Obstructed hemivagina and ipsilateral renal anomaly syndrome in an association with endometriosis: Role of Magnetic Resonance Imaging in diagnosis

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SUMMARY

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) is a rare congenital malformation of the female urogenital tract characterized by a triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal anomaly. It was formerly known as Herlyn Werner Wunderlich Syndrome (HWWS). The syndrome usually presents with cyclic pelvic pain following menarche. Endometriosis is a prevalent complication. Magnetic resonance imaging (MRI) helps in diagnosing OHVIRA syndrome and endometriosis due to its high contrast resolution and objectivity. We reported a 13-year-old girl who was evaluated for cyclic pelvic pain after her menarche at 12 years of age. Magnetic resonance imaging (MRI) revealed two separate uterine cavities, services and vaginae, indicating didelphys. The left uterine cavity is filled with fluid, and the left hemivagina is dilated and filled with hyperintense and hypointense fluid on T1 and T2, respectively, indicating blood products. Left hemivagina dilatation implicated the presence of an obstructing vaginal septum. A single left adnexal cyst lesion with blood products was suggestive of an endometriotic cyst. Additionally, the left kidney was absent. A uterine didelphys with left hemivagina obstruction, hematometra, hematocolpos, and the ipsilateral ovarian endometriotic cyst was diagnosed. A final diagnosis of OHVIRA syndrome or HWWS was made, considering that she had no left kidney.MRI is a suitable diagnostic tool for precise anatomical delineation of the uterus, cervix, and vagina in uterovaginal disorders such as OHVIRA syndrome. MRI can also properly evaluate endometriosis and adhesion.

INTRODUCTION

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome, formerly known as Herlyn Werner Wunderlich Syndrome (HWWS), was first reported in 1922. It is a rare condition of congenital malformation of the female urogenital tract involving müllerian and mesonephric duct anomalies. OHVIRA syndrome is a trifecta of ipsilateral renal abnormalities, obstructed hemivagina, and uterine didelphys symptoms.[1] The main clinical manifestations are cyclic pelvic pain and mass following menarche. Pelvic endometriosis and infection are prevalent complications. Early diagnosis is beneficial because this condition can be treated by vaginal septum excision and delay in diagnosis may worsen the associated endometriosis. Magnetic resonance imaging (MRI) is the modality of choice for diagnosing HWWS because of its high contrast resolution and objectivity.

CASE PRESENTATION

A 13-year-old girl was evaluated at our hospital for cyclic pelvic pain localized at the lower abdomen following her menarche (at 12 years old) as well as dark-brown discharge with a flow rate of approximately five menstrual pads per day. Previous medical history indicated no vaginal discharge, nausea, or vomiting before the current condition. Blood investigation revealed no significant changes in erythrocytes (4.63×10^6), leukocytes ($7 \times 10/\mu L$), hemoglobin (13.5 mg/dl), thrombocytes ($317 \times 10^3/\mu L$), prothrombin time (11.1 s; control: 11.0) and activated partial thromboplastin time (32.7 s; control 31.2). Her micturition is normal, with no substantial difficulties. Additional blood investigation revealed normal kidney function (blood urea nitrogen of 6 mg/dL and creatinine of 0.52 mg/dL) and normal blood electrolyte levels (sodium of 139 mEq/L, potassium of 3.7 mEq/L, and chloride of 105 mEq/L). Her family history indicated hymen imperforate in her younger sister.

