Case Reports

Mullerian Duct Anomaly: Two Patients in Adolescent Age in C. R. Gardi Hospital

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Clinical manifestations of Mullerian duct abnormalities vary greatly and can range from isolated abnormalities of a single reproductive organ to multiorgan aplasia. An early and precise diagnosis is essential because untreated symptoms may result in pelvic endometriosis, retrograde tubal reflux, and infertility. MRI and ultrasonography are the imaging modality that accurately helps in the diagnosis of mullerian duct anomalies. Two interesting cases came to C. R. Gardi OPD with Mullerian duct anomalies. The first case came with dysmenorrhea and a lower abdominal palpable mass upto 14 wk size was present, which was diagnosed to be bicornuate uterus, obstructed hemivagina. It is a diagnostic dilemma because menses were regular. Another case came with severe cyclic dysmenorrhea and was found to be a hematometra in the non-communicating rudimentary horn on the left side.

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Introduction

Female reproductive tract developmental diseases are known as mullerian duct anomalies (MDAs). MDA prevalence is 6.7%.[1] MDAs have diverse and complicated etiology. MDAs are developmental abnormalities that result from irregularities in the Mullerian ducts, which are the embryonic analogs of the uterus, fallopian tubes, and upper two-thirds of the vagina.[1] The etiology and pathophysiology of MDAs are poorly understood despite their clinical importance. Clinical manifestations of MDAs span a wide range, from isolated abnormalities of a single reproductive organ to multiorgan aplasia. They may coexist with additional syndromes. This case report discusses the most recent advancements in this area and fresh possibilities that may significantly increase our understanding of the causes of MDA and in MDA diagnosis, treatment, and prevention, including genetic counseling.

Patient 1

A 15-year-old girl experienced of excruciating cyclic pain in her abdomen during menstruation for the past two years. Lower abdominal palpable mass up to 14 wk size was present. Menarche at 13 year of age, secondary sex characters normal. The patient was lean and thin built. Breast- Tanner stage V. The gynecological examination revealed a grossly normal vulva, and the abdomen revealed no abnormality. On transabdominal ultrasound a bicornuate uterus with an anechoic collection measuring 9.9 x 8.8 x 8.1 cm approx. (375 mL) in right-sided endometrial and cervical cavity s/o hematometra, another anechoic cystic collection in right adnexa 3 x 4.2 x 4.7 cm approx. (25 mL) s/o? Right hydrosalpinx. MRI shows a developmental malformed uterus with two separate endometrial cavities and a complete septum extending into the cervix. Right-sided endometrial and cervical cavity shows hematometra measuring 8.6 x 8.6 x 6.8 cm in size, suggestive of obstructed right hemivagina. Right fallopian tube suggestive of hematosalpinx. Absent right kidney seen. We managed the patient by performing an exploratory laparotomy. Right-sided hematosalpinx, hematometra and hematocolpos incised and drained completely with right cuff salpingostomy. The vaginal opening was traced, resection of the transverse vaginal septum was done, and an opening was created between the vagina and cervix.

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Patient 2

A 17-year-old girl suffered cyclic abdominal pain followed by bleeding for 2.5 years. She reached menarche at the age of 14, and secondary sexual characteristics first manifested at the age of 12. She seemed physically slim and skinny, with stage V tanner breasts and typical axillary and pubic hair distribution. The gynecological examination revealed a vulva that was very normal. The ultrasound revealed an echoic collection with dense internal echoes on the left-sided endometrial and cervical cavities. According to an MRI, a functioning unicornuate uterus on the right side is connected to a non-communicating left-side rudimentary horn that measures around 7 x 6.3 x 6.4 cm in size. Metroplasty is used after an exploratory laparotomy to treat the condition. The non-communicating horn was cut, and the hematometra was drained. During a metroplasty, a catheter was placed from below, keeping the uterine cavity open.

Discussion

On the basis of sonography and MRI, the disease in our first patient, a 15-year-old girl, was identified to be a bicornuate uterus with unilateral hematocolpos, hematometra, and hematosalpinx with ipsilateral renal agenesis.

Our second patient, who had been experiencing significant abdominal pain for the previous five days, appeared with menstrual bleeding. On ultrasonography, both ovaries were discovered individually, and a congenital uterine abnormality (unicornuate) with a blockage and hematometra of the embryonic horn was also noted. It has a right-sided unicornuate uterus and Type II mullerian duct abnormalities found with hematometra in a non-communicating primordial uterine horn.

Congenital uterine or mullerian duct anomalies are an uncommon cause of infertility in women but are commonly treated for the syndrome when it develops. They are predicted to affect 0.1 to 0.5% of women, with a frequency of around 3% among women who have experienced numerous miscarriages. Reproductive issues, such as a greater prevalence of infertility, affect 25% of women with mullerian duct abnormalities.[2]

The lower third of the vagina develops independently from the urogenital sinus, whereas the uterus, fallopian tubes, cervix, and upper 2/3 of the vagina develop from the paired mullerian ducts. Because the Mullerian and metanephric ducts are concurrently affected by embryologic interruptions during the eighth gestational week, congested hemivagina and renal agenesis on the side of congestion are associated with uterine didelphys. Congenital malformations of the female genital tract, known as mullerian duct anomalies, are brought on by either unsuccessful resorption of the uterine septum or non-development or non-fusion of the Mullerian ducts.[3-5] These abnormalities are thought to occur between 0.5 and 5% of the life.[6] Most of these episodes are identified during menstruation. Early and precise diagnosis is essential since untreated episodes may result in endometriosis and retrograde tubal reflux. Later in age, it may also lead to obstetric problems and reduced fertility. Usually longitudinal and oblique, the obstructive vaginal septum found in this syndrome ranges in thickness from very thin to fairly substantial. The Müllerian ducts and the urogenital sinus have a vertical fusion dysfunction in the transverse vaginal septum. It is typically not accompanied by other urologic or Müllerian abnormalities and might be full or partial.[7] After menarche, it manifests as periodic abdominal pain with a steadily growing pelvic lump and regular menstrual periods. Distal occlusion, like in our scenario, may cause reversed menstruation with the emergence of a right hematosalpinx. The preferred method of treating blocked hemivagina is resection of the vaginal septum. Ultrasonography, sonohysterosalpingography, conventional and sonohysterosalpingography, and magnetic resonance imaging are imaging techniques used to identify this problem. The imaging modality of ultrasound, which is economical, noninvasive, and commonly approachable, correctly aids in the identification of this illness. Nevertheless, the vaginal septum is challenging to see on ultrasound and is best projected on MRI.

The challenge in these patients is the preservation of menstrual and child-bearing function. By proper reconstruction technique, we could preserve the uterus.

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