How to Diagnose and Manage Septated Obstructed Hemivagina and Ipsilateral Renal Agenesis: A Rare Case

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Abstract

Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) syndrome is a congenital disorder that occurs during formation of Mullerian ducts. Due to its rare prevalence, diagnosis is commonly delayed by physician. We present a case of a 31-year-old female complaining of infertility for ten years. Physical examination showed one portio with 9 cm distance from hymen and a cystic mass on right vaginal wall with 6 cm distance from hymen. USG exam shows a bicollis uterus didelphys, two cervixes, hematocolpos with multiple septa on the right hemivagina and hematosalphynx were found. OHVIRA syndrome has very small incidence rate and varied clinical manifestations. Diagnosis is challenging for this case. The unique finding in this case was multiple longitudinal septa. Ultrasonography and hysteroscopy are some of the imaging choices for diagnosing OHVIRA syndrome. We conclude that early diagnosis and implementation can reduce the number of complications of this condition. Surgery can be considered as treatment of this case.

Keywords: OHVIRA Syndrome; Multiple Septated Hemivagina; Sonographic Findings; Surgical Approach.

Introduction

Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) syndrome is a congenital disorder that occurs in the formation of the Mullerian ducts that are the precursors of upper fallopian tubes, uterus, cervix, and vagina. In this syndrome, a number of abnormalities in the female reproductive organs are generally found, including uterine didelphys, unilateral hemivaginal obstruction, and ipsilateral renal agenesis. In general, the external genitalia may have a normal shape, therefore the diagnosis is generally established after menarche.[1,2] The exact prevalence of this syndrome is uncertain, but some literature mentions the prevalence ranging from

0.1 to 6%. Generally, puberty is diagnosed due to the presence of some symptoms such as progressive abdomen and pelvic pain. Not uncommon in other age groups, the primary complaints of patients are infertility, pyometra, urinary obstruction, or swelling of the ischiorectal region.[1-3] Due to a relatively rare prevalence, a common delay in diagnosis occurs in this condition leading to an increased likelihood of complications in this case. We describe a case of a 31-year-old woman with a complaint of primary infertility. It is hoped that this case report will add a level of attention to the diagnosis of OHVIRA syndrome so that it can be enforced and obtained early execution.

Case Report

A 31-year-old female patient came to a Urogynecology clinic with chief complaint of inability to get pregnant for 10 years, having done intercourse regularly twice a week and having been married for 10 years. The patient was a referral

from an OBGYN specialist previously with a diagnosis of bicornuate uterus. The patient had already had HSG, ultrasound, and OH examination. The patient had a normal menstrual cycle and menstrual pain was present.

Physical examination obtained general status within normal limits. Gynecological physical examination obtained the vulva and urethra were within normal limits,1 portio was found with a distance of 9 cm from the hymen. There was cystic mass on the right wall of the vagina with the distance of 6 cm of the hymen [Figure 1]. Ultrasonography examination found bicollis uterus didelphys with the size of 8.03 x 2.95 on the left side, and 6.63 x 2.22 in the right side, 2 cervixes, hematocolpos septum on the right hemivagina with the thickness of the septum 0.9 mm, 0.43 mm, and 0.34 mm. On the right adnexa, the impression of hematosalphynx was obtained, measuring at 4.78 cm x 2.33 cm x 2,79 cm with a volume of 18.27 cm2 [Figure 1]. The patient was diagnosed with OHVIRA syndrome and planned for vaginal septum resection.

