ejpmr, 2023, 10(10), 361-364

SJIF Impact Factor 6.222

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Study

ISSN 2394-3211

EJPMR

UTERINE DIDELPHYS WITH OBSTRUCTED HEMIVAGINA AND IPSILATERAL RENAL ANOMALY: OHVIRA SYNDROME – A CASE REPORT

Shamrao Ramji Wakode and Sakshi Pramod Sharma*

Department of Obstetrics and Gynaecology, S.C.G.M.C., Nanded (MH), India.

***Corresponding Author: Sakshi Pramod Sharma**

Department of Obstetrics and Gynaecology, S.C.G.M.C., Nanded (MH), India.

Article Received on 04/08/2025 **Article Revised on 25/08/2023** **Article Accepted on 15/09/2023**

ABSTRACT

Introduction: OHVIRA syndrome is a rare abnormality of the urogenital tract characterized by the triad of obstructed hemivagina, uterine didelphys, and ipsilateral renal agenesis, which is generally diagnosed after menarche. It often creates a diagnostic dilemma as most of the patients with OHVIRA syndrome are diagnosed late due to the rare incidence and the non- specific clinical presentation. Another reason for delay in diagnosing this syndrome are due to usual onset of puberty and menstruation as there are different phenotypic variables. **Case presentation:** A 13-year-old female approached with complain of dull pain in lower abdomen since last 7 to 8 months. Patient gave history of menarche 10 months ago and her menstrual cycle was irregular and was associated with progressively increasing dysmenorrhea. Her USG abdomen and pelvis was suggestive of haematocolpos with agenesis of right kidney. CT scan report showed uterus didelphys with right renal agenesis and obstructed right hemivagina secondary to vagina. Vertical incision was made over the right vaginal section and the dark collected blood was drained. Post operatively patient was stable and repeat USG showed reduction in size of hematocolpos. Patient was discharged on day 5 after the procedure and was counselled regarding the need for septoplasty and further implications of her diagnosis. **Discussion:** The causes of OHVIRA syndrome are not fully understood so far, but it is considered as a result of multiple factors, including genetics, environment, and endocrine influences. When patients approach with dysplastic or absent kidneys, an attempt to diagnose uterovaginal anomalies must be made to allow early recognition and management of this rare entity. It must be considered as a differential diagnosis in women of any age with paramesonephric duct anomalies, pelvic pain, dysmenorrhea, and presence of masses in the genital tract. **Conclusion:** It's early diagnosis and timely treatment considerably improves the quality of life of these patients by reducing the severity of symptoms, decreasing the incidence of complications, and improving obstetric prognosis.

KEYWORDS: OHVARIA Syndrome, Uterine didelphys, obstructed hemivagina, ipsilateral renal anomaly.

INTRODUCTION

OHVIRA syndrome or Herlyn-Werner-Wunderlich syndrome is a rare congenital anomaly comprising of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis.[1] In 1922 Purslow, first reported this syndrome.[2] Thereafter, in 1971, Herlyn and Werner reported a case of renal agenesis with a blind hemivagina, and Wunderlich described the association of renal aplasia, a bicornuate uterus with a simple vagina and an isolated hematocervix in 1976.[3]

This syndrome is characterized by the triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal anomaly.[4] OHVIRA syndrome is mostly misdiagnosed and high index of suspicion is required in patients with Mullerian duct anomalies.[5] Mullerian duct anomalies have an incidence of 2–3%. While OHVIRA constitutes 0.16–10% of these.[6] Patients usually approach with indications such as, progressive dysmenorrhea and lower abdominal pain, however, other common indications like urinary retention and pelvic mass are also noticed.[7]

Most of the patients with OHVIRA syndrome are diagnosed late due to the rare incidence and the non-specific clinical presentation. Another reasons of delays in diagnosing this syndrome are due to usual onset of puberty and menstruation as there are a different phenotypic variable.

