International Journal of Pharmaceutical Research and Applications
Volume 9, Issue 4 July-Aug 2024, pp: 875-878 www.ijprajournal.com ISSN: 2456-4494

Earlydiagnosis and management Ofohvira Syndrome in an Adolescent: a Casereport

Dr Siriginedi Naga Spandana Priya[1*], Thrishnitha Dasari[2*], Kata Akshitha3*, Garlapati Rasagnya[4]

1.AssistantProfessor, Sri Venkateshwara Collegeof Pharmacy, Madhapur, Hyderabad
2. Pharm D Fourth Year, Sri Venkateswara Collegeof Pharmacy, Madhapur, Hyderabad
3. Pharm D Fourth Year,Sri Venkateswara Collegeof Pharmacy, Madhapur, Hyderabad
4. Pharm D Fourth Year, Sri Venkateswara Collegeof Pharmacy, Madhapur, Hyderabad

Date of Submission: 25-07-2024 Date of Acceptance: 05-08-2024

ABSTRACT:
The Ohvira syndrome also called as Herlyn-Werner-Wunderlich syndrome (HWWS), a very rare congenital anomaly of the urogenital tract involving both Müllerian ducts and Wolffian ducts.It is characterized by a triad of uterine didelphys, ipsilateral renal agenesis and obstructed hemivagina .This case study presents a 10-year old female who presented with severe left abdominal pain since 2-3days.Despite regular menstrual cycle , experienced decreased flow in last two cycles. On Hospitalization, Clinical examination revealed tender left side of abdomen. Diagnostic imaging via Ultrasonography confirmed Uterine didelphys, obstructed hemivagina Vagina, Ipsilateral renal agenesis and MRI showed Unilateral kidney (right side) Substantiating the diagnosis OHvira Syndrome.prompt surgical intervention was crucial in order to prevent further complications includes infertility, with the patient undergoing Hysterolaprascopy and Transverse vaginal septal resection. Intraoperative findings of bluish bulge (hematocolpos) near the perineum transverse vaginal septum. Postoperatively, the patient received antibiotics, analgesics facilitating a smooth recovery and discharge within two days. Overall, this study emphasizes the necessity of a comprehensive approach that includes early diagnosis, surgical expertise, and diligent postoperative care to effectively manage this time-sensitive condition.

KEYWORDS: OHvira syndrome, Herlyn -Werner -Wunderlich Syndrome, uterine didelphys, Ipsilateral renal agenesis, obstructed hemivagina, Hematocolpos

I. INTRODUCTION:

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome is a rare anomaly of Müllerian ducts. This syndrome also called as Herlyn-Werner Wunderlich(HWW) syndrome. It is an uncommon congenital anomaly of female urogenital tract which involves mesonephric (wolffian) and paramesonephric (mullerian) ducts, characterised by the triad of uterine didelphys, obstructed hemivagina, unilateral renal agenesis was first reported in 1922. [1,2]
The Wolffian ducts induce the normal development of the Müllerian ducts in addition to giving origin to the kidneys. Abnormal development of the Wolffian ducts leads to the unilateral renal agenesis and imperforated hemivagina associ ated with OHVIRA syndrome[3]Ipsilateral renal agenesis with partial vaginal septum on the same side and didelphys uterus is explained by embryologic arrest at the 8th week of gestation affecting simultaneously the mullerian and metanephric ducts[4]

The true incidence of OHVIRA syndrome is not precisely known, but the estimated overall incidence of Müllerian duct anomalies can vary from 0.1% to 3.8%.[1]

This syndrome, typically diagnosed in adolescent females around puberty (after menarche) which presents with symptoms such as pelvic pain, dysmenorrhea and the formation of a pelvic mass due to accumulation of menstrual blood in the obstructed hemivagina. When untreated, this may result in severe complications such as pyohematocolpos, pyosalpinx, or pelvic peritonitis, urinary obstruction, and long-term complications, such as endometriosis, pelvic adhesions, and increased risk of abortion or infertility.[5]

International Journal of Pharmaceutical Research and Applications
Volume 9, Issue 4 July-Aug 2024, pp: 875-878 www.ijprajournal.com ISSN: 2456-4494

