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Case Report
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A Case Report of Ohvira Syndrome

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Abstract

Background: Obstructed Hemivagina Ipsilateral Renal Agenesis (OHVIRA) syndrome, is a congenital defect of Mullerian ducts that is characterized by obstructed hemivagina, uterine didelphys, and ipsilateral renal agenesis.

Case Presentation: A 13-year-old female came with pain abdominal and a palpable mass in the abdomen, which was later diagnosed with hematometrocolpos and primary amenorrhea. Ultrasonography was suggestive but MRI findings confirmed uterine didelphys, obstructed hemivagina with left renal agenesis, along with being suggestive of OHVIRA syndrome. She underwent surgical intervention under general anesthesia with drainage of hematocolpos and resection of obstructive vaginal septum.

Conclusion: Intraoperative hysteroscopy and laparotomic exploration confirmed diagnosis with two uterine horns with no connection between the two. Patient then had substantial resolution in symptoms and success with a normal follow-up case management for reproductive health.

Keywords: OHVIRA syndrome; Uterine didelphys; Primary amenorrhea; Hematometrocolpos; Renal agenesis

Introduction

Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) syndrome, or Herlyn-Werner-Wunderlich syndrome, is an exceptionally rare, congenital condition associated with Müllerian duct development anomalies leading to concurrent female reproductive and urinary system anomalies. The global burden of OHVIRA syndrome can be as high as 3.8% and as low as 0.1% [1]. OHVIRA syndrome is primarily characterized by uterine didelphys, obstructed hemivagina, and unilateral renal agenesis.

Menstrual flow obstruction, leading to either hematocolpos or hematometra, is primarily associated with the clinical symptoms of OHVIRA syndrome. Both hematocolpos and hematometra can lead to cyclical severe pelvic pain, abdominal distension, and either delayed menarche or in some cases, primary amenorrhea in the adolescent female population [2].

Many patients may remain undiagnosed until menarche or until cyclical signs and symptoms manifest frequently enough to merit a medical consultation, including severe cyclical pelvic pain or abdominal masses presented as palpable masses. OHVIRA syndrome, while rare, is often diagnosed late due to the difficulty in establishing the diagnosis and the variability in presentation, which may ultimately lead to complications of endometriosis, pelvic inflammatory disease and in some cases, infertility. Diagnostic imaging is critical in detailing the anatomical defects of OHVIRA syndrome. The most common imaging modalities are MRI and transabdominal ultrasounds. MRI is of significant value because it has more contrast to visualize soft tissue and can provide high-resolution images of the pelvic anatomy. This information can help differentiate OHVIRA syndrome from other Mullerian anomalies, which is valuable for surgical planning [3]. Surgery is generally indicated when OHVIRA is suspected based on clinical presentation and imaging because it will ultimately relieve symptoms.

Surgically treating OHVIRA syndrome usually involves draining the hematocolpos and/or the hematometra, and resecting the obstructive vaginal septum. The role of hysteroscopy in preservation of the lower uterine segment is also helpful to visualize both uterine cavities to ensure that both didelphys uteri are with flow and any abnormalities previously seen are not obstructing flow, ensuring excellent generalizability with improved diagnostic accuracy. Surgical treatment allows for not only symptomatic relief but also avoids complications and maintains reproductive potential

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which is essential for young patients who may desire reproductive activity in the future [4].

Case Presentation

A 13-year-old female presented to the Department of Obstetrics and Gynaecology, NEIGRIHMS, with a two-week history of lower abdominal pain and a palpable mass in the lower abdomen without attained menarche. On physical examination, a firm, non-tender mass was palpable in the left lower abdomen consistent with a 16-week gestation size. Secondary sexual characteristics were Tanner stage V, indicating complete pubertal development. Local examinations revealed an oblique vaginal septum. Based on clinical presentations and examination findings, further imaging studies were performed to evaluate the congenital Mullerian anomalies. Ultrasonography of the abdomen revealed hematometrocolpos and a left-sided hematosalpinx, suggesting distal obstruction of the genital tract. A non-communicating right rudimentary horn was suspected with non-visualization of left kidney and gave impression suggestive of transverse vaginal septum with absent or ectopic left kidney. The left kidney was not visualized, which was concerning for renal agenesis. Magnetic resonance imaging demonstrated uterine didelphys, an obstructed left hemivagina with hematocolpos, and left-sided renal agenesis, confirming the diagnosis of OHVIRA syndrome (Figure 1). The patient underwent surgical intervention under general anesthesia, which included the drainage of hematocolpos and resection of the obstructive vaginal septum. Intraoperative hysteroscopy was also performed to visualize two non-communicating endometrial cavities (Figure 2). Laparotomy was done to confirm a well-developed left-sided uterus and ovary, and a smaller right uterus with its own fallopian tube and ovary. Laparoscopy could not be performed since the patient was of a thin build, and an appropriately sized Veress needle was not available. The procedure was well tolerated, and the patient experienced significant symptomatic relief postoperatively. The patient made an adequate recovery and was followed up to assess her reproductive health and whether she experienced any complications. This case illustrates the importance of considering OHVIRA syndrome in an adolescent presenting with primary amenorrhea with abdominal pain and pelvic masses because earlier recognition and management leads to relief of symptoms, management, and return to reproductive function.