Furthermore, the menstrual flow from the unobstructed hemivagina gives the appearance of usual menses. Therefore, accurate diagnosis and surgical treatment can be delayed for several months or even years.[8] Hence, when patients approach with dysplastic or absent kidneys, an attempt to diagnose uterovaginal anomalies must be made to allow early recognition and management. However, because the presentation is variable prompt recognition can be difficult. Though

renal agenesis is the classic presentation, other anomalies include renal dysplasia and ectopic ureters.[7]

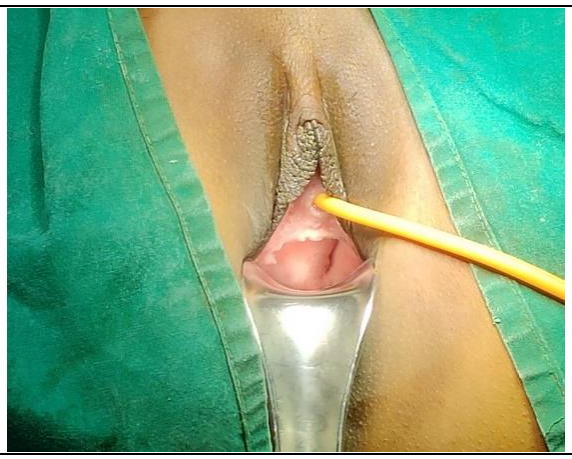
In patients who are suffering by OHVIRA, ovarian function is normal. The impact of this disorder, on fertility is not well known so far, but complications may arise due to endometriosis, developing pelvic abscess, and pelvic adhesions.[9] following report presents a case of this syndrome in a 13 year old female.

CASE REPORT

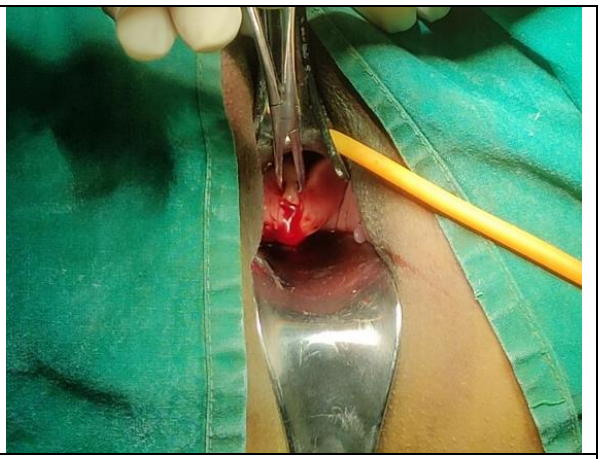
A 13-year-old female, approached to outpatient department with complain of dull pain in lower abdomen since last 7 months. Patient was apparently alright 7 to 8 months ago when she started experiencing a dull pain in lower abdomen which was gradual in onset and progressive in nature. Over the period of time it increased, hampering her daily activities. She had no history of burning micturition, diarrhoea, abdominal trauma, outside food consumption. She gave history of menarche 10 months ago. Her menstrual cycle was irregular and consisting of pain and clot passage. She was unmarried and was given analgesic in past as medical management but her symptoms were not relived.

Patient had no other significant past history. On examination patient was conscious, oriented and vitally stable. Secondary sexual characteristics were well developed and her USG abdomen and pelvis was suggestive of haematocolpos with agenesis of right kidney. As per the laboratory examination of patient, she had haemoglobin 11.2g/dl; total leukocyte counts 13000/mm^3; neutrophil 54%; lymphocytes 34%; eosinophil 1%; basophil 0%. Her blood urea was 18mg/dl; serum creatinine 0.7mg/dl; serum total bilirubin 0.6mg/dl; SGOT 24 IU/L; SGPT 16 IU/L. Blood group B+ve and ECG readings showed normal findings.

She was further posted for CT scan which was the leading evidence for diagnosis of OHVIRA Syndrome i.e. she had uterus didelphys with right renal agenesis and obstructed right hemivagina secondary to vagina septum with haematocolpos. Patient was posted for examination under anaesthesia after consent of her guardians. Intraoperative findings were similar to the radiological findings and a vertical incision was made over the right vaginal section and the dark collected blood was drained. Post operatively patient was stable and repeat USG showed reduction in size of hematocolpos. The postoperative management was urinary catheterisation for 24 hours, vaginal tampon for 24 hours, maintaining local hygiene and antibiotic regimen of injection Taxim 1gm intravenously twice a day along with injection Metronidazole 100 cc thrice a day. Her recovery was supplemented by giving multivitamins. Patient was discharged on day 5 after the procedure and was counselled regarding the further implications of her diagnosis, with follow up appointment for septoplasty.

