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RARE UTERINE ANOMALIES AS A CAUSE OF PERSISTENT CYCLIC PELVIC PAIN IN YOUNG WOMEN-CASE SERIES

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Abstract

Background: Young women have cyclic pelvic pain that does not improve with antispasmodic drugs, it may be caused by a variety of reasons. Also, among the less frequent but significant causes are uterine anomalies that involve having rudimentary or extra cavities. Even though ultrasonography is the first imaging test used for dysmenorrhea, its effectiveness may be reduced when the ultrasound is done through the abdomen. Because MRI offers clearer images of soft tissues and multiple views, it is the technique of choice in complicated cases. Unlike other studies, this one focuses on MRI findings linked to rare congenital uterine abnormalities in females experiencing severe, ongoing dysmenorrhea. The research also gives guidelines for imaging tests to help improve the accuracy of diagnosis.
Case Presentation: We present three cases of women under age 40 without previous pregnancies who had persistent pelvic pain and these cases were diagnosed with rare uterine malformations. The abnormalities found are: (1) a mass that looks like an accessory and is cavitated in the uterus, (2) MRKH syndrome and (3) a non-communicating rudimentary horn on the side of a hemiuterus (unicornuate uterus). In all of the cases, MRI helped to confirm the diagnosis and removing the abnormal structures through surgery greatly improved the patients' symptoms.

Conclusion: If chronic, cyclical pelvic pain cannot be managed with medicines in adolescent and young adult females, Müllerian duct anomalies and functional endometrial tissue should be considered. MRI is very useful in these cases, as it lets doctors precisely identify structures and help decide on the best treatment. Since the symptoms of dysmenorrhea can vary, it is important to use an imaging-based approach to identify any gynecological issues early and provide the right treatment quickly.

Keywords: Magnetic Resonance Imaging, Cyclic Pelvic Pain, Congenital Uterine Anomalies, Accessory and Cavitated Uterine Mass, Mayer–Rokitansky–Küster–Hauser Syndrome, Unicornuate Uterus

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Background

Many young females with persistent pelvic pain and varying degrees of obstruction in their outflow tract may have Müllerian duct anomalies which are often overlooked.

The symptoms could be no menstruation at all or normal cycles, depending on how severe the condition is. MRI is the main tool for assessing these anomalies noninvasively, since it offers a better way to find and recognize their features. The system used for classifying congenital As shown in Table 1, the European Society of Human Reproduction and Endoscopy (ESHRE) and the European Society for Gynecological Endoscopy (ESGE) give information on abnormalities of the female genital tract.

The first test used for young females with menstrual pain is usually transabdominal ultrasound. It is useful for discovering abnormalities of the Müllerian system, yet it does not have the needed detail to clearly define the uterine zones, the precise morphology of the uterus, cervix and vagina or the condition of the fallopian tubes and the connection between the uterine buds and the ovaries. Using T2-weighted MRI, you can clearly see the parts of the uterus and confirm if the uterine cavity has a normal function. Performing MRI along the uterine axis is vital and it may include a 3D T2-weighted pelvic sequence for better visualization of the pelvis. In addition, coronal images that cover a wide area are necessary for checking renal abnormalities which are often connected to Müllerian problems.

Table 1: Uterine Anomalies Classification (ESHRE/ESGE System)

Code	Primary Uterine Class	Subtype Description
U0	Anatomically Normal Uterus	Uterine morphology is within normal anatomical limits.
U1	Dysmorphic Uterus	a) T-shaped uterine cavity b) Infantile uterus c) Other structural variations that reduce endometrial volume
U2	Septate Uterus	a) Partial septation of the uterine cavity b) Complete septation of the endometrial canal
U3	Bicornuate Uterus	a) Partial division of the uterine corpus b) Complete division c) Bicornuate uterus with residual septum

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U4	Hemi-Uterus	a) Presence of a rudimentary uterine horn with or without communication to the main uterine cavity b) Absence of a rudimentary horn
U5	Aplastic Uterus	a) Presence of a rudimentary uterine structure (communicating or non-communicating) b) Complete absence of uterine cavity
U6	Unclassified Müllerian Anomalies	Congenital malformations not encompassed by categories U1 to U5.

