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OHVIRA SYNDROME COMPLICATED WITH UPPER GENITAL TRACT INFECTION: A CASE REPORT

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ABSTRACT

Introduction and importance: Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) is a rare congenital urogenital malformation with uterus didelphys, obstructed hemi-vagina, and renal agenesis. **Case presentation:** we report a 18-year-old female admitted with lower abdominal pain with smelly clotted vaginal progressing in the context of a complicated upper genital tract infection in whom MRI examination confirmed the diagnosis of OHVIRA syndrome. The early diagnosis and management help prevent complications such as upper genital tract infection and pelvic peritonitis. **Clinical discussion:** The most frequent symptom of OHVIRA syndrome involves isolated hematocolpos. MRI is the recommended diagnostic imaging method. Routine laparoscopy is not required. Vaginal septectomy is an effective surgical technique for relieving symptoms and preventing potential complications in the majority of OHVIRA patients. **Conclusion:** OHVIRA syndrome must be suspected in any patient, especially adolescents, presenting with non-specific pelvic pain.

KEYWORDS: OHVIRA syndrome – Müllerian congenital malformation – Didelphic uterus – Hematocolpos – Renal agenesis.

INTRODUCTION AND IMPORTANCE

Obstructed hemivagina ipsilateral renal agenesis (OHVIRA) syndrome is a complex congenital Müllerian malformation. Clinical symptoms of this syndrome are characterized by didelphic uterus, obstructed hemivagina and ipsilateral renal agenesis. This involves abnormal development of the Müllerian and Wolffian ducts during genital formation in female embryos.[1,2] The estimated incidence of OHVIRA syndrome amounts to 0.16%-10% of all cases of Müllerian malformations.[3] We report a rare case of OHVIRA syndrome in a 18 years old girl who presented with vague abdominal pain accompanied by a smelly clotted discharge with vomiting and episode of fever. Our case highlights the importance of early diagnosis and treatment to mitigate the complications of this syndrome that can lead to upper genital tract infection or, in more severe cases, pelvic peritonitis, especially if neglected or if there is a delay in diagnosis.

CASE PRESENTATION

A 18-year-old nulligravida girl presented with lower abdominal pain with associated smelly clotted vaginal discharge of 3-months duration with two episode of fever at 39 degrees. The patient reports having irregular menstrual cycles with moderate pelvic pain during each cycle. No prior use of oral contraception. Menarche occurred at the age of 15. She had no notable medical or surgical history. She reported dyspareunia without post-coital bleeding or associated urinary symptoms. Our initial inspection and clinical examination found GCS 15/15 with a blood pressure of 100/60 mmHg, including tachycardia at 100 bpm, temperature at 39degrees. Her physical examination revealed a mild lower abdominal tenderness without rebound or guarding. The vaginal examination found a bulge in the right upper portion of the vagina with smelly purulent leukorrhea. Complete blood count was done, finding a haemoglobin value of 12.5 gm/dl, leukocytes 20.000/ µl, C-reactive protein (CRP) 325 mg/L. Also a vaginal sample was taken and the culture result was positive for staphylococcus saprophyticus sensitive to amoxicillin and clavulanic acid resistant to ceftriaxone and doxycycline. The serologies for chlamydia and gonococcus were negative. Additionally, the cytobacterial study of the urine was sterile. An ultrasound (US) showed a large mass in the pelvis measuring 70x60mm suggesting obstructed hemivagina with hematocolpos (Figure 1) with left pyosalpinx (Figure 2). A magnetic resonance imaging (MRI) scan confirmed the diagnosis of right sided OHVIRA syndrome. The patient had uterus didelphys and a right-sided obstructed hemivagina with hematocolpos (Figure 3) and associated right ipsilateral renal agenesis. The patient was placed on intravenous amoxicillin and clavulanic acid at 1 gram every 8 hours,

metronidazole at 500 mg every 12 hours and gentamicin at 320 mg every 24 hours, until achieving 48 hours of fever-free status. A 48 hour follow-up blood count was conducted, showing a leukocytes count of 14.000/µl and a CRP level of 140 mg/L. After achieving fever-free status and under anesthesia, a vaginal examination revealed a bulge in the right upper side of the vagina caused by accumulated menstrual blood (hematocolpos). The vaginal septum was excised, the hematocolpos was drained, and a saline vaginal wash was performed. A 14 Fr Foley catheter was left in place for 24 hours until the bloody discharge had fully cleared. The patient was discharged on the third postoperative day on oral amoxicillin and clavulanic acid at a dose of 1gr every 8 hours, and metronidazole at a dose of 500 mg every 12 hours for a total duration of 21 days. Recovery was uncomplicated. Follow-up after 4 weeks showed no abdominal pain with no abnormal vaginal discharge with one normal menstrual cycle. A follow-up vaginal sample was taken and returned sterile. An US was performed too showing no residual collection. The patient was scheduled for a follow-up twice a year for a total duration of two years.