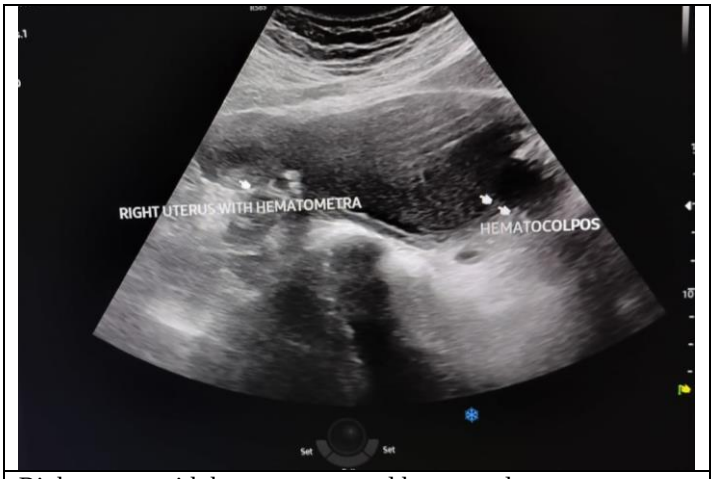


1. obstructed right hemivagina secondary to vagina septum.



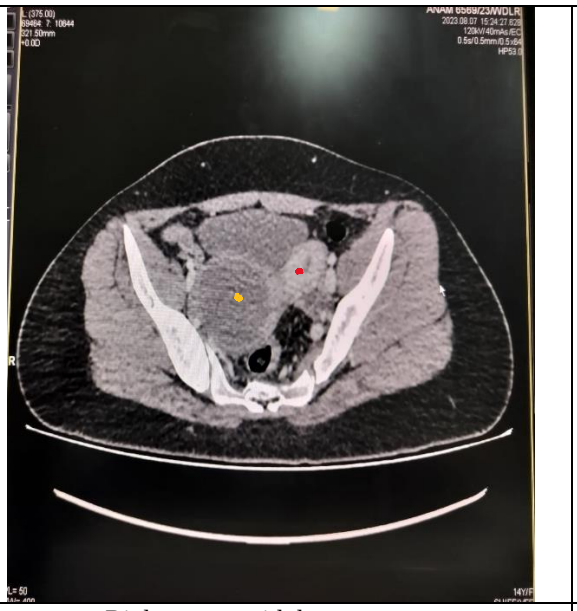
2. Hematocolpos drained by taking vertical incision on the bulging vaginal septum.

Fig. - 1 & 2: Intraoperative images.

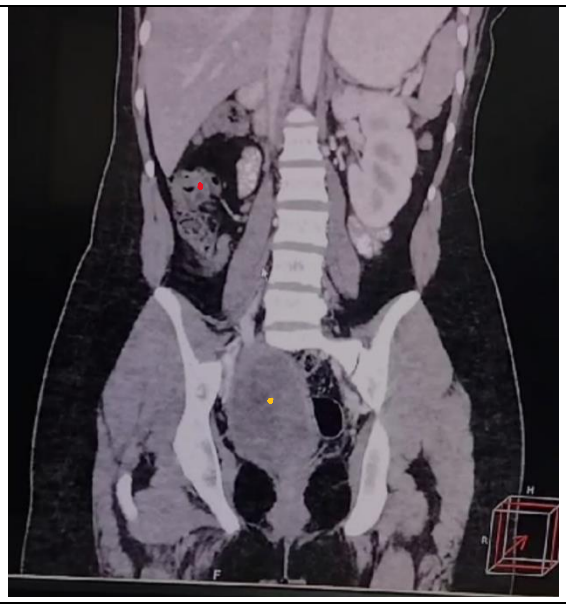


Right uterus with hematometra and hematocolpos

Fig. - 3: Ultrasound image of OHVIRA syndrome.



Right uterus with hematometra
Left uterus



Right uterus with hematometra
Right renal agenesis

Fig. – 4 & 5: CT Scan images of OHVARIA Syndrome.

DISCUSSION

Female genitourinary tract malformations are common: they are found in approximately 3–4% of the general population and in 15% of patients experiencing recurrent miscarriage.[10] In contrast OHVIRA syndrome characteristically occurs in the setting of uterine didelphys or rarely a septate uterus. Most widely reported urological anomaly is the Renal agenesis. Uterus didelphys results from fusion failure of Mullerian ducts.

