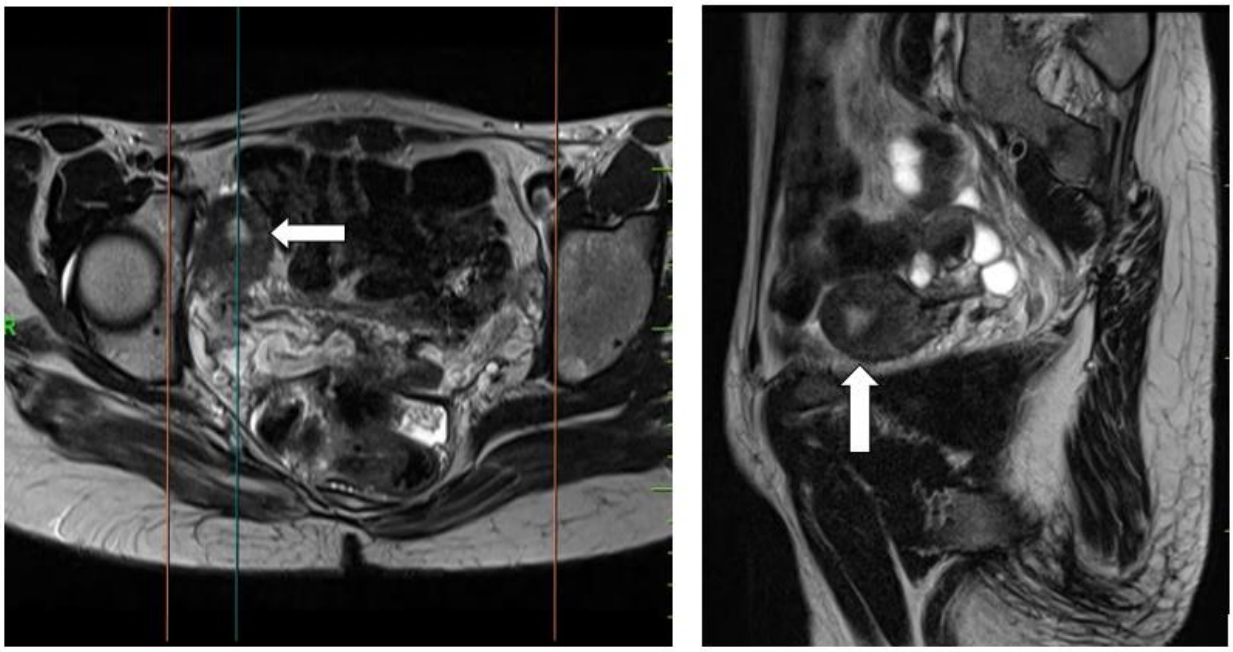

Fig 1: Axial & sagittal T2WI showing right rudimentary horn adjacent to right ovary [11]

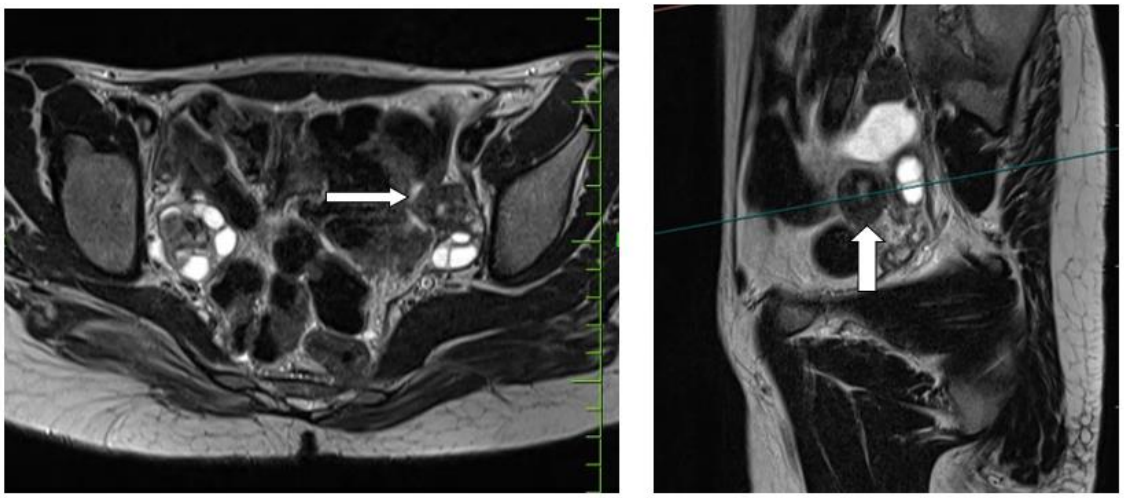

Fig 2: Axial & sagittal T2WI showing left rudimentary horn adjacent to left ovary [12]

