Case Series

Magnetic Resonance Imaging Evaluation of Wolffian Duct Anomalies – OHVIRA and OSVIRA

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Abstract

Herlyn–Werner–Wunderlich syndrome in females, also known as obstructed haemivagina and ipsilateral renal agenesis (OHVIRA), and Zinner's syndrome, also known as OSVIRA in males, are two rare congenital syndromes affecting the urogenital tract. OHVIRA is obstructed haemivagina and ipsilateral renal agenesis and OSVIRA abbreviation stands for obstructed seminal vesicle and ipsilateral renal agenesis. For a long, these syndromes were considered different entities, owing to our poor understanding of underlying embryopathogenesis; however, in light of the recent acceptance of the Wolffian origin of the vagina in its entirety, these two syndromes are considered to be cut from the same cloth. In this case series, we present magnetic resonance imaging features of two cases each of OHVIRA and OSVIRA and discuss the similarities in embryopathogenesis behind them while debunking the old concepts. A deep understanding of embryopathogenesis allows a clinician and radiologist to predict the disease manifestation, and its outcome and also acts as a guide to screen for potential subtle defects, which would otherwise have been missed.

Keywords: *Magnetic resonance imaging, obstructed haemivagina and ipsilateral renal agenesis, OSVIRA, Wolffian duct anomalies*

Introduction

The genitourinary system develops predominantly from mesonephric ducts (also known as Wolffian duct) in males and paramesonephric duct (also known as Müllerian duct) in females.[1] Development starts from the 4th week of intrauterine life and both these ducts are found in a foetus. As the foetus grows, the Müllerian duct regresses in a female foetus, owing to the absence of testosterone and the Wolffian duct regresses in the male foetus. Anomalies in this process of development lead to a diverse gamut of congenital defects in the genitourinary system.[1] Here, we are illustrating magnetic resonance imaging (MRI) findings of two such rare anomalies, namely Zinner's syndrome in males and obstructed haemivagina and ipsilateral renal agenesis (OHVIRA) in females. Zinner syndrome, first described in 1914, has only a limited number of reported cases till now.[2] It is a constellation of unilateral ejaculatory duct obstruction, ipsilateral seminal vesicle obstruction with cyst and ipsilateral renal agenesis, abbreviated as OSVIRA, arising as a result of faulty development of the male urogenital tract.[2] Herlyn–Werner–Wunderlich syndrome, also known as OHVIRA, abbreviated for obstructed haemivagina and ipsilateral renal agenesis, is associated with faulty embryogenesis of the female genital tract.[3-5] The exact embryological basis of the syndrome has always been disputed; however, a deeper understanding of embryopathogenesis has led to the conclusion that OHVIRA and OSVIRA are syndromes with similar underlying pathogenesis having unique phenotypic presentations in opposite genders.[6-9] These are rare yet important causes of infertility in individuals. Through this case series, we are

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describing the embryological basis and the MRI features of both syndromes.

Case Series

We are illustrating four cases of Wolffian duct anomalies, two males and two females, who underwent MRI on a 3T SEIMENS 'SOMATOM' machine.

Patent one, with abdominal pain and fever, underwent ultrasonography (USG), which revealed, non-visualisation of the left kidney in the left renal fossa. A multi-loculated cystic lesion with low-level internal echoes, likely haemorrhagic in the left ovary, and loculated collection in the pelvis. MRI done for further evaluation revealed left-sided obstructed haemivagina with haematocolpos, uterine didelphys with left pyometra and left haematosalpinx, left ovarian haemorrhagic cyst and ipsilateral renal agenesis, as shown in Figure 1a-c.

Patient two is being worked up for infertility. USG reported non-visualisation of the right kidney in the right renal fossa and a pelvic loculated collection. Further evaluation with MRI revealed obstructed right haemivagina with large haematocolpos, uterine didelphys with right haematometra, right-sided haematosalpinx and ipsilateral renal agenesis, as shown in Figure 1d-f. However, due to excessively dilated right haemivagina, the right ovary could not be imaged, which was found to be compressed and displaced during intraoperative exploration. This represents a limitation of MRI to delineate pelvic structure in cases of excessively dilated structures.