According to Fedele *et al.*, in most cases (72.4%), OHVARIA syndrome presents with a classic variant involving a uterus didelphys, unilateral genital obstruction, and ipsilateral renal abnormality. The causes of OHVIRA syndrome is not fully understood so far, but it is considered as a result of multiple factors, including genetics, environment, and endocrine influences. The syndrome arises due to an arrest in the development of the mullerian and mesonephric ducts during week 8 of pregnancy. Failure of the Mullerian ducts to fuse, results in uterine didelphys, while the ipsilateral kidney agenesis results from initial high-level obstruction at the pelvic ureteric junction.[11]

Diagnosis of OHVIRA syndrome, is usually based on abdominal pain after menarche due to increased blood retention caused by obstructed hemivagina. However, the demarcation between the dilated cervix and vagina is normally unclear, which makes diagnosis of OHVIRA syndrome problematic. In several cases this syndrome remains unrecognized because of normal menstruation through the unobstructed hemivagina. Such cases are treated as simple dysmenorrhea and overlooked in routine practice. However, if this syndrome not treated in time, retrograde menstruation may result in tubal

hematoma and endometriosis in the abdomen, subsequently may lead to infertility.[12] Therefore, timely surgery of the vaginal septum is preferable. A worthy operational classification for Mullerian anomalies has been reported by Mane et. al., 2010.[13] This classification is focused at making the surgical management of such cases easier.

CONCLUSION

OHVARIA Syndrome must be considered as a differential diagnosis in women of any age with paramesonephric duct anomalies, pelvic pain, dysmenorrhea, and presence of masses in the genital tract. Early diagnosis and timely treatment considerably improve the quality of life of these patients by reducing the severity of symptoms, decreasing the incidence of complications, and improving obstetric prognosis.

REFERENCES

1. Moufawad G, Ayed A and Sleiman Z. Reconsidering the best minimally invasive approach for patients with herlyn-wernerwunderlich syndrome: Should we push the frontiers for a better outcome? *Front. Surg, 2023; 10: 1158753.*
2. Purslow, C.E. A case of unilateral haematocolps, haematometria, and haematosalpinx. J. Obstet. Gynaecol. Br. Emp, 1992; 29: 643. [CrossRef]
3. Sambedna, Amit Kumar, Mukta Agarwal, Nimisha Agrawal, Sudwita Sinha. OHVIRA syndrome with Orofacial clefts: a rare association. *Int J Reprod Contracept Obstet Gynecol, 2020 Mar; 9(3): 1306-1309.*
4. Utama Bobby Indra. Obstructed Hemivagina and Ipsilateral Renal Anomaly – A case report. Mecedonian Journal of Medical Sciences, 2022; 2(9C): 15-18.
5. Yadav P, Prashanth V, Hazarika P. A rare case: OHVIRA syndrome (obstructed hemivagina and ipsilateral renal anomaly) with unicornuate uterus. J. Evid. Based Med. Healthc, 2019; 6(10): 819-821.
6. Mandurkar Suyash and Guruprasad Pednekar. A case report on OHVIRA syndrome managed in Goa medical college. International Journal of Clinical Obstetrics and Gynaecology, 2019; 3(5): 283-285.
7. Jha Swati and Shahram Abdi. Diverse Presentations of OHVIRA Syndrome: A Case Series. Clinical Pediatrics, 2021; Vol. 60(1): 6–8.
8. Nigam, A., C. Raghunandan, R. Yadav, S. Tomer, R. Anand OHVIRA syndrome: rare cause of chronic vaginal discharge in an unmarried female congen it Anom, 2011; 51(3): 153–155. http://dx.doi.org/10.1111/ j.1741-4520.2010.00293.x Kyoto.
9. Muraoka A, Tsuda H, Kotani T, Kikkawa F. Severe hemoperitoneum during pregnancy with obstructed hemivagina and ipsilateral renal anomaly syndrome: a case report. *J Reprod Med*, 2016; 61: 290-294.
10. Ryosuke Arakaki, Kanako Yoshida, Junki Imaizumi, Takashi Kaji, Takeshi Kato, Takeshi Iwasa. Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome: A case report. International Journal of Surgery Case Reports, 2023; 107: 108368.
11. Mohamed Amin Elgohary, Rushikesh Naik, Mohamed Elkafafi, Hussein Hamed, Yaser Ali. Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome: A case report. Journal of Pediatric Surgery Case Reports, 2023; 95: 102662.
12. Guducu N., G. Gonenç, H. Isçi, A.B. Yigiter, I. Dunder, Herlyn-Werner-Wunderlich syndrome—timely diagnosis is important to preserve fertility, J. Pediatr. Adolesc. Gynecol, 2012; 25: e111–e112.
13. Mane Shivaji B., Pankaj Shastri, Nitin P. Dhende, Abu Obaidah and Himanshu Acharya, et al. Our 10-year experience of variable mullerian anomalies and its management, Pedatric surgery International, 2010; 26(8): 795-800.