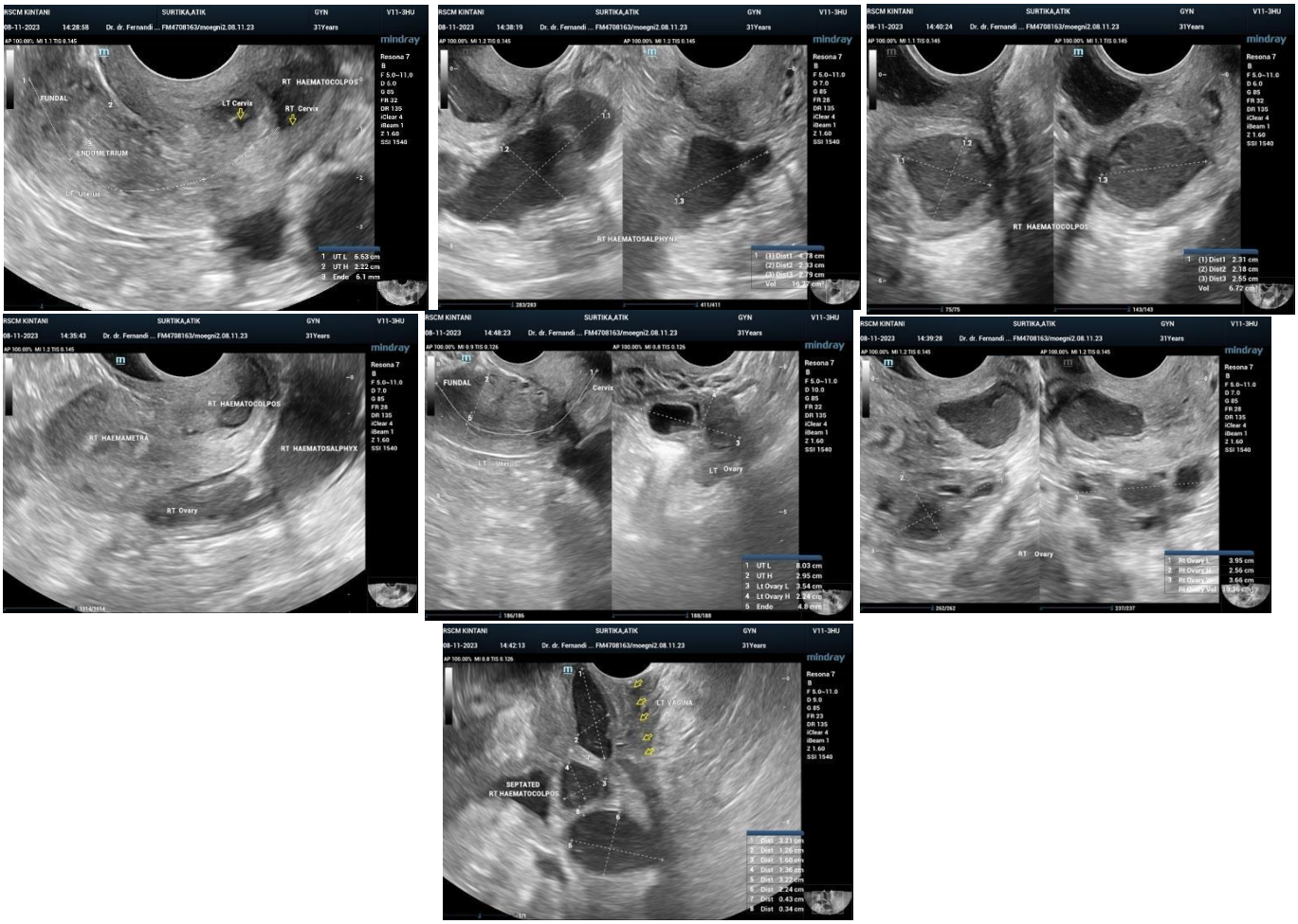

Figure 1: Gynecological Ultrasound.

At the time of the surgery, the septum was identified, marking the lateral and medial septums, gradual septum inscriptions were performed and the sewing continued, and the observation of the 2 cervicals was evaluated [Figure 2].

