Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) Syndrome Associated with Bicornuate Uterus with Recurrent Hematopyo-metra with Reactive Thrombocytosis, A Case Report

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ABSTRACT

The development of the female urinary and genital system is deeply linked at the embryological level. About 10% of infants are born with abnormality of the genitourinary system, and anomalies in one system are often mirrored by anomalies in another system. The OHVIRA syndrome is a rare set of anomalies affecting the female urogenital tract. The syndrome consists of obstructed hemivagina with ipsilateral renal agenesis with uterine anomalies. The true incidence of the OHVIRA syndrome is not precisely known, but according to the available literature it is estimated between 0.1-3.5 percent of all Mullerian anomalies. It is mostly reviewed in western literature under pediatric surgery, as the median age of presentation is 12.5 years (birth-25yrs), but in our setup it usually falls into the gynecologists purview as the age of presentation is in early childbearing age. A 22 year old married patient was admitted in our hospital as a referred case of abdominopelvic mass with dysmenorrhea. The patient after complete evaluation was shown to have a uterus bicolliis unicornis, with obstructed hemivagina absent right kidney with unilateral right hydrosalpinx and reactive thrombocytosis. Here, we present the diagnostic modalities that helped us reach the diagnosis of this rare syndrome, its associated features and surgical management of an adult patient in a low cost setting.

Key words: Hemivagina, Ipsilateral Renal Agenesis, Hematopyo-metra, Thrombocytosis.

CASE REPORT

A 22 year old married female non-diabetic, normotensive was referred to us with the chief complaints of amenorrhea 2 months, pain lower abdomen and foul smelling vaginal discharge for 1 month. She was admitted in our hospital on 3rd November 2010. The patient was in her usual state of health 2 months back when she started with pain in her lower abdomen around the expected date of menstruation. She was admitted in our hospital on 3rd November 2010. The patient was in her usual state of health 2 months back when she started with pain in her lower abdomen around the expected date of menstruation. The patient had previous history of dysmenorrhea, so she did not seek immediate medical help, the pain was not relieved with medication and it grew in intensity. The patient consulted a local gynecologist who performed a preliminary pregnancy test which was negative and referred her to our center as a case of pelvic mass with pain. The patient also noticed a foul smelling vaginal discharge which was dark in colour. As per history the patient reached menarche at the age of 14 years. Her cycles were irregular since menarche with scanty flow. Cycles were associated with pain lower abdomen which was colicky in nature, radiating to her back and was relieved with medication. The patient is a nulligravida with married life of 11 months, non-smoker and non-alcoholic with positive history of constipation and dyspareunia. Other medical and surgical history was not significant.

On examination, the patient was conscious, cooperative, well oriented to time, space and person. Her vitals were maintained except for a temperature of 99 °F. She had pallor, chest and CVS examination was unremarkable. Per abdomen the patient had an 18 weeks mass arising out of her pelvis which was dull on percussion, non-tender, tense, symmetrical and its lower margins could not be made out. Per speculum the patient had foul smelling thick blackish discharge and the cervix could not be identified. Per vaginum
the exact uterus size could not be appreciated, there was bogginess in the anterior vaginal wall.

A working diagnosis of Hematopyometra was made and the patient’s investigations were sent. The patient was put on broad-spectrum antibiotics and was given two packed cell transfusions. Laboratory findings revealed hemoglobin of 7g/dl, TLC 12,000/mm3. Trans-abdominal sonography demonstrated Bicornuate uterus with Pyometra and right Adenexal mass with absent right Kidney.

An MRI and CT abdomen was planned to delineate the anatomy in greater detail. MRI using T1 and T2 weighted axial, coronal and sagittal images revealed Bicornuate uterus with right Hydrosalpinx with a non-communicating right horn with Hematocolpos on the left side. (Picture 1, 2 & 3) The left horn was seen communicating with the common cervical canal. Bilateral multiple follicular cysts were present in both the ovaries.

The patient was then planned for laprotomy and as is routine, the patient was subjected to detailed pre-anesthetic investigations which revealed that the patient had a platelet count of 7lac/mm3, other investigations including renal function tests were within normal limits. The patient continued on antibiotics and bone marrow aspiration was done. The bone marrow cytology revealed mild Granulocytic Hyperplasia with Reactive Thrombocytosis with Normoblastic Erythropoiesis.

The patient was continued on antibiotics. The patient’s platelets showed a decreasing trend and finally the patient was fit for surgery. Before laprotomy, examination under anesthesia was performed. A transverse vaginal septum was found which was excised. A single cervix was identified through which uterine cavity was sounded. The left cavity was found communicating with the cervical canal.
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Intraoperatively a **rectovesical ligament** was found, separating the two cornua of the uterus, which was excised. (Picture 4 & 5) To correlate the findings of MRI and **Ultrasound** a vertical incision was given on the anterior surface of right cornua. The findings of imaging studies were confirmed and non-communicating cornua was seen on the right side with **myometrial hyperplasia**. The horn was then excised and **ipsilateral salpingectomy** carried out in view of **hydrosalphinx**. **Ovarian drilling** was done in view of the multiple follicular ovarian cysts and keeping in mind her fertility status. An intraabdominal drain was placed, which was removed on the fifth post-operative day. Postoperatively the patient did well on antibiotics and received one more packed cell transfusion. The stitches were removed on the eighth postoperative day.

