Case Report

Müllerian Duct Fusion Anomaly Associated with Unilateral Renal Agenesis

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The purpose of this study was to check for the associated anomalies present in a suspected case of Müllerian duct fusion anomalies. Almost one-third of women with a renal anomaly will have Müllerian duct anomalies. MRI plays an important role in evaluating abnormal uterine morphology with other association like unilateral renal agenesis and vertebral anomaly. In this case report we present two cases of rare Müllerian fusion anomaly associated with unilateral renal agenesis.

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Key words **: Müllerian Duct anomaly.**

Congenital uterine anomaly occurs due to abnormal formation, fusion or reabsorption of the Müllerian ducts during fetal life. The process may be partial or total and affect one or multiple parts of the female urogenital tract[1].

Müllerian duct anomalies is a rare developmental anomalies which occur in about 7% of the general population and in almost one-third of women with a renal anomaly[2].

Case 1

A 19-year-old female unmarried patient referred to Department for ultrasound pelvis and MRI pelvis due to menstrual cramps and progressive pain in the hypogastric region. She had started menstruating for 8 months. Menstruation is irregular with associated mild pain during menstruation. Abdominopelvic ultrasound scan revealed uterus didelphys with right hematocolpus measuring ~38mm × 28mm × 51mm, obstructed right hemivagina. The right kidney was not visualized. MRI (Fig 1) revealed two uterine horns with two external os opening into two separate vagina. Marked distention of right hemivagina with fluid levels due to transverse band at lower third right hemivagina and right renal agenesis were noted. Patient underwent surgical management for obstructed hemivagina and the pre-operative findings confirmed the imaging findings of Herlyn-Werner-Wunderlich Syndrome (HWWS).

Case 2

A 25-year-old female married patient referred to Department for MRI pelvis due to menstrual cramps, progressive pain in the hypogastric region and right iliac fossa. Physical examination showed tenderness in right iliac fossa and a palpable mass. Pelvic ultrasound scan report showed complex right adnexal cyst with bicornuate uterine morphology. Patient was referred to MRI for better characterization of adnexal pathology. MRI (Fig 2) revealed two uterine horns with single cervix and single vagina. Marked distention of right cornua of uterus and the opening of right cornua into the cervix is not clearly visualized. There is a right ovarian hemorrhagic cyst with scoliosis of lumbar spine with hemi L2 vertebra with absent right kidney. Final diagnosis is Bicornuate unicollis uterine morphology with obstructed right cornua associated with hematometra. Right renal agenesis with L2 hemivertebra and right ovarian hemorrhagic cyst. The

Editor's Comment :

- This study aimed to identify anomalies associated with Müllerian duct fusion anomalies. About one-third of women with renal anomalies also have Müllerian duct anomalies. MRI is essential for evaluating abnormal uterine morphology and its associations, such as unilateral renal agenesis and vertebral anomalies. All women with Müllerian duct fusion anomalies should be screened for renal agenesis. Radiological investigations, including ultrasound and MRI, are crucial for assessing uterine and obstructive uterovaginal anomalies.

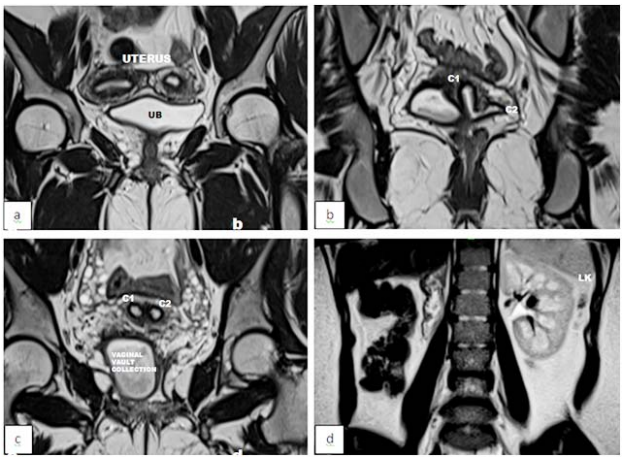

Fig 1 — (a) T2 coronal images showing two separate cornua of the uterus. (b) T2 coronal images showing two separate vagina, with heterogenous collection in the right vaginal vault. (c) T2 coronal images showing right vaginal heterogenous collection with a septum at lower aspect, left vaginal vault is patent. (d) T2 coronal images showing absent right kidney with hypertrophy of left kidney.

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patient was advised surgical management for cystic lesion but the patient opted for conservative treatment and we could not do follow up of the patient.