Cervical Anomalies Classification

Code	**Cervical Morphological Description**
C0	Cervix is structurally and functionally normal
C1	Presence of a fibrous or muscular septum within the cervical canal (septate cervix)
C2	Duplication of the cervix with two structurally normal cervical canals
C3	Absence of one cervical canal (unilateral cervical aplasia)
C4	Complete congenital absence of the cervix (cervical aplasia)

Vaginal Anomalies Classification

Code	**Vaginal Morphological Description**
V0	Vagina is structurally normal
V1	Presence of a longitudinal, non-obstructing septum within the vagina
V2	Longitudinal vaginal septum causing partial or complete obstruction
V3	Presence of a transverse vaginal septum or an imperforate hymen
V4	Congenital absence of the vaginal canal (vaginal aplasia)

One of the uncommon and overlooked uterine malformations is the accessory and cavitated uterine mass which is a separate cavity lined with working endometrial and myometrial tissue, apart from the usual uterine cavity. MRKH syndrome is yet another condition that starts in embryonic development and comes from the

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incomplete development of the paramesonephric ducts, causing a range of effects on the Müllerian structures. Both types of anomalies are capable of forming uterus-like formations with different levels of development and they often cause similar problems when it comes to diagnosis because they sometimes look the same as a unicornuate uterus with a rudimentary horn that does not communicate with the main uterus. The series gives special attention to the

MRI features that make it possible to tell apart these challenging diseases.

Case Presentation

Case 1

For the last seven months, a 17-year-old girl has been suffering from lower stomach discomfort. She had menarche at the age of sixteen. In the beginning, her symptoms were believed to belong to primary dysmenorrhea; however, the patient did not respond to antispasmodic medicines.

USG done afterwards showed a round, well-defined cystic lesion on the right lateral side of the uterus with thin walls and small particles inside it. There was no contact seen between the endometrial cavity and the uterine horn which could mean it is a non-communicating rudimentary uterine horn. MRI of the pelvic area was done to better explain the reported findings. An MRI revealed that the cyst is clearly round, thick-walled and appears enhanced outside near where the right round ligament enters. The wall of the lesion was the same brightness as the myometrium on T1-weighted pictures and darker than the myometrium on T2-weighted views, whereas the inside contents had hemorrhagic traits (Figure 1). It appears that the endometrium was not involved and there was no thickening in the uterus area. Very similar to people, both uterine horns were present and developed normally and the ovaries appeared normal as well.

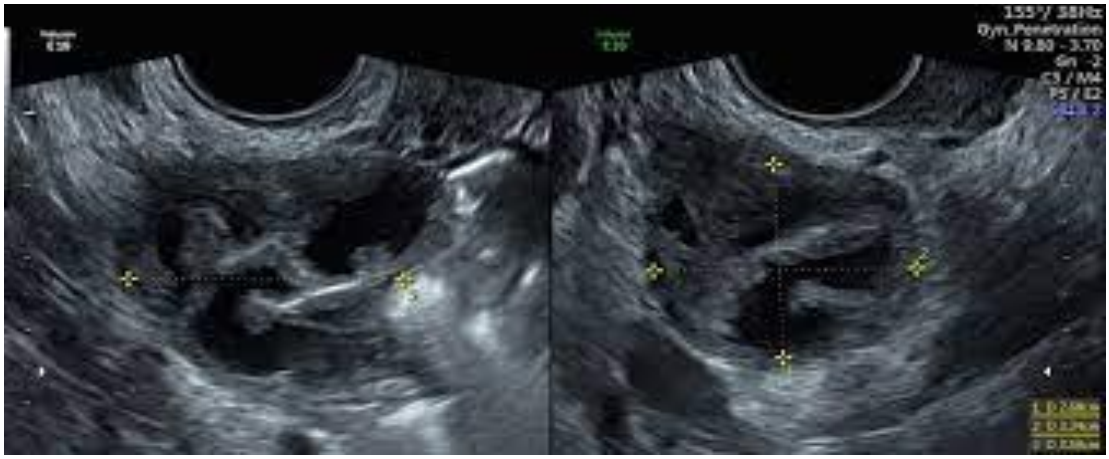

Figure 1: MRI of A 17-year-old female presented with a history of cyclic lower abdominal pain

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Its unique radiologic appearance and the usual site in an otherwise normal uterus, the patient was found to have ACUM (ESHRE/ESGE U6C0V0). During December 2021, the patient was treated with laparoscopic ACUM removal to clear all the bothersome symptoms.