The patient was discharged on oral progesterone (Norethisterone) to control her cycles and was asked to come for follow up. On follow up the patient came with withdrawal bleeding 1 week after stopping of progesterone with no dysmenorrhea. Currently the patient is doing well and hopeful of a pregnancy.

### DISCUSSION

The **OHVIRA** syndrome was first described in 1922 as **Herlyn-Werner-Wunderlich Syndrome**. In one study it was quoted that 300 cases of **OHVIRA** syndrome have been reported since 1922, emphasizing the need to report adequately diagnosed and managed cases.

**OHVIRA** syndrome classically occurs in the setting of **Uterine Didelphys** or, more rarely a **Septate uterus**. Renal agenesis is the most widely reported urological anomaly. **Uterus Didelphys** results from fusion failure of **Mullerian ducts**. A good working classification for Mullerian anomalies was published by Shivaji B. Mane Pankaj Shastri. It aims at making the surgical management of such cases easier. To our discretion, this classification is better than the other often quoted academic ones, as it points towards the surgical management of such cases.

The presentation is around the age of menarche in the western setup, but literature shows a whole spectrum of presentations from **Amenorrhea**, **Infertility**, and **Chronic per vaginal discharge** to **pregnancy complications**. With

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<th>Group</th>
<th>Description</th>
<th>Anomaly</th>
<th>Procedure</th>
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<tr>
<td>IA</td>
<td>Complete absence of uterus and vagina</td>
<td>Mayer–Rokitansky–Kuster–Hauser (MRKH) syndrome</td>
<td>Bowel vaginoplasty</td>
</tr>
<tr>
<td>IB</td>
<td>Absence of lower third of vagina</td>
<td>Partial Mullerian agenesis</td>
<td>Pull-through vaginoplasty and bowel vaginoplasty</td>
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<td>2</td>
<td>Asymmetric obstructed duplicated uterus, cervix and vagina</td>
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<td>Combined laparotomy and hysteroscopic treatment</td>
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<tr>
<td>3</td>
<td>Asymmetric obstructed duplicated uterus and cervix with single vagina</td>
<td>Cervical atresia</td>
<td>Combined hysteroscopic and laparoscopic treatment</td>
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<td>4</td>
<td>Active Mullerian remnant in ARM patient</td>
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the advent of various imaging modalities the diagnosis has become simplified but the discretion of the treating surgeon is still the most reliable tool.

The surgical management currently favours a single stage procedure with fertility conserving methods. Literature exists in favour of novel percutaneous procedures such as *Percutaneous obstructed hemivagina access (POHVA)* to the difficult obstructed hemivagina in the *OHVIRA* syndrome.

Our setup limits the options like *POHVA* which we can offer to these patients. However the end point that the patient leads a relatively morbidity free life is ensured with our kind of management. The optimal approach is still adolescent screening for *mullerian defects*. But, this approach can only be feasible when the adolescent clinics shall be operational in our country. Suad Gholoum, Promod S. puligandla et al in one of the largest reviews of *OHVIRA/HWWS* reported the median age of presentation as 13 years.

Our patient presented at 22 years of age and her disease had advanced to *hydrosalpinx* and may have landed her in *Septicemia* if not intercepted. Her other complaint was naturally a concern for her fertility, and it has been reported by Sarac A, Demir MK that *HWWS* is a rare cause of infertility. For the radiological diagnosis, a multimodality approach seems to work best. Boram Han, MD, Christopher N. Herndon, MD et al published the first ever case report where transabdominal ultrasound, CECT and MRI were performed in a single patient. Similar studies were done by us and helped lead us to the final diagnosis. However, in our opinion the variants of *mullerian defects* are many, as is exemplified by the presence of a non-communicating horn of the *bicorneuate* uterus in this case. Thus, the discretion of the treating physician is very important to reach at the final diagnosis. The predilection of right sided anomalies has also been reported and studies done on animal models attribute this to the right side being more susceptible to hypoxic damage (perhaps the precocious development of *mitochondria* on the left side of the embryo results in higher energy reserves and less tissue damage due to hypoxia). Laparoscopy has been mentioned in the literature as the procedure of choice when surgery is being considered. But as stated earlier, the low cost setting limited our access to operative laparoscopy. Open surgery is also justified in our case in view of the extent of disease. The combined abdomino-perineal approach aims at preserving the patient’s fertility. Similar conservative approach has been advocated by TSUCHIYA TAKEHIKO (Teikyo Univ, Mizonokuchi Hosp) et al where they recommend fertility preservation whenever possible. The outcomes of pregnancy in these patients reveal 87% go on to have a successful pregnancy, while abortions occur in 23% of the patients, 15% have preterm births, and 62% have full-term pregnancies and uncomplicated deliveries.

**REFERENCES**