Discussion

Müllerian duct normally fuses between the 6th and 11th weeks of gestation to form the uterus, fallopian tubes, cervix and proximal two-thirds of the vagina.

Any Interruption of the Müllerian duct during the *fusion* process gives rise to uterine didelphys and bicornuate uterus subtypes of Müllerian ductal anomalies[3].

There is a close embryologic relationship between the development of the urinary and reproductive organs, hence Müllerian ductal malformation are associated with urinary tract anomaly. Renal anomalies occur most frequently in patient with unicornuate (29.5%) and didelphic uterus (29.1%). Lower rates of abnormal renal tract (11.7-15.2%) were noted in women with uterine agenesis, septate or bicornuate uterus. Congenital absence of one kidney has been the most common urologic anomaly associated with obstructive uterovaginal anomalies[4].

According to the ASRM 2016 guidelines (American Society for Reproductive Medicine Classification System), the bicornutate uterus is defined as an external fundal indentation of greater than 1 cm and the uterine didelphys is defined as two separate uterine bodies with duplication of the cervix[2].

Bicornuate uterus accounts for approximately 10% of MDA and it occurs due to incomplete or partial fusion of the Müllerian ducts. The duplicated endometrial cavity may be associated with cervix duplication (bicornuate bicollis) or be without cervix duplication (bicornuate unicollis).

Uterus didelphys results from complete failure of Müllerian duct fusion. Each duct develops fully with duplication of the uterine horns, cervix and proximal vagina. Transverse hemivaginal septum may be seen in proximal duplicated vagina which results in ipsilateral obstruction and hematometrocolpos[3].

Herlyn-Werner-Wunderlich Syndrome (HWWS) is a rare Müllerian duct anomalies with mesonephric duct anomalies with the triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis. The incidence of didelphys uterus, related to HWW, is approximately 1/2,000 to 1/28,000 and it is accompanied by unilateral renal agenesis in 43% of cases[4].

Patients may be asymptomatic until menarche but may present with progressive pain due to hydrometrocolpos and hemivaginal obstruction within the first year of menstruation. The most common clinical presentation is abdominal pain, cyclical dysmenorrhoea at puberty and abdominal mass secondary to hematocolpos and normal menstrual periods[5,6].

This condition can be managed by full resection of the vaginal septum to relieve the obstruction and also helps to prevent the development of further complications, and restore functionality of genital system. Laparoscopic hemihysterectomy may be a effective alternative treatment for patients with a didelphic uterus with a hypoplastic cervix. Ipsilateral hysterectomy is recommended in cases

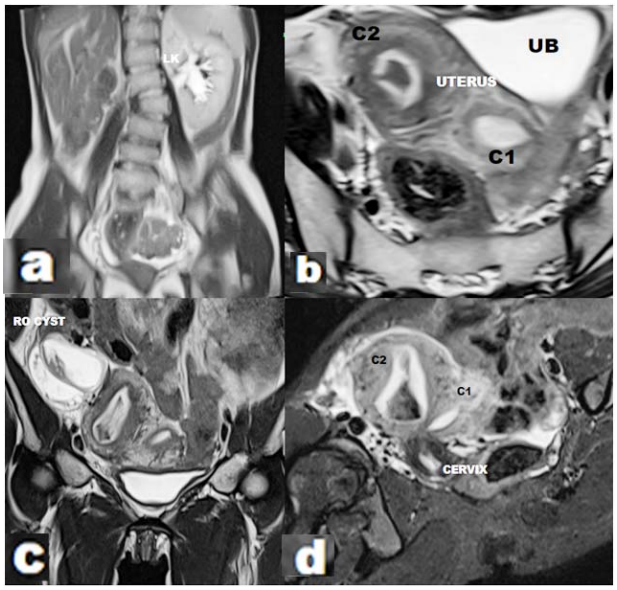

Fig 2 — (a) T2 coronal images showing absent right kidney and L2 hemivertebra. (b) T2 axial images showing two separate uterine cornua with collection in right uterine cornu. (c) T2 coronal images showing two separate uterine cornua with collection in right uterine cornua and a right ovarian hemorrhagic cyst. (d) T2 fat suppression oblique coronal showing single cervix with two separate uterine cornua.

of cervical atresia, because resection of the septum would not relieve obstructed symptoms[7].

Conclusion

All women with Müllerian duct fusion anomalies should be checked for presence or absence of renal agenesis. Radiological investigation including Ultrasound and MRI plays an important role in assessing the type of uterine and obstructive uterovaginal anomalies.

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