Physical examination revealed lower abdominal bulging. Her most recent ultrasound examination revealed uterine enlargement, bicornuate, left ovarian cyst (4.95 × 3.95 cm), and uterine hematometra. MRI revealed two separate uterine cavities, cervices, and vaginas, indicating didelphys (Figure 1). The left uterine cavity was filled with fluid, and the left hemivagina was dilated and filled with hyperintense and hypointense fluid on the T1-weighted image (T1WI) and T2WI, respectively (Figure 2), indicating blood products. Left hemivagina dilatation implicated the presence of an obstructing vaginal septum. A single left adnexal cyst lesion with blood products indicated an endometriotic cyst. Additionally, the left kidney was absent (Figure 3). Following the laparoscopy diagnosis, we discovered a didelphic uterus, left hemi uteri (size 10×7×5 cm), right hemi uteri (size 7×5×4 cm), and multiple endometriotic nodules on the peritoneum and diaphragm wall. The patient underwent surgical intervention of septectomy, which consisted of needle aspiration resulting in approximately 150 ml of dark-red fluid. The septectomy was performed at 10 cm in length with ligation and sent to the Pathology Anatomy unit. After 3 days of care, the patient was diagnosed with uterine didelphys with left hemivagina obstruction, hematometra,

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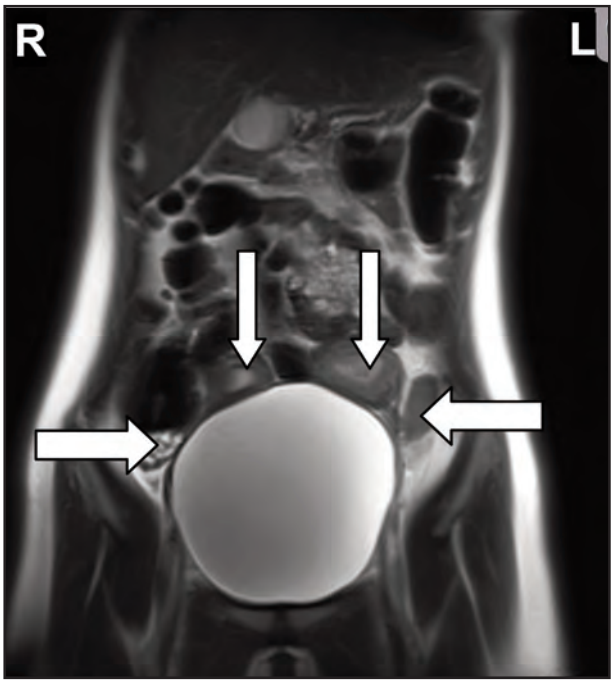

Fig. 1: Coronal T2WI MRI shows two separate uterine cavities (thin arrows) with normal right ovary and left endometrioma (thick arrows)

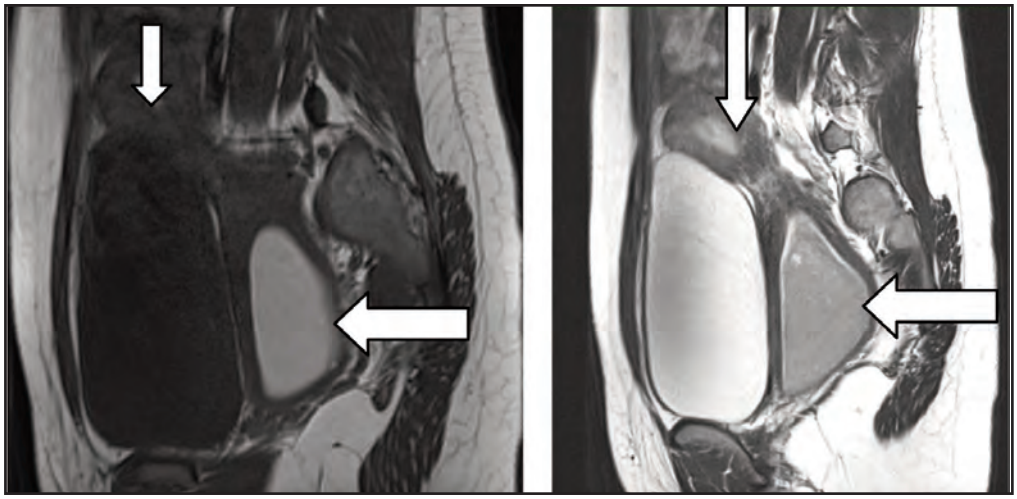

Fig. 2: Sagittal T1WI MRI [A] and T2WI MRI [B] show left uterine cavity (thin white arrows) with dilated hemivagina (thick white arrows) showing a hyperintense signal on T1WI (thick white arrow) and hypointense signal on T2WI/T2 shading (thick white arrow), suggestive of blood products within the uterus (hematometra, thin white arrow), and within the vagina (hematocolpos, thick white arrows)

hematocolpos, and ipsilateral ovarian endometriotic cyst. A final diagnosis of OHVIRA syndrome or HWWS was established, considering that she had no left kidney. No complaints were raised postoperatively, and the patient's daily activities returned to normal.