With these findings in both cases, a diagnosis of OHVIRA was made with accompanying features of retrograde menstruation.

The third case is of a male patient, being evaluated for lower abdominal pain, and after preliminary investigation, an MRI was planned. MRI revealed left-sided obstructed ejaculatory duct, left seminal vesicle cysts and ipsilateral renal agenesis, as shown in Figure 2a-c. These findings led to a diagnosis of Zinner's syndrome.

The fourth case is that of a male patient being evaluated for unrelated complaints. On computed tomography, incidental findings of seminal vesicle cysts and right renal agenesis were reported, which raised a suspicion of Zinner's syndrome, as shown in Figure 2f. To confirm, an MRI was done, which revealed right-sided obstructed ejaculatory duct, right seminal vesicle cysts and ipsilateral renal agenesis, as shown in Figure 2d and e. However, the patient had no history of infertility.

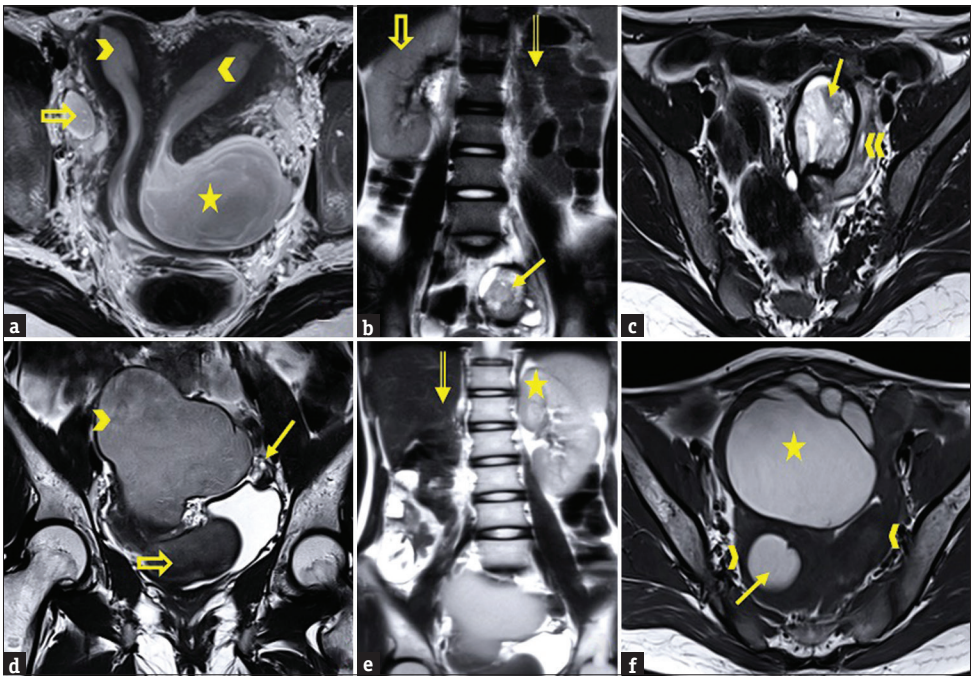

Figure 1: Magnetic resonance imaging (MRI) images of Herlyn–Werner–Wunderlich syndrome, aka obstructed haemivagina and ipsilateral renal agenesis (OHVIRA) syndrome. (a-c) A constellation of features of OHVIRA in patient one, (a) MRI T2 axial maximum intensity projection image showing uterine didelphys (arrowhead), left obstructed haemivagina with haemato-pyometra (star) and right simple ovarian cyst (block arrow), (b) MRI T2 coronal image depicting left renal agenesis (double arrow), normal right kidney (block arrow) and left ovarian haemorrhagic cyst (arrow), (c) MRI T2 axial image showing left ovarian haemorrhagic cyst (arrow) with left haematosalpinx (double arrowhead), (d and e) A constellation of features of OHVIRA in patient two, (d) MRI T2 coronal image shows large right haematosalpinx (arrowhead), obstructed right haemivagina with haematocolpos (block arrow) and normal left ovary (arrow), (e) MRI T2 coronal image shows right renal agenesis (double arrow) and normal left kidney (star), (f) MRI T2 axial image shows uterine didelphys (arrowhead), right haematometra (arrow) and right haematosalpinx (star)

