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CASE REPORT

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Obstructive hemivagina with ipsilateral renal agenesis in a 16-year-old African girl with diabetes: A case report

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Abstract

Background: The obstructed hemivagina with ipsilateral renal agenesis (OHVIRA) syndrome is a type of Müllerian duct anomaly (MDA) resulting from abnormal development of Müllerian ducts. Its features typically become apparent at puberty when girls present with menstrual abnormalities. The varied presentation of MDAs presents diagnostic and management challenges.

Case presentation: A 16-year-old diabetic African girl presented to Savannah Healthcare with cyclical lower abdominal pain and foul-smelling per vagina discharge. Magnetic resonance imaging (MRI) showed complete duplication of the uterus, cervix, and vagina (uterine didelphys) with an obstructed left hemivagina with hematocolpos. The septum obstructing the hemivagina was resected with drainage of the hematocolpos. A follow-up abdominal renal ultrasound showed the absence of the left kidney.

Conclusion: Careful clinical examination and MRI are essential for diagnosing OHVIRA syndrome. Prompt surgical intervention improves outcomes.

Keywords: hematocolpos, OHVIRA, renal agenesis, septoplasty

Introduction

Müllerian duct anomalies (MDAs) result from abnormalities in embryological differentiation and development of the Müllerian ducts, typically identified at menarche when patients present with menstrual disorders (1). If not corrected, they cause persistent menstrual, sexual, and reproductive challenges (2). The OHVIRA syndrome, also known as the Herlyn-Werner-Wunderlich syndrome is a type of MDA whose presentation may result in delayed diagnosis and suboptimal treatment (3). This case highlights the diagnostic, management, and outcome challenges of an atypical presentation of Müllerian duct anomalies and highlights their impact on quality of life.

Case presentation

A 16-year-old nulliparous African girl, with well-controlled diabetes, presented to Savannah Healthcare with a two-month history of foul-

smelling vaginal discharge that started following a cervical dilatation procedure for stenosis and haematometra. She did not respond to multiple courses of antibiotics. She reported social withdrawal due to persistent malodor. She presented to another facility with cyclical lower abdominal pain and discomfort since her menarche two years earlier. An ultrasound showed cervical stenosis and hematometra, necessitating the cervical dilatation procedure. Her diabetes, diagnosed at 14 years old, was well controlled with insulin. Her other medical or surgical history was unremarkable.

A vaginal examination revealed normal external genitalia and a reddish foul-smelling vaginal discharge. A bulging cystic mass was observed on the left aspect of the vagina ending just below the hymen. A hemivagina with a hematocolpos was suspected. A pelvic magnetic resonance imaging (MRI) demonstrated a class III Müllerian duct anomaly with uterine didelphys, complete duplication of the cervix and uterus, and an obstructed left hemivagina with hematocolpos due to a distal transverse septum (**Figure 1-2**).

FIGURE 1

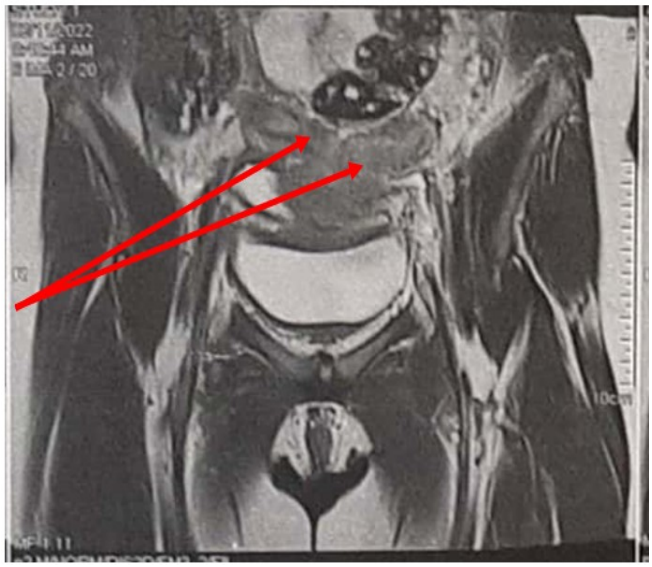

Figure 1: Pelvic magnetic resonance imaging (MRI) showing complete duplication of the uterus (red arrows).

