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Full Length Research Article

OHVIRA SYNDROME – A CASE REPORT

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ABSTRACT

Back ground: OHVIRA syndrome is a very rare congenital Uro-genital anomaly associated with both the Mullerian (paramesonephric) and Wolffian (mesonephric) systems. It is characterized by a triad, OHV – Obstructed Hemi vagina, IRA—Ipsilateral Renal Agenesis with associated uterus didelphs. Mullerian Duct anomalies 0.8-4% of the women who came for evaluation of infertility. OHVIRA syndrome-0.1-3.5% of the Mullerian Duct Anamoly.
Aims: To discuss and analyse the radiological and embryological co-relation in OHVIRA syndrome and study the genesis of both the urinary and genital systems.
Materials and Methods: A 17 year old girl, nulligravida, presented to Kamineni Institute of Medical Sciences, Narketpally, with progressive development of cyclic lower abdominal discomfort and a large abdomino- pelvic mass. She was thoroughly investigated including the 3 imaging modalities like USG, CECT and MRI which was in favour of OHVIRA syndrome.
Observation: On examination revealed a large, tense, protuberant mass extending slightly beyond her umbilicus, corresponding to 20weeks of gestation.
Conclusions: An early correct diagnosis is the goal to relieve the symptom and prevent complications caused by retrograde menstruation which may lead to endometriosis and subsequently to conception abilities; a two step vagino-plasty can be performed to correct the anomaly.

INTRODUCTION

The prevalence of congenital Mullerian duct anomalies is reported to be 1 % (Ashton *et al*., 1988). The syndrome of obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) is a rare congenital anomaly of the Mullerian ducts (paramesonephric) and Wolffian structures (mesonephric). It usually includes unilateral renal anomalies and uterine didelphs (Kimble *et al*., 2009). Most of the women suffering from OHVIRA syndrome are diagnosed late due to the rare incidence and the non- specific clinical presentation. Moreover, the menstrual flow that comes from the patent unobstructed hemi-vagina gives the appearance of normal menses. Consequently accurate diagnosis and surgical treatment can be delayed for several months or even years. OHVIRA could present with lower abdominal pain, severe dysmenorrhea, a pelvic or vaginal mass, abnormal vaginal discharge, inter-menstrual bleeding (Nigam *et al*., 2011), acute retention of urine, fever, vomiting (Mandava *et al*., 2012), infertility and abdominal swelling or complication with pregnancy and labour (Shavell *et al*., 2009). Here we present an interesting case report of OHVIRA.

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Case report

A 17 year-old female, unmarried nulli-gravida presented to our clinic with, Left lower quadrant bulging and pain. Pain showed cyclical association with onset of menses and progressive lower abdominal distension, Increased urinary frequency and urgency in addition to constipation. The patient attained menarche at the age of 12 years and had regular menstrual cycles, once every 28 days with 3–5 days' duration of menstrual flow associated with dysmenorrhoea. O/E Temperature: A-febrile, Vital signs, WBC, Serum Creatinine: Normal, Pregnancy test: Negative, Laboratory tests, including complete blood count and urinalysis, hormonal profile and a pregnancy test was negative. General physical examination showed no abnormalities with well developed breasts and normal feminine axillary and pubic hair distribution. Per Abdominal Examination shows, Large, tense, protuberant

mass extending slightly beyond her umbilicus corresponding to 20 weeks of gestation. Abdominal/pelvic ultrasound (2D/3D) revealed two widely divergent well developed uterine horns (hemicorpora); the left horn was larger than the right one (uterus didelphs). Each hemiuterus was relatively reduced in size. The left horn measures 46.5 mm in length, 32.0 mm in width with a volume of 24.9 ml. The right horn measures 43.0 mm in length, 24.0 mm in width with a volume of 15.1 ml. There were two normally developed cervices adherent together. There was a curved partial longitudinal vaginal septum originating from the right side of the left cervix and attached to the right lateral vaginal wall. The left hemi hematocolpos was measuring 18cmx11cm; it was filled with an echogenic fluid, chocolate like material. The left fallopian tube was distended (hydrosalpinx). Both ovaries appeared normal in size. The left kidney was not visualized. Intravenous excretory urography (IVP) showed no visualization of the left kidney and left ureter with mild compensatory hypertrophy of the right kidney.