DISCUSSION

Congenital malformations, also known as Müllerian Duct Anomalies (MDAs), originate from improper müllerian duct development. This may be due to complete agenesis, defective vertical or lateral fusion, or resorption failure. Properly classifying MDA is important because the associated risks of poor pregnancy outcomes and treatment can vary widely among anomalies. The American Society of Reproductive Medicine developed the most prominent classification system.[2]

The OHVIRA syndrome, which was first identified in 1922 and is now known as the HWWS, combines a vaginal septum with type III müllerian anomalies.1 The incidence has been observed in 0.1–3% of various case studies.[3] The classic renal manifestation of OHVIRA syndrome is ipsilateral renal agenesis. Reports also indicated additional kidney anomalies, including crossed fused ectopias, duplicated kidneys, rectovesical bands, and dysplastic kidneys. Renal agenesis is the classic presentation, but other anomalies include renal dysplasia and ectopic ureters.[3,4] The müllerian ducts' lateral

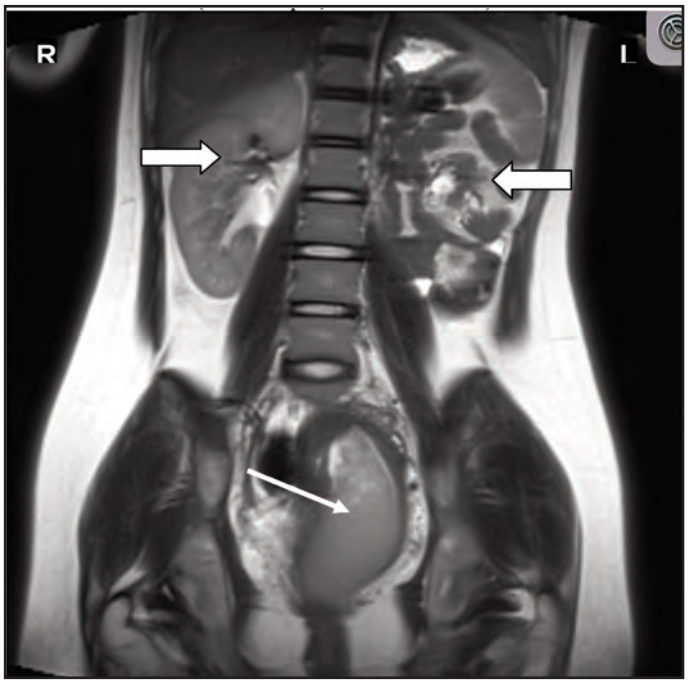

Fig. 3: Coronal T2WI MRI shows dilatation of left hemivagina filled with blood (thin arrows), normal right kidney and absence of left kidney (thick arrows)

nonfusion causes an asymmetric obstruction in uterine didelphys, usually accompanied by renal agenesis on the side of the obstruction.[4] Notably, renal agenesis is located ipsilateral to the dilated uterine cavity in patients with OHVIRA syndrome or HWWS. The caudal parts of the müllerian ducts fuse to form a single double cavity uterus at 8–12 weeks of fetal development. The septum recedes by 20 weeks of pregnancy, resulting in a single cavity. Hence, delayed development of the müllerian and mesonephric ducts at 8 weeks of gestation may cause an anomaly known as uterus didelphys.[5]