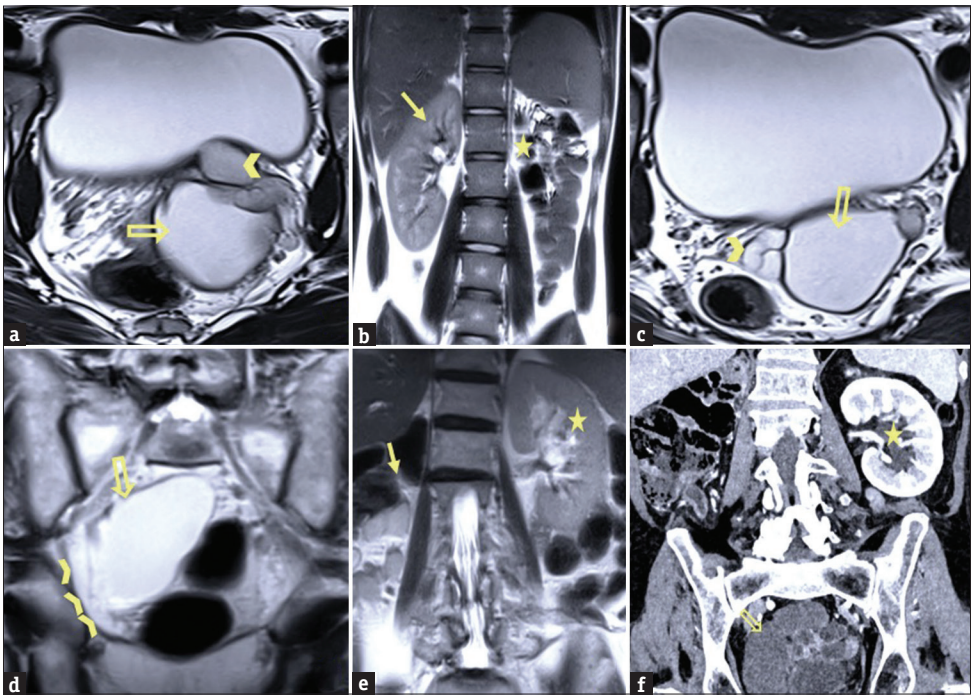

Figure 2: Magnetic resonance imaging (MRI) images of Zinner's syndrome, aka OSVIRA syndrome. (a-c) Constellation of features in Zinner's syndrome in case 3, (a) MRI T2 axial image showing left obstructed ejaculatory duct (blocked arrow) and left seminal vesicle cysts (arrowhead), (b) MRI T2 coronal image shows single right-sided kidney (arrow) and left renal agenesis (star), (c) MRI T2 axial image shows normal right-sided seminal vesicle (arrowhead) and left seminal vesicle cyst (blocked arrow), (d and e) MRI images of incidentally diagnosed case of Zinner syndrome, (d) MRI T2 coronal image shows dilated obstructed right ejaculatory duct (arrowheads) and right seminal vesicle cyst (blocked arrow), (e) MRI T2 coronal image shows solitary left kidney (star) with right renal agenesis (arrow), (f) Contrast-enhanced coronal computed tomography image showing solitary left kidney (star) and right seminal vesical cyst (arrow)

Out of four cases, one presented with infertility, two with lower abdominal pain, which was related to the anomaly, and one was incidentally diagnosed. Thus, Wolffian duct anomalies have an array of presentations, including, but not limited to, infertility.

A summary of the cases is given in Table 1.

Discussion

In this case series, we are discussing MRI findings of two rare structural congenital malformations, OSVIRA and OHVIRA.[2,3,5] With its excellent soft-tissue resolution, MRI is the mainstay to making an accurate diagnosis, but even with its cardinal role in diagnosis, MRI has its limitations. MRI, in most cases, is unable to delineate the vaginal septum and thus cannot classify OHVIRA and its types. In cases of excessively dilated haemivagina, evaluation of pelvic structures by MRI is suboptimal. These limitations are overcome by laparotomy, which provides additional information on the vaginal septum and pelvic structures and helps us to evaluate the pelvis in toto.