Understanding the etiology and pathophysiology of OHvira syndrome reveals insights into its clinical management and outcomes. The precise cause of this condition is unknown, genetic, environmental and endocrine factors include hormonal influence on Müllerian duct development. The Wingless integrated (Wnt) genes have been implicated in Müllerian duct development but its role in this syndrome is unknown. HWWS is characterised by aberrant development of one Müllerian duct and normal development of the other, resulting in a didelphys uterus (two distinct uterine cavities). obstructed hemivagina, A transverse vaginal septum or vaginal atresia (lack of a normal vaginal opening) block one hemivagina,the collection of menstrual blood in the obstructed hemivagina causing hematocolpos, which can result in pelvic pain and mass impact.[6]

The clinical presentations in patients with ohvira syndrome usually present at post menarche with symptoms, dysmenorrhea,pelvic or vaginal mass. The collection of menstrual blood in the obstructed hemivagina due to this obstruction causes hematocolpos, rarely hematosalpinx..Pelvic pain is the most common presenting symptom (90%) followed by an abdominal mass (40%), abdominal pain,Patients can also present at a later age with abnormal and foul-smelling vaginal discharge due to pyocolpos, vomiting, fever , Acute urinary retention(AUR).furthermore blood reflux into abdominal cavity may result in endometriosis which is further complicated by pelvic adhesions and increased risk of abortion and infertility.[7]
initial clinical diagnosis is incorrect in majority of the cases as it's incidence is rare and misleading signs and symptoms includes pelvic pain, recurrent urinary tract infections, irregular menstrual cycles,vaginal discharge, Dyspareunia, Hematocolpos.[8]

The Diagnosis of OHVIRA syndrome requires a multimodal approach, which includes a detailed history of patient, meticulous examination, and appropriate imaging studies. It is diagnosed late due to its rare incidence and non-specific clinical manifestations and also the menstrual flow from the unobstructed hemivagina gives the appearance of normal menstrual flow[9]

Diagnostic tools include Ultrasound, Ultrasonography (USG) which is frequently the initial imaging modality to evaluate suspected mullerian duct anomalies, require expert interpretation to confirm the diagnosis and guide timely surgical intervention crucial within the narrow window of salvageability,

MRI (magnetic resonance imaging), the gold standard available to provide detailed imaging of pelvic anatomy and in confirming presence of uterine didelphys, hemivagina and associated hematocolpos.it has 100%accuracy in detecting mullerian duct anomalies.[10,11]Other diagnostic procedures include CT scan, laparoscopy can help in confirming the diagnosis and detection, HSG (hysterosalpingography) was used in the past quite often. Obstructed anomalies, if not treated timely and appropriately may lead to chronic complications and detrimental psychological effect, thereby diminishing a woman's quality of life. Awareness of these anomalies, having familiarity with their symptoms, and maintaining a high level of suspicion can facilitate early diagnosis and timely treatment, helping to preserve reproductive potential.[12]

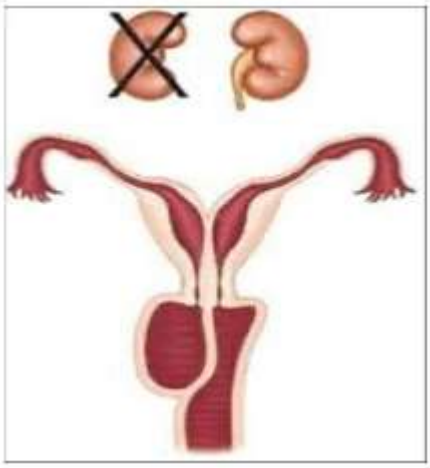

Fig:1: pictorial presentation of triad of Herlyn-Wenrer Wunderlich Or OHVIRA syndrome (Obstructed Hemivagina, Ipsilateral renal agenesis with uterine didelphys)

The management of OHvira syndrome, comprises of either resection or excision of the vaginal septum or drainage of hematocolpos.[13,14].In this case, the patient underwent Hysterolaproscopy and transverse vaginal septal resection. The Hysteroscopy, enables direct visualisation and treatment of the uterine cavity and the obstructed hemivagina. Laparoscopy, provides a view of pelvic organs and helps to evaluate and manage associated anomalies. The delays in the management can result in complications such as endometriosis and infertility.

