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Obstructed Hemivagina and Ipsilateral Renal Anomaly, An Uncommon Entity

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Abstract

Obstructed hemivagina and ipsilateral renal anomaly also known as OHVIRA-Herlyn Werner-Wunderlich (HWW) syndrome is a rare female urogenital anomaly. There is delay in diagnosis because of regular menstruation and nonspecific abdominal pain. Clinical presentation begins in post-menarche young girl with lower abdominal pain, with formation of pelvic mass. Its management needs to focus on timely, right diagnosis and management. Present case had right sided hematometrocolpos with right sided atresia of the cervix, vaginal septum and right renal agenesis in a eighteen years old girl who presented with cyclical abdominal pain, distension of abdomen increasing since one year. Ultrasonography revealed uterine didelphys, with 6.6 x 3.5 cms size collection with internal echoes at cervical region and right sided hematometra. Right kidney was not visualized in right renal fossa. MRI pelvis was suggestive of uterus didelphys with two separate uterine horns and one cervix seen on left side. Right-sided uterine horn and cervix were dilated as one sac and showed an hourglass appearance. Under anesthesia examination revealed vaginal bulge on right side and normal left part of vagina, with a single cervix. There was septum from right vaginal wall to cervix on left side, confirming the clinical and imaging diagnosis. Hematometrocolpos was drained, septum excision was done, and margins of septum were everted and fixed on lateral vaginal wall. Right-sided hematometrocolpos with right-sided atresia of the cervix with vaginal septum along with right renal agenesis, were diagnosed by clinical examination, added by imaging techniques and managed with surgical intervention. Because of rarity the case is being shared.

Keywords: HVIRA - herlyn werner-wunderlich syndrome; renal agenesis; hematometrocolpos; diagnosis; management

Introduction

Obstructed hemivagina and ipsilateral renal anomaly also known as OHVIRA -Werner-Wunderlich (OWW) syndrome is a rare female urogenital anomaly. It was initially described by Herlyn-Werner in 1971 and later in 1976 Wunderlich described an association of right renal aplasia with a bicornuate uterus and single vagina associated with hematocolpos. It is believed to result from the combination of Mesonephric and Mullerian ducts anomalies [1, 2]. The exact incidence of this syndrome is unknown, but it is estimated to be 0.1%to 3.8% of all Mullerian anomalies [3]. There is delay in diagnosis because of regular menstruation and nonspecific abdominal pain. Later collection of blood above vaginal septum and in uterus increases severity of pain, which brings the patient to hospital. Its management needs to be focused on early, correct diagnosis and septoplasty, which relieves pain and prevents further complications such as endometriosis, infertility and others [4].

Case Summary

An eighteen years old girl was admitted to the gynecology wards because of cyclical abdominal and pelvic pain, with feeling of increasing distension 0f abdomen over one year. The pain used to get worse every day with menstruation for which she used to take non-steroid anti-inflammatory drugs. She had menarche at age of 14 years and her last menstrual period was a month back, with previous regular cycles, needing 2 pads/ day during menstruation. Menstrual pain exaggerated in the last one year. Neither bowel & bladder abnormalities were reported nor were found on clinical examination. With due consent, speculum examination revealed bulge on right side of vagina near left side cervix, and on vaginal examination palpable mass was felt in upper vaginal quadrant. Her Hemoglobin was 8.6 gm% and no abnormality was detected in day-to-day investigations. Ultrasonography revealed uterine didelphys with 6.6 x 3.5cms collection with internal echoes present at cervical region on right side, suggestive of right sided hematometrocolpos. Right kidney was not visualized in right renal fossa. MRI pelvis was suggestive of uterus didelphys with two separate uterine horns but single cervix visible. Right side uterine horn and cervix were dilated, as one sac and

showed hour glass appearance measuring I10x 32 x29 mm with T2 hypo intense and TI hyper intense collection with narrowing at the level of vagina. Also noted was oblong shaped tubular structure posteriorly having similar signal intensities - likely representing hematometrocolpos and hematosalpix. Uterus and cervix were normal on left side. Right renal agenesis was revealed. Patient was examined under anesthesia also which showed right vaginal bulge with normal left part of vagina and single cervix were present. Large bore needle was inserted in vaginal bulge and old blood around 300ml was drained, Septum excision was performed, and margins were everted with vagina on right vaginal wall. Post-operative management was urinary catheterization, valval hygiene, and antibiotics.

Discussion

The specific causes and etiology of OHVIRA syndrome remains unknown. It is thought to be a representation of the abnormal development of the paramesonephric (Mullerian) and Mesonephric (Wolffian) ducts. Consequently, unilateral renal agenesis with imperforate hemivagina may be caused because of developmental abnormality of the caudal part of one of the Wolffian ducts [1,3]. The Mullerian duct is shifted laterally and unable to fuse with the contralateral duct on the side where the Wolffian duct is lacking, resulting in a didelphys uterus. The misplaced Mullerian duct that cannot come into contact with the urogenital sinus centrally develops into blind spot giving rise to an imperforate or blocked hemivagina, whereas the contralateral Mullerian duct gives rise to vagina and 75% of these defects are connected to an obstructing vaginal septum which was present in the case being shared, as oblique septum [4,5]. The girl presented with dysmenorrhea and was clinically suspected to be having unilateral Mullerian anomaly and the diagnosis was confirmed by Sonography and MRI as others have reported [6,7]. The most frequent associated non genital anomaly is renal agenesis invariably present with the side of genital anomaly, as was present in the case being shared. The cyclical increasing abdominal pain secondary to hematrometrocolpos because of retained partially clotted menstrual blood in the obstructed hemivagina and ipsilateral renal anomaly syndrome are typically discovered shortly after menarche because dysmenorrhea is common in this age and menstrual flow from normal side gives the feeling of normality. This syndrome initially may remain undiagnosed and patient reports when severity of pain increases and may be bladder symptoms occur. Restoration of menstrual flow relieves the symptoms and cures the patient and the same was done in the case being shared.

Conclusion

A case of uterus didelphys with unilateral oblique vaginal septum with haemetrocolpus and ipsilateral renal agenesis is being shared because of its rarity. Case was clinically suspected and was finally diagnosed by adding USG, MRI and examination under anesthesia and treated surgically.

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