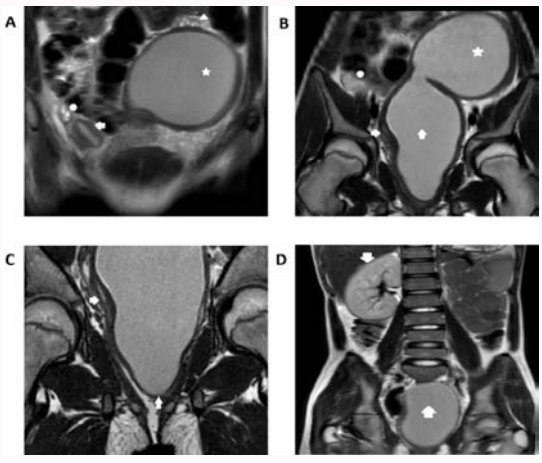

Figure 1: A: Star distended left horn of uterus, triangle left ovary, left arrow compressed right horn of uterus; B: Right arrow compressed right hemivagina, upward arrow-distended left hemivagina, right arrow compressed right hemi vagina, upward arrow obstructed and distended left hemivagina; C: Right arrow compressed right hemivagina, upward arrow obstructed and distended left hemivagina; D: Downward arrow right kidney with non-visualization of Left kidney.

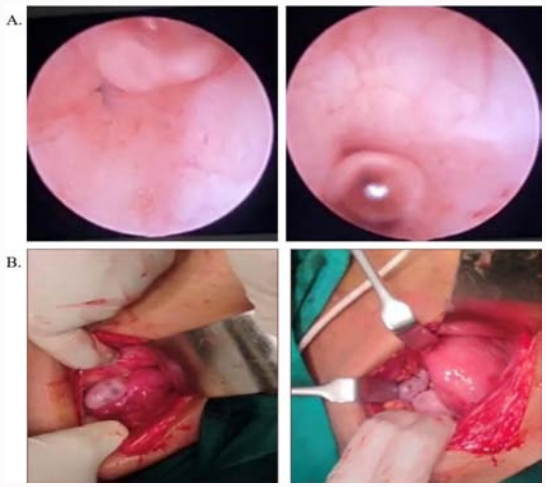

Figure 2: A: Hysteroscopic view B: Laparotomy view.

Discussion

OHVIRA syndrome generally presents in adolescent females when they are a menarche, and symptoms of severe abdominal pain and pelvic mass may lead to consideration of the continued understanding of OHVIRA syndrome [5]. Most of the evidence regarding OHVIRA has come from adult and post-menarche studies. Currently, there are only a handful of retrospective studies with small populations that have shown OHVIRA had distinct features in pre-menarche patients [6]. The recognition of OHVIRA syndrome is critical in order to prevent complications of hematocolpos, hematometra, endometriosis, and infections. Treatment delays and/or inaccurate diagnoses can result in long-term chronic pelvic pain, pelvic adhesions, endometriosis and/or compromised reproductive function. Hence, sudden onset abdominal pain in young female patients with uncharacteristic severe symptoms or altered menstrual patterns should raise suspicion of as yet undiagnosed OHVIRA syndrome.

OHVIRA is best evaluated by imaging, as the obstructed hemivagina distorts anatomy and makes it difficult to find by ultrasound. Given the rarity of the condition, and the similar presentations with other pelvic pathologies, the diagnosis of OHVIRA syndrome may not be established until imaging and medical evaluation reveal symptoms. Imaging is essential in order to diagnose OHVIRA syndrome, especially MRI, for its ability to provide clear and accurate visualization of the reproductive and urinary system which can establish the triad of OHVIRA syndrome [7]. Surgical intervention is often the preferred treatment, involving drainage of hematocolpos and septum resection to relieve obstruction.8 This approach provides for a relief of symptoms, avoids some further complications; and ultimately improves the future potential for fertility. The positive outcome in this case highlights the important role of timely surgical management for patients with OHVIRA syndrome and reinforces the additional need for healthcare providers to be cognizant of this rare syndrome, so that delays in diagnosis can be reduced and these patients can benefit from improved outcomes.

This case illustrates the clinical and operative management of OHVIRA syndrome in a young premenarchal adolescent - an

uncommon demographic for diagnosis. By giving an image with the MRI findings, in addition to intraoperative hysteroscopy and laparotomy confirmation, we are afforded a reasonable, complete diagnostic picture. The patient being clinically menarchal despite having developed secondary sexual characteristics reiterates the importance of considering OHVIRA in the early adolescent patient presenting with abdominal pain and how early diagnosis/intervention could prevent issues later down the road and possibly preserve fertility.

Conclusion

Timely diagnosis of OHVIRA syndrome is critical to reducing the risk of long-term reproductive and urological sequelae. The best approach for management involves a multidisciplinary team who accurately identify the syndrome through imaging, and performs appropriate surgical intervention as indicated. Early identification with clinical suspicion of OHVIRA is particularly important in adolescents with complaints of unexplainable pelvic pain and menstrual disorders.

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