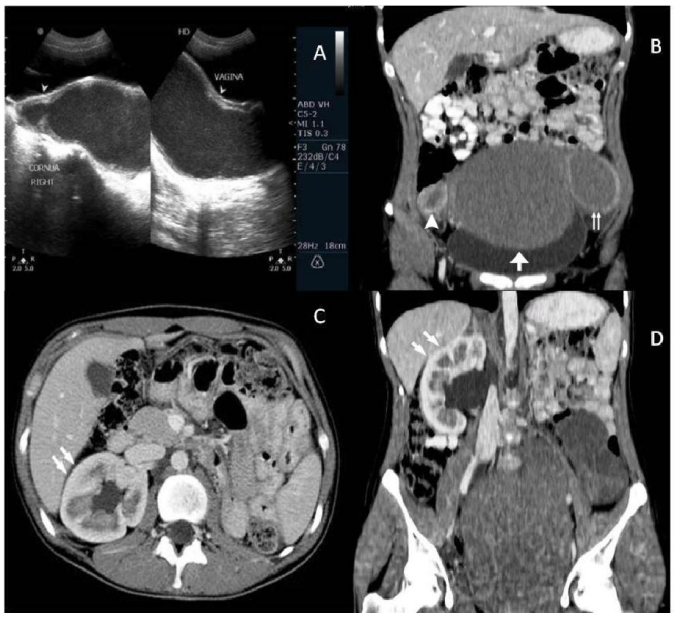

Figure1 A: USG image of a large hematometrocolpos shows a markedly dilated vagina, approximately 18x11cm containing echogenic material consistent with blood. b): Coronal CECT shows blood filled dilated vagina (↑), normal right hemiuterine horn (↑ head), distended left hemiuterine horn (↑↑), suggesting blood, an absent left kidney, a midline pelvic mass approximately 11x12x18cm, connected in the left pelvis consistent with a hydrometrocolpos and left hydrosalpinx. C) Axial CECT shows a mildly hypertrophied right kidney and an absent left kidney. D) Coronal CECT shows a mildly hypertrophied right kidney and an absent left kidney.

Magnetic resonance imaging (MRI) showed two separate uteri and cervices with longitudinal vaginal septum and an obliterated left side that was seen distended and filled with blood signal displaying high signal on T1 and T2 W images with a small linear blood filled tract extending from the left lateral wall of the cervix to the blind end (Fig. 2B**)**.

On the basis of the imaging findings of unilateral renal agenesis, uterus didelphys, unilateral obstructed hemivagina with resultant hematometrocolpos and hydrosalpinx, the case was diagnosed as OHVIRA, or Herlyn–Werner–Wunderlich syndrome.

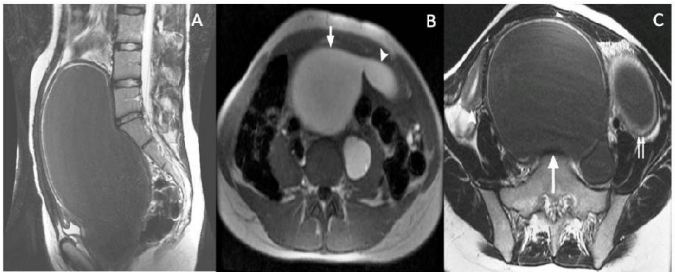

Figure 2 a) Sagittal T_2 weighted MRI, shows a dilated hemivagina with low T_2 - signal-intensity contents and corresponding high T_1 - signal intensity consistent with blood. b) Axial T_1 WI shows left hydrosalpinx containing high T_1 signal intensity contents consistent with blood products, originating from a dilated hemivagina. c) Axial T_2 WI shows a dilated vagina with low T_2 and high T_1 signal intensity contents consistent with blood products, normal appearing right uterine horn and dilated left uterine horn with low T_2 signal intensity blood products.

A diagnostic laparoscopy was performed to determine the extent of hematometra and hydrosalpinx and to assess the adnexae for any endometriosis resulting from retro-grade menstrual flow. Laparoscopic findings confirmed the abdominal/pelvic ultrasound and MRI findings, but it did not reveal any endometrial peritoneal deposits over the right fallopian tube and right ovary and in the recto-uterine pouch. The methylene blue test instilled through the visible cervix was +ve on the right tubal side demonstrating a lack of communication between uterine cavities. The patient was treated with hemi-vaginal septal resection and draining of hematometrocolpos and a 12 Foley catheter was left inside the cavity for one week to reduce adhesion. Antibiotics were continued for further 5 days. The postoperative course and follow up for 9 months were uneventful.

DISCUSSION

Embryology: Development of the Mullerian system: Initially both male and female embryos have two pairs of genital ducts[12] a) Mesonephric (Wolffian) duct, b) Paramesonephric (Mullerian) duct. The paramesonephric duct arises as a longitudinal invagination of the coelomic epithelium on the anterolateral surface of the urogenital ridge which develops in the 5th week of intra uterine life. Cranially the duct opens into the abdominal cavity with a funnel like structure, caudally it runs first lateral to the mesonephric duct, then crosses it ventrally to grow caudomedially. In the mid line it comes in close contact with with paramesonephric duct from the opposite side. Untill the 7th week the gonads, (male and female) do not acquire differentiation. The gonads appear as a pair of longitudinal ridges which are formed by the proliferation of the coelomic epithelium and a condensation of the underlying mesenchyme of the urogenital ridge (Sadler *et al*., 2010). The two paired Mullerian ducts develop into the fallopian tubes, uterus, cervix and the upper 2/3 of the vagina. The ovaries and the lower 1\3 of the vagina do not develop from the Mullerian system (Dutta, 2011). The Mullerian duct anomalies occur from defects in one of the 3 stages of development of the Mullerian ducts (Fertil Steril, 1998). Organogenesis, Lateral Fusion, Septal resorption, Lateral