The round ligament attaches frequently to these accessory cavities which are on the right side of the uterus and lined with myometrium. These conditions usually show up in females who have never been pregnant and are younger than 30. At present, it is suggested that ACUM happens when the female gubernaculum fails to function normally or when Müllerian remnants are present in a place other than the womb. To diagnose ACUM, one will find: (ione accessory cavitated mass; (ii) a cavity containing a functional endometrium; (iv) blood degradation products in the accessory cavity; (iii) a uterus, fallopian tubes, and ovaries that appear normal; and (v) no adenomyosis in the uterine tissue, unless a small amount is discovered adjacent to the accessory cavity.

The following are some differential diagnoses for ACUM:

- **Non-communicating rudimentary uterine horns** in other Müllerian anomalies. Taking into account the normal structure of the uterus is useful for telling apart ACUM from a hemiuterus with the banana-shaped horn. Such lesser functional horns are identified by the absence or small size of the uterus and other Müllerian structures in MRKH syndrome.
- **Uterine leiomyoma with cystic or red degeneration**, Such cases display several larger lesions pushing into the abdominal interior and sometimes histopathology is used to confirm the nature of these growths in ACUM.
- **Focal cystic adenomyosis**, Is mostly reported in females having children more than one time, between 35 and 40 years of age, presenting with poorly defined lesions that are far from the junction zone and do not create big masses on the endometrium, commonly combined with adenomyosis in the uterus.
- **Cornual ectopic pregnancy**, even though MRI can show a location very similar to ACUM's, it clearly shows that the gestational sac is positioned unevenly, has a small (<5 mm) thick rim and is more than 1 cm from the lateral side of the endometrium. In medical settings, secondary amenorrhea and a positive test for pregnancy help to confirm this disease.

Case 2

A 14-year-old girl was brought for evaluation because she had primary amenorrhea which had lasted six months and severe, non-stop cyclic pain in the lower abdomen. Her secondary sexual features looked normal and her hormone levels were not significant.

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The first pelvic ultrasound imaging revealed a tubular cystic formation with incomplete partitions and echoes within, as well as a distinct, well-shaped, thick-walled cyst in the right adnexa. The uterus showed no clearly visible parts in the ultrasound. The MRI scout also helped to learn that there was a simple horn in the right hip containing bright fluid on T1- and T2-weighted images which was confirmed by its absence of darkening on T1 fat-saturated images, pointing to hematometra. At the same time, next to this, a large round lesion near the right ovary had incomplete walls and shared the same imaging features typical of hematosalpinx. There was no trace of cervix or vagina in the patient, but both ovaries were seen to be normal (as shown in Figure 2). The renal and lumbosacral spinal areas were not affected by any abnormalities.

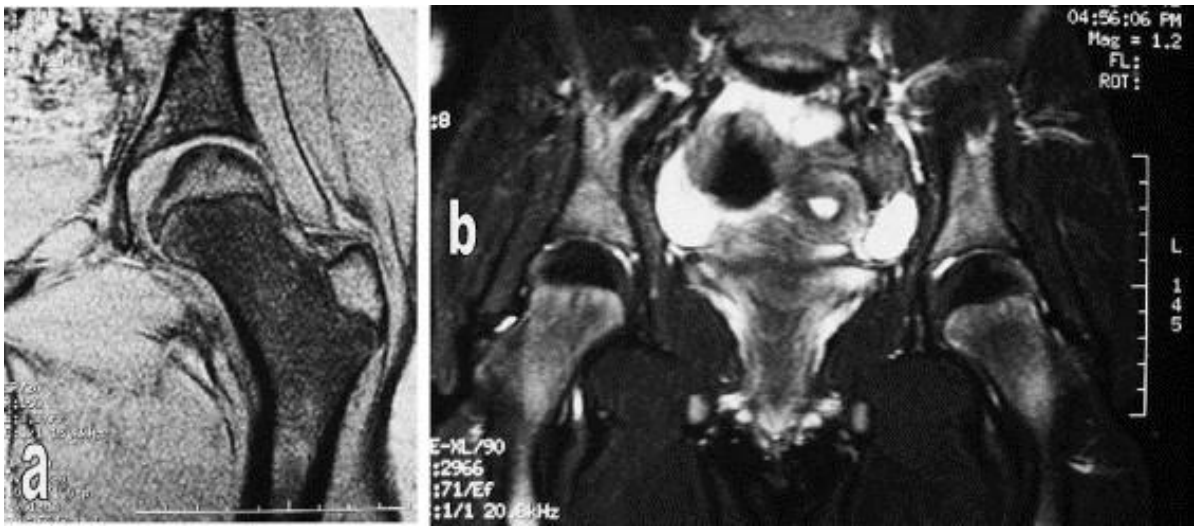

Figure 2: MRI of pelvic of a 14-year-old female was evaluated in February 2022 for primary amenorrhea accompanied by severe, unrelenting cyclic lower abdominal pain of six months' duration

With a functional right uterine horn with hematometra and hematosalpinx, as well as cervical and vaginal absence, the condition was determined to be atypical MRKH (ESHRE/ESGE U5aC4V4). The patient is booked for a procedure called laparoscopic excision of the working rudimentary horn.