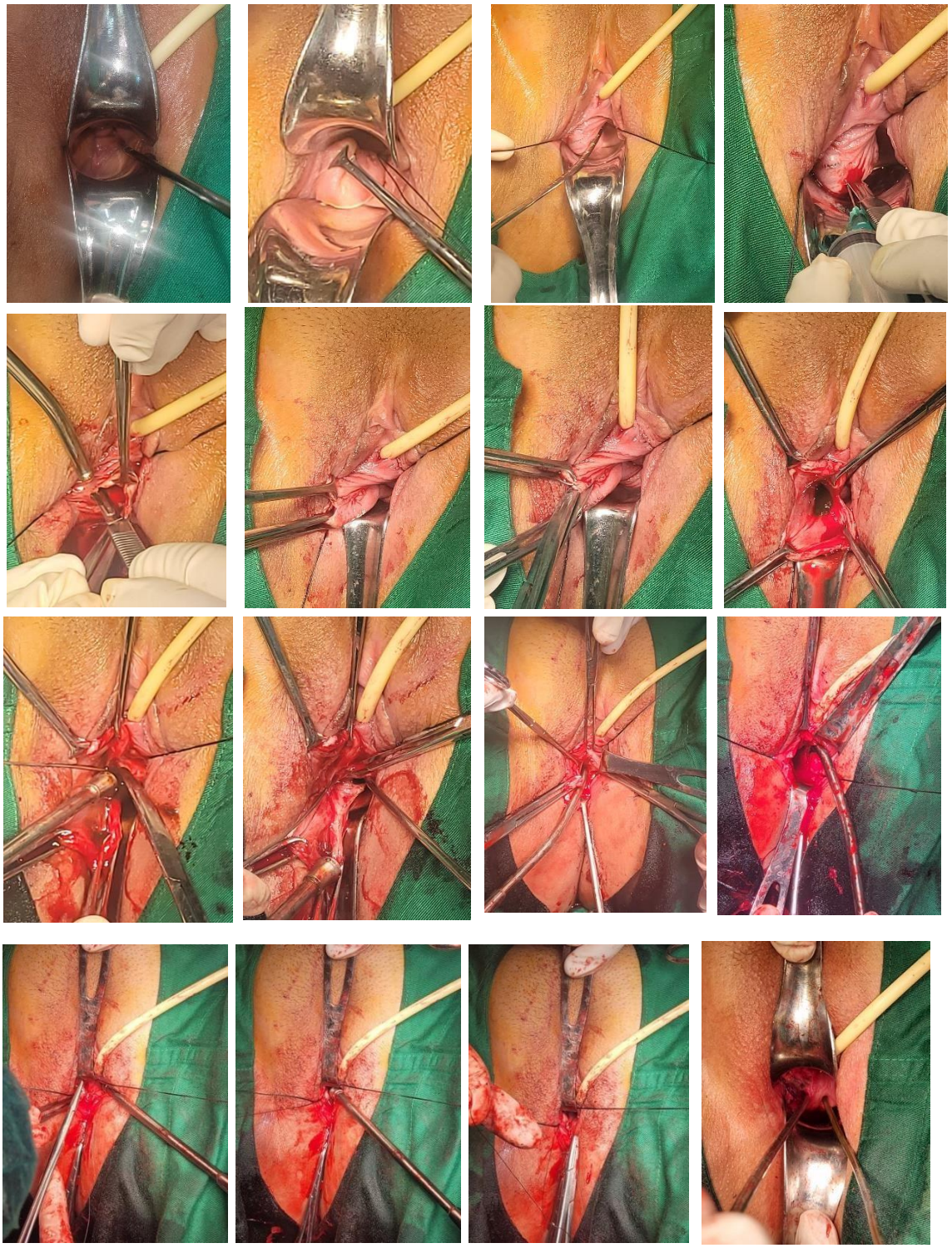

Figure 2: Intraoperative findings.

Follow-up examination was done a month after the surgery. The patient had no post-surgical pain and no other complaints. She also had normal menstruation after the surgery with duration of seven days. Gynecological physical examination showed that the vulva and urethra were within normal limits. Two cervical portio were seen and sondage was able to be inserted to both portio [Figure 3]. Ultrasonography examination found bicollis uterus didelphys with no hematocolpos and hematometra present [Figure 4].

Figure 3: Postoperative sondage.

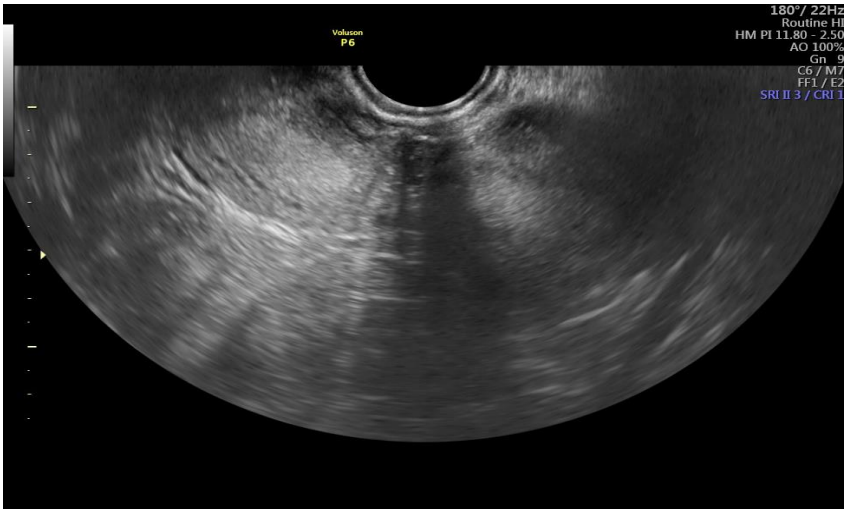

Figure 4: Postoperative ultrasonography.

Discussion

OHVIRA syndrome is part of the group of Mullerian ducts anomalies. This condition can occur in the female reproductive organs due to the absence of embryogenesis imperfections of the mullerian ductus and the paramesonephric. The incidence of anomalies of the Mullerian duct varies between 2%-3% with a frequency of 1 in 200 females, or 1 in 600 fertile females. OHVIRA syndrome has an incidence rate of 5% of the total cases of anomaly of the Mullerian ducts, with uterus didelphys being the main finding generally obtained.