Her laboratory investigations including complete blood count, renal function tests, and c-reactive protein (CRP) were unremarkable. Examination under anesthesia (EUA) plus a corrective surgery was scheduled. Intraoperatively, foul-smelling discharge that builds up as soon as it is emptied was noted. The right vaginal wall and cervix were grossly normal. On the left aspect was a longitudinal septum ending as a terminal transverse septum enclosing a bulging cystic cavity. There was a traumatic opening along the longitudinal septum through which the discharge was oozing. Upon excision of the transverse septum, a left vaginal cavity was noted, filled with foul-smelling old blood. After draining it, a left cervix was noted, and a normal sounding obtained. She was noted to be menstruating through both cervices.

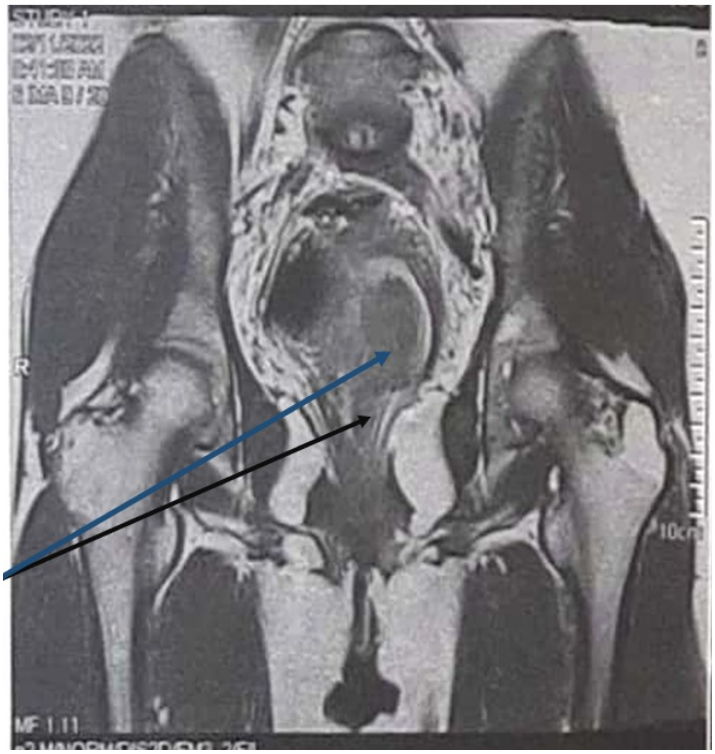

FIGURE 2

Figure 2: Pelvic magnetic resonance imaging (MRI) showing hematometra in the left uterus (blue arrow) and hematocolpos in the left hemivagina (black arrow).

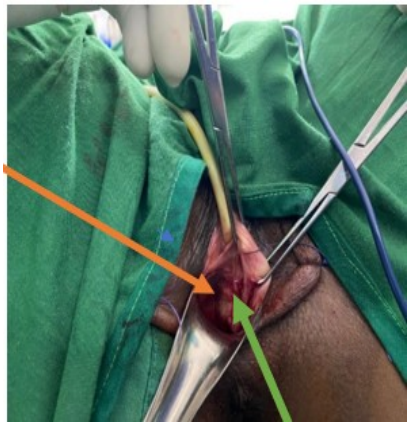

FIGURE 3: A

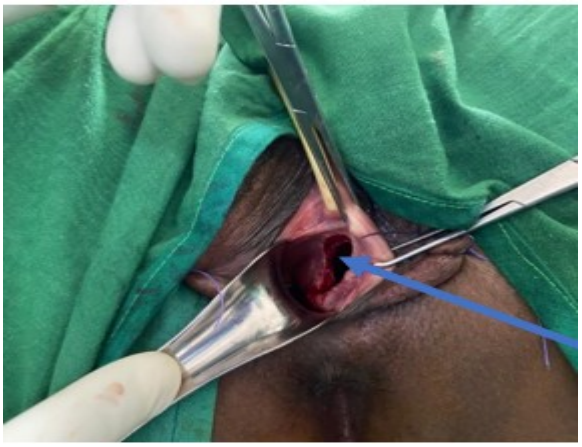

FIGURE 3: B

Figure 3: Gross surgical image (**A**) showing right open vaginal canal (orange arrow) and obstructed left hemivagina with hematocolpos that appear as a left building mass (green arrow); (**B**) Post-excision left hemivagina (blue arrow).