Non-specific symptoms, such as recurrent pelvic discomfort or dysmenorrhea caused by increasing distension of the blocked hemivagina, usually accompany menarche. A timely, correct diagnosis is crucial because quick treatment alleviates the problem, prevents additional consequences from chronic obstructed hematocolpos, and preserves reproductive potential. Improper information about the condition, frequent menstruation in the context of incomplete vaginal outlet obstruction, and delayed hematocolpos expansion were all cited for the delays in diagnosis. This case is very encouraging because the patient was diagnosed one year after menstruation. The age of the type of vaginal malformation, obstructed or unobstructed, is important for diagnosis. The average age of diagnosis is 12.8 years in cases with total hemivaginal blockage, compared with 20.6 years in cases of incomplete obstruction.[6]

This patient also had an endometriotic cyst (endometrioma) on one side with hemivaginal obstruction and multiple endometriotic nodules on the peritoneum and diaphragm wall. The relationship between pelvic endometriosis and HWWS remains unclear. Sampson's theory of retrograde menstruation and implantation is thought to explain most issues although no single theory of the etiology of endometriosis can explain all pelvic endometriosis cases. Regarding the association of endometriosis with müllerian anomalies, endometriosis is not more frequent in patients with müllerian anomalies as a whole but is more frequent with müllerian abnormalities with outflow obstruction, hematosalphynx, hematometra, or hematocolpos.[7] Jung et al. revealed that 100% of ovarian endometrial cysts were ipsilateral to the vaginal septum. Long-term continuous reverse menstrual flow has been predictive of pelvic endometriosis onset and progression. HWWS was associated with an increased risk of pelvic lesions.[8]

Ultrasound is frequently used as a screening modality for suspected müllerian duct abnormalities, but it is far inferior to MRI (able to differentiate with T1WI and shading on T2WI) because of its ability to differentiate details (e.g., cysts vs. Endometriosis). MRI rather than computed tomography should follow because the latter involves radiation exposure, and many questions remain unanswered because of its limited soft-tissue resolution. MRI is a good technique for examining the usually complex MDAs. The significant difference in uterovaginal anomalies is associated with multiplanar capabilities, better tissue characterization, and a wide range of views. Precise anatomical delineation of the uterus, tubes, cervix, and vagina is essential. Furthermore, MRI can quickly detect any concurrent renal and urethral abnormalities and define the contents of occluded cavities (e.g., simple fluid vs. blood). The health of these reproductive organs and the presence of endometriosis, pelvic inflammation, and adhesions can be determined through MRI.[9] MRI is an effective diagnostic support tool for identifying anatomical abdominal abnormalities with the right choice of protocol sequences to obtain optimal MRI.[9]

In general, T2-weighted sequences are considered the cornerstone of pelvic MRI because of their superiority in delineating the zonal anatomy of the uterus. T2-weighted sequences should include axial images and those parallel to the long axis of the uterus for better characterization of the external uterine contour, such as the adnexa and ovaries, for MDA classification. Targeted axial or coronal T2-weighted MRI of the abdomen allows assessment of associated urinary tract anomalies. T1-weighted images with excellent soft tissue contrast between the myometrium and the overlying fat may be helpful for characterization. More importantly, fat-saturated T1WI is instrumental in demonstrating blood products of a subacute stage in hematometroclopos or glandular components of deep infiltrative endometriosis.[9,10]

The primary treatment for OHVIRA syndrome is surgical intervention, specifically vaginal septum excision, which removes the blockage. One-stage vaginoplasty, which includes entire septum excision, is performed in a single procedure. Surgery reduces the risk of pelvic endometriosis because of retrograde menstrual seeding.[7] The patient's renal function was normal in this case, but it must always be evaluated because of renal agenesis on the side of the hemivagina obstruction. The history of the younger sister experiencing imperforate hymen needs to be further investigated to determine any recessive inheritance pattern.

CONCLUSION

OHVIRA syndrome or HWWS is characterized by a triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal anomalies. MRI is a suitable diagnostic tool for precise anatomical delineation of the uterus, cervix, and vagina in uterovaginal disorders such as OHVIRA syndrome. Endometriosis, which is a common complication of OHVIRA syndrome, can be properly evaluated using MRI.

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