Characteristic of this condition consists of three findings: uterus didelphys, hemivagina obstruction, and the presence of ipsilateral renal agenesis.[4-6] Diagnosis is a challenge for clinicians facing this case. Diagnosis becomes a challenge due to its non-specific and varied clinical manifestations. Reported in some cases found inconsistent manifestations associated with existing uterine abnormalities. The abnormalities of the uterus can be the didelphys (62.5 per cent), the septa (22.5 per cent) and the bicornuate (15%). In the case of a septa, the whole is a complete septa separated up to the level of uterine ostium. In some cases, a unilateral vaginal septum was found (75%), with the proximal part ended at the cervix, most unilaterally at the right (53%) and lateral left (46%). The case we found was interesting due to unusual clinical manifestations. In our case, the clinical manifestation is a multiple vaginal septum in longitudinal position.[5,7]

MRI modality is still the standard for diagnosing cases of OHVIRA syndrome. On modality MRI can provide a detailed anatomical picture, including contours, intrauterine morphology, and fluid observation specifically. In addition, MRI modality can also evaluate clinical manifestations of pathologies such as endometriosis, adhesions, and renal abnormalities. The use of sonography and hysteroscopy in cases of OHVIRA syndrome can be considered because of the rapid diagnosis and arrange surgical approach correctly. In this case, an ultrasound and hysteroscopy are performed to evaluate anatomy efficiently and quickly to establish a diagnosis quickly and not delay treatment.[7,8] Implementation of this condition has the principle to eliminate the symptoms of the existing symptoms. Therefore, surgical intervention remains an effective and more common choice to be chosen by clinicians. The resection of the part of the septum where the obstruction occurs is the key to the enforcement procedure. In general, patients will recover completely after vaginal resection. In general, this action is best performed during the menstrual phase due to the enlargement of the hematocolpos section which gives a better viewing space and anatomical visualization. In general, if this case is diagnosed and performed properly, then will have a good outward appearance. In this case, a septum resection is performed one by one to see the presence of multiple septum images on the diagnostic imaging.[9,10] OHVIRA syndrome is one of the very rare cases, which makes this case sometimes out of a clinic's reference diagnosis. With the increase in this case as an appeal diagnosis for the clinic, then this case will be easier to detect earlier and reduce the number of complications that occur due to delayed diagnosis and execution.[2,3]

Conclusion

OHVIRA syndrome is a condition of the Mullerian ductal anomaly group. The incidence of this syndrome is very small and has varied clinical manifestations. Diagnosis is a challenge for the clinic facing this case. Early diagnosis and implementation can reduce the number of complications of this condition. Surgery can be considered in the execution of this case.

References

1. Li X, Liu T, Li L. Herlyn-Werner-Wunderlich syndrome and its complications: A report of two cases and literature review. Radiol Case Rep 2021 Jun;16(8):2319-2324.

2. Girardi Fachin C, Aleixes Sampaio Rocha JL, Atuati Maltoni A, das Chagas Lima RL, Arias Zendim V, Agulham MA, et al. Herlyn-Werner-Wunderlich syndrome: Diagnosis and treatment of an atypical case and review of literature. Int J Surg Case Rep 2019;63:129-134.

3. Horst W, de Melo RC, Theilacker G, Schmitt B. Herlyn-Werner-Wunderlich syndrome: clinical considerations and management. BMJ Case Rep 2021 Mar;14(3):e239160.

4. Piccinini PS, Doski J. Síndrome de Herlyn-Werner-Wunderlich: Relato de caso. Rev Bras Ginecol Obstet 2015;37(4):192-196.

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

6. Mabuchi S, Hayashida H, Kubo C, Takemura M, Kamiura S. Herlyn-Werner-Wunderlich syndrome (HWWS)-associated gynecological malignancies: A case report and literature review. Gynecol Oncol Rep 2022 Jul;43:101051.

7. Zhang H, Ning G, Fu C, Bao L, Guo Y. Herlyn-Werner-Wunderlich syndrome: diverse presentations and diagnosis on MRI. Clin Radiol 2020 Jun;75(6):480.e17-480.e25.

8. Liu M, Zhang L, Xia Y, Huang X, Ye T, Zhang Y, et al. New Consideration of Herlyn-Werner-Wunderlich Syndrome Diagnosed by Ultrasound. J Ultrasound Med 2021 Sep;40(9):1893-1900.

9. Baños Cándenas L, Abehsera Davó D, de la Peña Rodríguez de Medina M, Martín Orlando J, López Díaz AC. Diagnosis and gestational follow-up in a patient with Herlyn-Werner-Wünderlich syndrome, a case report. Taiwan J Obstet Gynecol 2019 Jul;58(4):560-565.

10. Roziana R, Nora H, Maharani CR, Yeni CM, Dewi TP, Rusnaidi R, et al. Herlyn-Werner-Wunderlich syndrome: Challenges in diagnosis and management. Narra J 2023 Aug;3(2):e223.