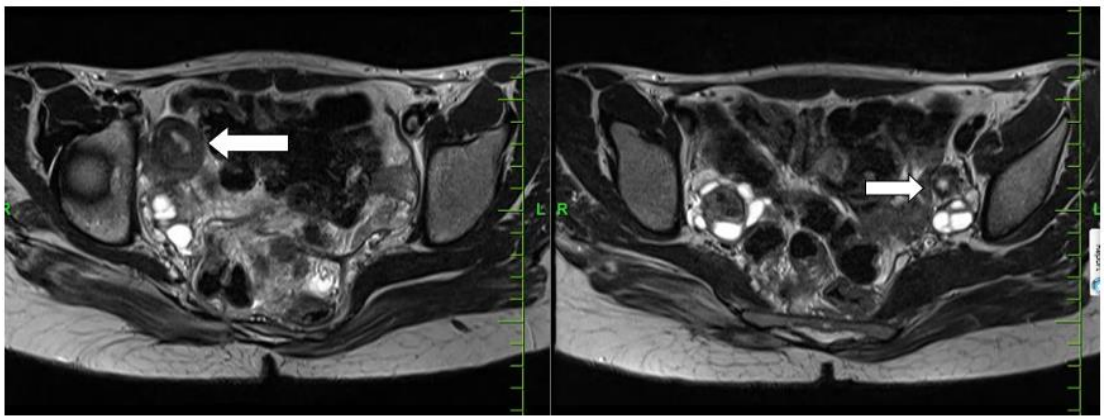

Fig 3: Axial T2WI showing both rudimentary horns [13]

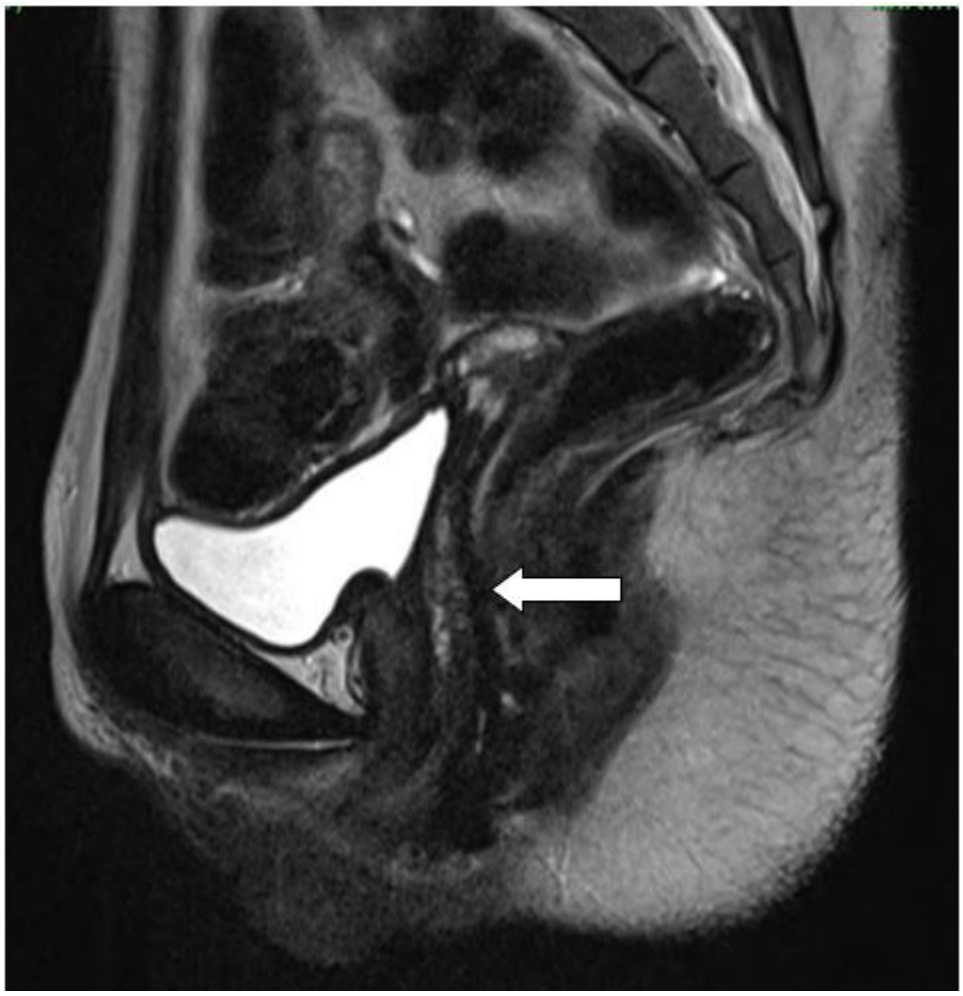

Fig 4: Sagittal T2WI showing hypoplastic blind ended vagina [14]

DISCUSSION

MDA (mullerian duct anomalies) are a fertility obstacle for young adult women. These patients usually manifest psychological consequences – such as anxiety, lower self-esteem, and lower life quality – associated with this diagnoses [5, 6]. As the main outcome of the MDA includes partial or complete vaginal agenesis, uterine and cervical abnormalities – for instance, the cervix aplasia/ hypoplasia – primary amenorrhea is the main reason women go to find medical assistance [9, 10].

MDA with bilateral rudimentary uterine horns and small vagina is an unusual form of Müllerian agenesis. The treatment is based on medical therapy, which includes hormonal and non-hormonal medication, such as analgesics and surgical correction of uterine malformation depending upon the signs and symptoms presented, besides the uterus anatomy and patient's history [7, 8].

On reviewing the literature, we could find very few cases of rudimentary horns with cervico-vaginal agenesis. A similar case has been reported in 2019 where a 28 year-old women had communicating rudimentary uterine horns with cervico-vaginal agenesis.

The anatomical correction of the syndrome is creating a new vaginal canal through surgical or non-surgical procedures in order to allow the patient to perform sexual activities. However, vaginal elongation should wait until the patient has been oriented and feels secure and emotionally stable about the procedure and expresses the desire to proceed. Surgical procedures with removal of the uterine remnants also aim to avoid endometriosis development.

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