Fusion. The process during which the lower segments of the paired Mullerian ducts fuse to form the uterus, cervix and the upper 2/3 of the vagina is termed as lateral fusion. Failure of fusion results in anomalies such as bicornuate uterus, or didelphs uterus. The term vertical fusion is used to refer to fusion of the ascending sino vaginal bulbs with the descending Mullerian system (i.e. fusion of lower 1/3 and upper 2/3 of the vagina). Complete vertical fusion forms a normal patent vagina while incomplete vertical fusion results in an imperforate hymen. Development of vagina; The endodermal cloaca is partitioned by a urorectal septum into a dorsal rectum and a ventral primitive urogenital sinus (Dutta, 2011). The primitive urogenital sinus is divided by the openings of the mesonephric ducts into cephalic and caudal parts (Dutta, 2011). The caudal part forms the definite urogenital sinus and the lower dilated phallic part presents a Mullerian eminence which is formed by the caudal ends of the paramesonephric ducts. In the females the peripheral part persists as the hymenal membrane and the central part disappears to form the hymenal orifice. Hence the persistence of the central part of the Mullerian eminence results in imperforate hymen leading to cryptomenorrhoea (Dutta, 2011). The ovaries and the lower vagina are not derived from the Mullerian system. The ovaries are derived from the germ cells that migrate from the primitive yolk sac into the mesenchyme of the peritoneal cavity and subsequently develop into ova and supporting cells. Development of the urinary system; The kidney develops from two sources: Metanephric mesoderm which provides the excretory units and Ureteric bud which gives rise to the collecting system, Failure of development of these gives rise to renal agenesis (Sadler *et al*., 2010).

Causes: Hereditary and genetics: The WNT4 gene encodes glycoproteins that serve as signalling molecules during early development. Normally the absence of Anti Mullerian Hormone (AMH), gene is SOX9 will trigger stabilization of the Mullerian system and regression of the wolffian system leading to development of the female reproductive tract. WNT4 gene is the ovary determining gene. Without WNT 4gene the Mullerian system is either deformed or absent (Dutta, 2011). The female reproductive tract develops at the same time and close to the urinary tract and kidneys from the intermediate mesoderm. As a result, developmental problems in the female reproductive tract sometimes occur with problems in other areas, including the urinary tract, kidneys, such as pelvic kidney, absent kidney, duplication of the collecting system, or ectopic ureters (Rackow and Arici, 2007) or multicystic kidney (Shavell *et al*., 2009). The OHVIRA syndrome is classically associated with uterus didelphys (type III, American Society for Reproductive Medicine classification) or rarely a complete septate uterus (type V) (Rackow and Arici, 2007). Early detection of Mullerian anomalies is important for counselling and planning the proper management and helps in preventing complications and preserving future fertility. Although, MRI is widely and effectively used in the diagnosis of uterine anomalies, especially the vaginal septum with approximate 100% accuracy (Kimble *et al*., 2009).

Treatment

Two stage vaginoplasty in the form of drainage of the haematocolpos in one operation followed by another operation to re-sect the septum is the classic treatment option[7]. Single stage vaginoplasty, advocated in our case, in the form of drainage of the collected blood, complete septum resection followed by suturing of the lateral vaginal wall was proposed to be a suitable alternative to the two stage procedures without any complications. However, postoperative stenosis, recurrence of haematometra[8] and infection are significant possibilities necessitating a second operation. To overcome this possibility, different treatment modalities have been tried to reduce the need of a second operation and minimize the risk of postoperative re -obstruction such as the use of vaginal moulds, dilators[9] and coated trachea-bronchial stent.

Conclusion

Ohvira syndrome is an uncommon congenital anomaly with clinical significance; It is typically associated with didelphs uterus with 2 cervices and 2 vaginas, one of which is obstructed. The obstruction usually occurs on the same side as the renal anomaly. We reported a rare congenital anomaly of female reproductive tract of a 17 year old girl, associated with ipsilateral renal agenesis, because female reproductive tract develops at the same time and close to urinary system from the intermediate mesoderm. Imaging particularly MRI, as in this case plays a major role in diagnosis which could be missed clinically. An early correct diagnosis is the goal to relieve the symptom and prevent complications caused by retrograde menstruation which may lead to endometriosis and subsequently to conception abilities; a two step vagino-plasty can be performed to correct the anomaly.

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