We are reporting a case of 10-yearold female who came to the emergency department

International Journal of Pharmaceutical Research and Applications
Volume 9, Issue 4 July-Aug 2024, pp: 875-878 www.ijprajournal.com ISSN: 2456-4494

with complaints of severe abdominal pain since 2-3 days.

CASE STUDY – 1:

A 10-Yr-old female presented to the emergency department with chief complaints of severe pain in the left lower abdomen, pelvic region since 2-3days.There was no history of fever, dysmenorrhea, or dyspareunia. The patient attained her menarche 5 months back. she has been having regular menstrual cycles every 28 days and lasting for 3–4 days, secondary sexual characters were developed as per the age. But decreased flow in last two cycles and painless. She denied any dysmenorrhea or menorrhagia. The patient had no comorbidities, was not on any medications, had no drug allergies, and had no prior surgical history. Her family History found to have similar diagnosis in paternal Aunt.No history of cancer in family.

II. DISCUSSION:

This case report details the presentation, diagnosis and treatment of 10-yearold female who was admitted to the hospital with complaints of severe pain in the left lower abdomen, pelvic region since 2-3 days. The patient also experienced the decreased menstrual flow in last two cycles, painless. The initial examination revealed the P/A: Tenderness, with no apparent increase in temperature and the rest of systemic examination was unremarkable.

On Laboratory examination, the general condition was fair,patient blood group found to be B positive, BMI was16.7kg/m2 and her vitals were as follows BP:100/70 mmhg, PR:82b/min, Hb: 14.1%gm%platelet count: 304x10 3/mm3, RBS: 107mg%., HCV – negative, slight increase in PT-14.1 and INR- 1.05.

An Ultrasonography (USG) showed vagina hemivagina 68x19mm with transverse septum 1.2cm from perineum, ipsilateral renal agenesis, uterine didelphys, both ovaries are normal. A magnetic resonance imaging (MRI) scan showed unilateral kidney. The diagnosis of left-sided obstructed hemivagina with obstructed blood collection and associated left ipsilateral renal agenesis (OHVIRA syndrome) with uterine didelphys was confirmed by Ultrasonography. The consent was obtained for Hysterolaparoscopy and transverse vaginal septal resection. During procedure, the laparoscopy findings are uterus Didelphys and bilateral fallopian tubes with normal ovaries and the hysteroscopy, right hemivagina seen and right cervix visualised whereas left Hemivagina and left cervix not visualised, A bluish bulge (hematocolpos) noted near the Perineum-transverse vaginal septum. No undue bleeding noted, vaginal pack was placed in situ.

Based on the patient's clinical presentation and diagnostic criteria,patient is provisionally diagnosed as OHVIRA syndrome(obstructed hemivagina with Ipsilateral renal agenesis).

Post- operatively, the patient received IV antibiotics (IV Taxim -O 100mg BD), pain management(IV paracetamol 100mg BD)(IV UKET 30mg SOS). The patient recovered well after surgery and no complications. The patient was discharged on the second postoperative day with the following medications:

Taxim-O 100mg twice a day after food for 5 days, pantop 20mg once a day before breakfast for 5 days, paracetamol 500mg twice a day after food for 3 days and Premarin vaginal cream 0.625mg 1 fingertip once a day for 4 weeks. She was advised to follow up at the Gynaecology department after 1 week and vaginal pack to be removed after 1 day.

III. CONCLUSION:

This case of a 10-year-oldfemale with complaints of pain in left lower abdomen since 2-3 days which emphasises the essential need for quick diagnosis and treatment. Early recognition, timely identification, rapid assessment, and promptsurgical intervention are vital for maintaining normal function and avoiding complications.The patient's successful recovery and discharge within three days highlight the effectiveness of timely surgery and appropriate postoperative care. Ongoing follow-up is crucial to monitor for any complications and ensure the overall health of the patient. This is the rare presentation which highlights the importance of educating individuals about the complications of OH vira which includes vaginal discharge, dysmenorrhea, abdominal pain, pelvic infections, menstrual irregularities, infertility, endometriosis. Early presentation to the emergency department can significantly enhance the outcomes of individuals. Hence, awareness and early diagnosis with adequate and timely treatment and psychological counselling can prevent further complications and improves the quality of life.