Septoplasty was performed by complete excision of the transverse septum and excision of the longitudinal septum to approximately the proximal third. The vagina was packed with iodine-laden gauze for 24 hours, and prophylactic antibiotics were administered. The patient was discharged the following day. A follow-up transabdominal renal ultrasound found a missing left kidney confirming the diagnosis of OHVIRA syndrome. During the two weeks of postsurgical review, she had no vaginal discharge. She subsequently reported normal menses and resumed school.

Discussion

Female genital tract development is a complex and highly regulated process involving cellular differentiation, migration, fusion, and canalization. During fetal development, the paired Müllerian ducts fuse in an organized manner to form the fallopian tubes, uterus, cervix, and upper two-thirds of the vagina. Development abnormalities at any stage leads to abnormal development or failure to develop these structures, leading to Müllerian duct anomalies (4). These rare anomalies affect approximately 1% of all women and 3% of women with poor reproductive outcomes (5). The OHVIRA syndrome comprises a triad of didelphys uterus, obstructed hemivagina, and ipsilateral renal agenesis. It is a rare MDA with an incidence of 1 in 2000 to 1 in 28000 women, accounting for 0.16%-10% of MDAs (6). According to the 2013 European Society of Human Reproduction and Embryology and European Society for Gynaecological Endoscopy (ESHRE/ESGE) consensus on the classification of female genital tract anomalies, it is classified as U3bC2V2 (6).

The disorder is usually discovered during puberty or shortly thereafter when girls present with menstrual abnormalities, including pelvic pain and swelling due to retained menstrual blood in the hemivagina (5). Because of the rare nature and nonspecific presentation, diagnosis can be delayed, increasing the risks of complications such as endometriosis, pyocolpos, and infertility (3). This case initially presented to a different facility with cyclical abdominal pain and discomfort since menarche and was managed as a case of cervical stenosis, then presented later with a foul-smelling vaginal discharge for two months which was treated with antibiotics without improvement. The discharge was noted to be old blood that collected in the obstructed left hemivagina and escaped through an opening that was probably iatrogenically created on the longitudinal septum during cervical dilatation. Cases presenting with excessive mucopurulent discharge from pyocolpos have also been previously reported with suggestions that the two vaginal cavities can communicate through a partially fenestrated septum leading to secondary infection of the blood retained in the obstructed hemivagina with pus formation (6). A closer clinical examination and high index of suspicion made us suspect a hemivagina with haematocolpos, hence the request for an MRI, which has been touted as the most sensitive diagnostic method (7).

The clinical course of the pathology is unique for each patient, and the treatment is based on specific presentation. However, most cases are treated through excision of the vaginal septum with a good outcome (8). When treatment is done early, there is a good prognosis and prevention of complications such as endometriosis, pyocolpos, and infertility (9). Timely treatment also improves the quality of life. The patient reported resolution of symptoms and improved quality of life. She had no more cyclical lower abdominal pains, and the foul-smelling vaginal discharge that stopped her from staying in school or social gatherings also stopped. In this case, we noted that the unusual presentation of MDAs carries diagnostic, management, and outcome challenges, with an impact on quality of life.

The diabetes placed her at a higher risk of infection thus she required more prompt diagnosis and treatment. She was diagnosed with diabetes at 14 years of age and was being managed as type 1 diabetes mellitus. While it could have been a coincidence that the girl also had diabetes, previous studies have reported associations between

mature onset diabetes of the young (MODY) and genitourinary anomalies (9). In another case report, a teenage girl was diagnosed with renal and Müllerian anomalies at 12 years then with diabetes at 16 years and her genetic evaluation reported an HNF1B gene mutation pointing to possibilities of genetic mutations in association with both renal and Müllerian anomalies (10). Whether this case was a MODY rather than type 1 diabetes mellitus is unknown. Due to limited resources, this case was not genetically evaluated.

Conclusion

OHVIRA syndrome, though rare, is an important cause of menstrual irregularities in adolescents. Clinical evaluation and MRI are essential for diagnosis, and timely surgical correction improves outcomes. Further research is warranted to explore potential associations between OHVIRA syndrome and diabetes in adolescents.

Informed consent for publication

Informed consent for publication was obtained from the patient's next of kin.

Declarations

Conflicts of interest

The authors have no conflicts of interest to declare.

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