A main sign for MRKH syndrome is when someone experiences no menstruation at puberty, alongside usual secondary sexual development due to ovarian activity. MRI allows healthcare professionals to look closely at various problems such as having the uterus entirely or partly missing or uterus structures that are not well-developed. In the rudimentary uterus, the uterine corpus and cervix are not normally differentiated, the uterus is situated in the pelvis around the ovary and it is connected with a constant relationship to the caudal wall. MRI is better at showing these abnormalities than a transabdominal ultrasound.

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Case 3

A 16-year-old woman came to the clinic in October 2021, complaining of ongoing menstrual pain for eight months which continued even though her hormones and sex organs were normal. At the beginning of the test, the ultrasound showed that the uterus had two different cavities and inside the right cavity, there was anechoic fluid echoed by some thin lines. It was found in the right adnexal region that a defined tubular anechoic structure with incomplete separation or septation, was present. Given these results, it was likely that the patient has hemi-uterus with right-sided accumulation of both blood in the womb lining and in the fallopian tube or hematometra and hematosalpinx.

Additional evaluation with MRI of the pelvic area showed that the left part of the uterus shaped like a banana was there, showing standard parts and linked to a normally formed cervix and vagina. Nearby, a big cavity lined by the myometrium was identified in the right side of the pelvis and it was attached to the left uterus by a fibrous tissue. On all these sequences, the cavity showed heterogeneous hyperintensity which is compatible with hematometra located in a non-connected right rudimentary uterine horn. A hematosalpinx was seen on the right side too. Both of her ovaries were viewed from both sides and seemed normal.

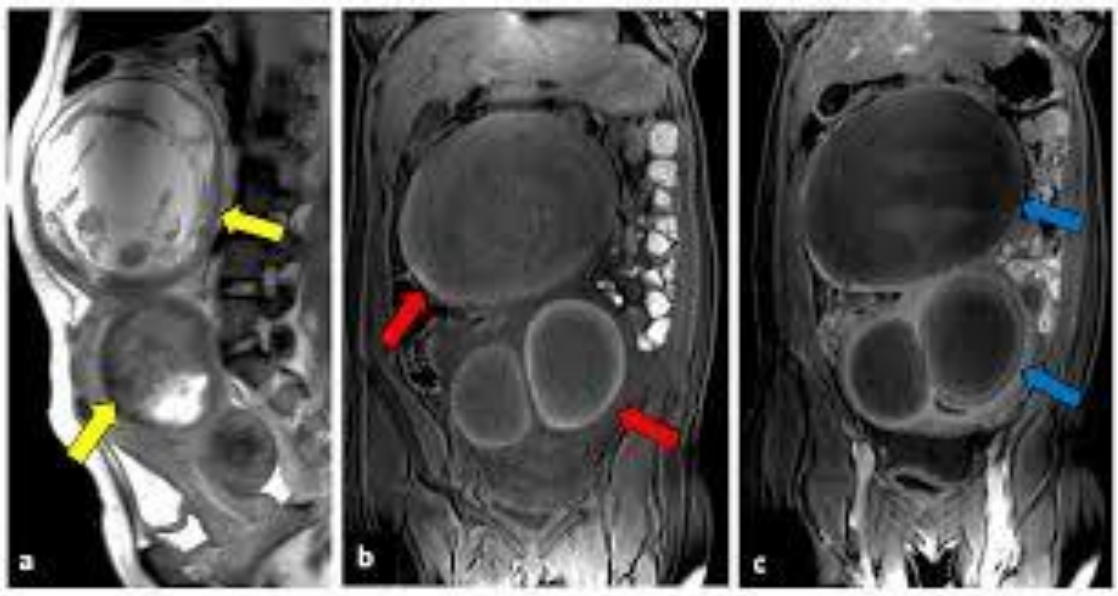

Figure 3 MRI of pelvic of a 16-year-old female presented with persistent dysmenorrhea lasting eight months, despite normal hormonal profiles and secondary sexual characteristics

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The presence of a hemi-uterus with a functional and non-communicating right rudimentary horn was therefore verified, while hematometra and ipsilateral hematosalpinx (with the ESHRE/ESGE code U4aC0V0) further confused the situation. All symptoms were eliminated when the patient had laparoscopic surgery in January 2022 to remove the obstructed rudimentary horn.