Diving into the embryology, the male urogenital system is derived from the mesonephric duct, and the female is derived from the paramesonephric duct, with the vagina having a disputable origin.[2,3,6] Till recently, it was accepted that the vagina has a dual origin. The upper part arising from Müllerian duct and the lower part from the sino-vaginal bulbs, but with recent advances, this concept has been replaced by the new concept of Wolffian origin of the vagina in its entirety.[6,9] First proposed by Acién's[7] and confirmed by Sánchez-Ferrer *et al*.,[8] this concept has put forward a new horizon of understanding pathologies and syndromes, including the vagina. One such syndrome is OHVIRA.[9]

OHVIRA is a rare Müllerian anomaly associated with complex urogenital anomaly.[3,4] All the components of this syndrome can be explained by this new hypothesis. Considering OHVIRA as a Wolffian duct anomaly, a faulty mesonephric duct development leads to renal agenesis/dysgenesis due to failure of induction of metanephric blastema. Due to the abnormal release of growth factors from an anomalous mesonephric duct, there is a disturbance in the positioning of the paramesonephric duct, leading to non-fusion and other uterine anomalies. Vaginal dysgenesis is seen, considering its origin from the mesonephric duct.[6-9] Females with OHVIRA syndrome are at increased risk of endometriosis due to recurrent retrograde menstrual flow, and approximately 40% have an ectopic vaginal insertion of the ureter.[3]

Table 1: Summary of cases

	Age (years)/ gender	Presenting complaints	Infertility history	Hallmark features of Wolffian agenesis	Associated Müllerian duct anomalies	Complications
OHVIRA						
Case 1	12/female	Lower abdominal pain with fever	Absent	Left obstructed haemivagina Left renal agenesis	Uterine didelphys	Left haematocolpometera Left haematosalpinx Left ovarian haemorrhagic cyst
Case 2	21/female	Infertility	Present	Right obstructed haemivagina Right renal agenesis	Uterine didelphys	Right haematocolpometera
OSVIRA						
Case 3	16/male	Lower abdominal pain	Absent	Left obstructed ejaculatory duct Left seminal vesicle cyst Left renal agenesis	None	None
Case 4	80/male	Right upper quadrant pain	Absent	Right obstructed ejaculatory duct Right seminal vesicle cyst Right renal agenesis	None	None

OHVIRA=Obstructed haemivagina and ipsilateral renal agenesis

Due to this development, one can now say that Zinner's syndrome is a counterpart of OHVIRA syndrome, sharing the same embryo pathogenesis of anomalous Wolffian duct development.[2,9] For time immemorial, Zinner's counterpart in females has been considered Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome, a group of anomalies consisting of uterine and vaginal dysgenesis, resulting from anomalous development of Müllerian duct.[3,6] However, the caveat here is that MRKH is of Müllerian origin and Zinner's syndrome is of Wolffian origin. Thus, in light of recent developments, OHVIRA syndrome is considered the female counterpart of Zinner's syndrome.[2,3,6,9]

Correct knowledge of embryopathogenesis is cardinal while reporting cases of congenital anomalies, as anomalous development of a structure can give rise to a spectrum of findings, of which many might be subtle. However, a precise working knowledge of embryopathogenesis, helps radiologists and clinicians to have a targeted approach and screen for the spectrum in toto, leading to prompt, accurate and complete reporting, which in turn leads to favourable patient outcomes.

Conclusion

In this case series, we have tried to bring out the similarities between Zinner's and OHVIRA syndrome, which share the same embryopathogenesis of anomalous Wolffian duct development, in males and females, respectively, contrary to the old concept of Müllerian origin of OHVIRA syndrome. Thus, OHVIRA syndrome and Zinner's syndrome are two sides of the same coin. Wolffian duct anomalies have a varied clinical presentation, thus MRI, with its soft-tissue resolution, plays a cardinal role in accurate diagnosis and management.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Author's contributions

LB, SA - Concept and manuscript writing; LB, SA - Acquisition and interpretation of imaging studies; All authors have critically reviewed the manuscript.

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Conflicts of interest

There are no conflicts of interest.

Data availability statement

Data will be available from corresponding author upon reasonable request.

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