Clinical Discussion

OHVIRA syndrome, known as Herlyn-Werner-Wunderlich syndrome[4], it's a rare form of Müllerian tract abnormality. Müllerian malformations include a wide variety of developmental anomalies that result from the defective fusion or regression of the Müllerian ducts during foetal development.[1,3] The incidence of OHVIRA is low, occurring in 1 per 2000 to 1 per 28000 women.[1] These patients are usually asymptomatic until menarche, at which point they may present symptoms such a development of a mass secondary to hematocolpos, pelvic or vaginal pain and, less commonly, abdominal pain, vomiting, fever, abnormal vaginal discharge, or urinary retention.[2] The defining triad of this syndrome includes a didelphys uterus, obstructed hemivagina, and ipsilateral renal agenesis.[5] While renal agenesis is the most common renal anomaly associated with this syndrome, other renal malformations, such as dysplastic kidneys and renal duplication, have also been reported.[6]

Ultrasound is typically the first line of investigation for pelvic symptoms due to its accessibility. However, MRI is the recommended diagnostic imaging method for assessing Müllerian duct abnormalities. It provides detailed about the shape of uterine cavity, cervical and vaginal abnormalities also associated renal anomalies. It assists with surgical management.[6,7] Laparoscopy remains the gold standard for investigating gynecological congenital abnormatlities, but it is only used when MRI is inconclusive or unavailable.[8]

The treatment for OHVIRA involves vaginoplasty. This surgery includes draining the obstructed hemivagina and resecting the septum to restore normal vaginal function. This was the procedure used to treat our patient, we realised excision of the vaginal septum under general anesthesia with drainage of the hematocolpos. A long-term follow-up is planned to monitor the condition of the vaginal septum, hematocolpos and her future fertility.

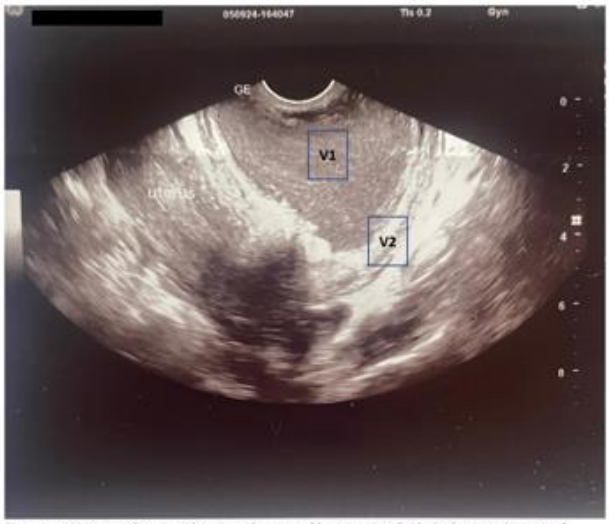

Figure 1: US image showing showing obstructed hemivagina (V1). And a nonobstructed hemivagina (V2)

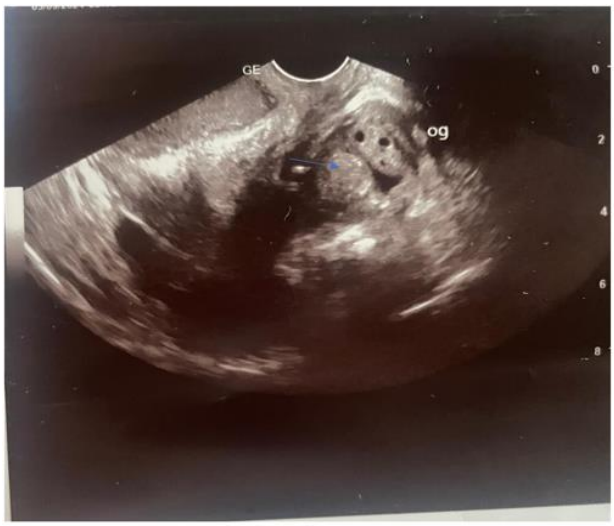

Figure 2: US image showing a left pyosalpinx (blue)

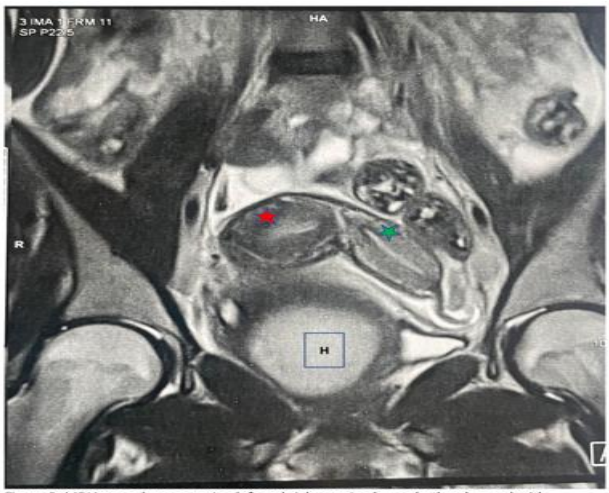

Figure 3: MRI image demonstrating left and right uterine horns (red and green) with hematocolpos (H)

CONCLUSION

Our case highlights a rare case of OHVIRA syndrome, featuring a didelphys uterus accompanied by an obstructed hemivagina and ipsilateral renal agenesis. Early diagnosis as well as an appropriate early treatment are crucial to prevent complications including chronic pelvic pain, pelvic infections and infertility. Gynecologists, pediatricians and radiologists should be aware of OHVIRA syndrome and maintain a high level of suspicion for this presentation when evaluating adolescent patients with non-specific abdominal or pelvis symptoms. Our work has been reported in line with the SCARE Guidelines 2023 criteria.[9]

CONFLICTS OF INTEREST

The authors declare that they have no competing interests relevant to the content of this article

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ETHICAL APPROVAL

Ethical approval is not applicable. The case reports are not containing any personal information.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.
Please see consent section in instructions to authors for further information.

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