If just one Müllerian duct grows properly, the uterine structure is known as a hemi-uterus. This condition may result in a rudimentary horn that is functional or it may exist without one, leaving a non-functional or aplastic cavity instead. Among the other possible diagnoses to look into are endometriosis, adenomyosis and obstructive vaginal.

For example, young girls might be born with obstructed vagina in only one side and related anomalies of the kidney, what is known as OHVIRA syndrome. A detailed examination and focused imaging are necessary for properly diagnosing and treating patients who keep experiencing cyclic pelvic pain.

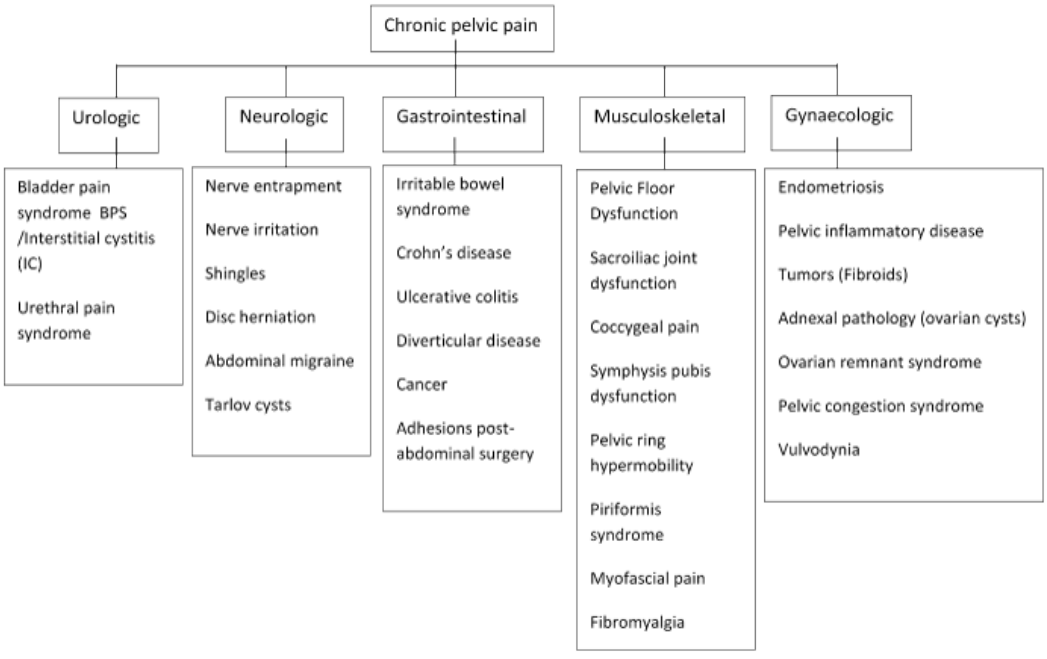

Figure 4: Graphical Presentation of Chronic Pelvic Pain

Conclusions

The possibility of diseases in the uterus with blocked cavities that are lined with the endometrium should always be looked into in young females suffering from chronic pelvic pain. MRI is needed to diagnose these conditions. As dysmenorrhea can have many nonspecific symptoms, it is

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important to look at the uterus, cervix and vagina using radiology and examine the distance between the functional cavities, uterus and ovaries for accurate diagnosis. Although ultrasound can detect the existence of non-communicating cavities, MRI is the best method to see if the rudimentary uterine horns or accessory cavities work. Accurate diagnosis of the problem allows surgeons to perform the right operation which usually results in eliminating the symptoms.

Abbreviations

- MRI: Magnetic Resonance Imaging
- ACUM: Accessory and Cavitated Uterine Mass
- MRKH: Mayer–Rokitansky–Küster–Hauser Syndrome
- ESHRE: European Society of Human Reproduction and Embryology
- ESGE: European Society for Gynaecological Endoscopy
- OHVIRA: Obstructed Hemivagina with Ipsilateral Renal Anomaly
- T1WSE: T1-Weighted Spin Echo
- T2WSE: T2-Weighted Spin Echo
- T1WFS: T1-Weighted Fat Saturated

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