REFERENCES:

[1]. Han BH, Park SB, Lee YJ, Lee KS, Lee YK. 2013. Uterus didelphys with blind hemivagina and ipsilateral renal agenesis

International Journal of Pharmaceutical Research and Applications
Volume 9, Issue 4 July-Aug 2024, pp: 875-878 www.ijprajournal.com ISSN: 2456-4494

(Herlyn–Werner–Wunderlich syndrome) suspected on the presence of hydrocolpos on prenatal sonography. Journal of Clinical Ultrasound: JCU 41:380–382.

[2]. R Del Vescovo, S Battisti, V Di Paola, CL Piccolo, RL Cazzato, I Sansoni, et al. Herlyn-Werner-Wunderlich syndrome: MRI findings, radiological guide (two cases andliterature review), and differential diagnosis BMC Med Imaging. 2012 Mar 9;12:4.

[3]. Gungor Ugurlucan F, Bastu E, Gulsen G, Kurek Eken M, Akhan SE. OHVIRA syndrome presenting with acute abdomen: a case report and review of the literature. Clin Imaging. 2014;38: 357–359. doi: 10.1016/j.clinimag.2013.12.011. pmid:24461469

[4]. Kim TE, Lee GH, Choi YM, Jee BC, Ku SY, Suh CS, et al. Hysteroscopic resection of the vaginal septum in uterus didelphys with obstructed hemivagina: a case report. J Korean Med Sci2007. Aug;22(4):766-769 10.3346/jkms.2007.22.4.766

[5]. Orazi C, Lucchetti MC, Schingo PM, Marchetti P, Ferro F. Herlyn-Werner-Wunderlich syndrome: uterus didelphys, blind hemivagina and ipsilateral renal agenesis. Sonographic and MR findings in 11 cases. Pediatr Radiol. 2007 Jul;37(7):657-65.

[6]. Mullen RD, Behringer RR. Molecular genetics of Müllerian duct formation, regression and differentiation. Sex Dev 2014;8(5):281–296. DOI: 10.1159/000364935.

[7]. Dhar H, Yasser AR, Illham H. Uterusdidelphys with obstructed hemi vagina, ipsilateral renal agenesis and right pyocolpos: a case report. Oman Med J. 2011;26:447-50.

[8]. Arikar II, Harma M, Harma MI, Bayar U, Barut A. Herlyn-Werner-Wunderlich syndrome (uterus Didelphys, blind hemivagina and ipsilateral renal Agenesis): A case report. J Turk Ger Gynecol Assoc 2010;11:107-9.

[9]. David A, Gudi SN, Shankar R. Herlyn–Werner–Wunderlich syndrome: premenarche. JSouth Asian Feder Obst Gynae 2017;9(2):207–210. DOI: 10.5005/jp-journals-10006-1496.

[10]. Braverman P, Breech L. Committee on adolescence. Clinical report—gynaecologic examination for adolescents in the paediatric office setting. Pediatrics 2010;126(3):583– 590. DOI: 10.1542/peds.2010- 1564.

[11]. Carrington BM, Hricak H, Nuruddin RN, et al. Mullerian duct anomalies: MR imaging evaluation. Radiology 1990; 176:715–720. DOI: 10.1148/ radiology.176.3.2202012.

[12]. Zhu L, Chen N, Tong JL, et al. new classification of Herlyn-Werner-Wunderlich syndrome. Chin Med J (Engl) 2015; 128:222–225. DOI: 10.4103/0366 - 6999.149208.

[13]. Jaiprakash T, Saxena RK, Pandey P. Obstructed hemivagina with ipsilateral renal anomaly (OHVIRA) syndrome - a rare congenital anomaly. J Genit Syst Disord. 2013; 2:2.

[14]. Han B, Herndon CN, Rosen MP, Wang ZJ, Daldrup-Link H. Uterine didelphys associated with obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome. Radiol Case Rep. 2010;5:327.