Sonographic diagnosis of Obstructed Hemivagina and Ipsilateral Renal Anomaly Syndrome: a report of two cases

Abstract

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome is a rare urogenital anomaly. Patients typically present at puberty, shortly after menarche with increasing pelvic pain, dysmenorrhea and pelvic mass. There may be a known history of unilateral renal agenesis. Diagnosis can usually be established by ultrasound or magnetic resonance imaging. Resection of the vaginal septum is the treatment of choice. Early diagnosis and treatment can relieve symptoms, prevent complications and preserve fertility. We present two cases of OHVIRA syndrome diagnosed by ultrasound to promote recognition of this rare but important condition.

Keywords: Herlyn-Werner-Wunderlich syndrome, obstructed hemivagina, OHVIRA syndrome, renal anomaly, ultrasound, uterine didelphys.

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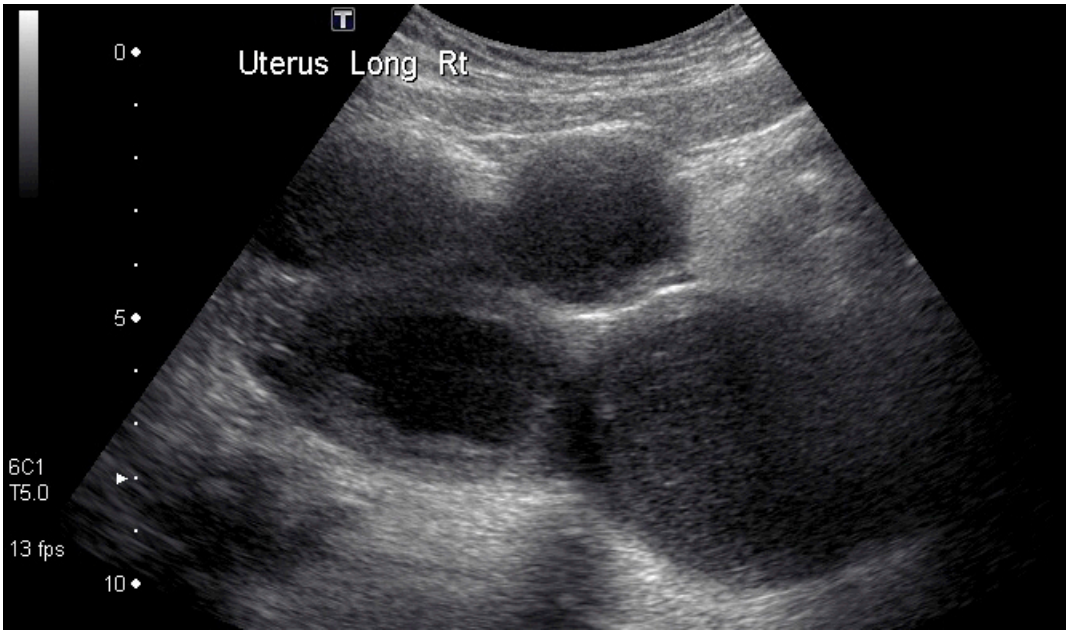

Figure 1: Sagittal scan showing right hemiuterus with distended cavity and vagina containing echogenic material.

Introduction

The combination of obstructed hemivagina and uterine didelphys was first reported in 1922.[1] The triad of uterine didelphys with obstructed hemivagina and ipsilateral renal agenesis has been referred to as Herlyn-Werner-Wunderlich syndrome with the first paper in English to use this term published in 2006.[2] The acronym OHVIRA (obstructed hemivagina and ipsilateral renal anomaly) syndrome was proposed in 2007 to allow inclusion of other uterine and renal anomalies.[3] The incidence of uterine didelphys related to this syndrome is approximately 1/2,000 to 1/28,000, and it is accompanied by unilateral renal agenesis in 43% of cases.[4] It has been estimated that there would be only four to five cases of uterine didelphys and obstructed hemivagina per year in Australia.[5] Early diagnosis and surgical intervention in OHVIRA syndrome is important to relieve symptoms, prevent complications and preserve future fertility.[6] We present two cases of OHVIRA syndrome seen and managed at our institution to promote recognition of this rare urogenital anomaly.

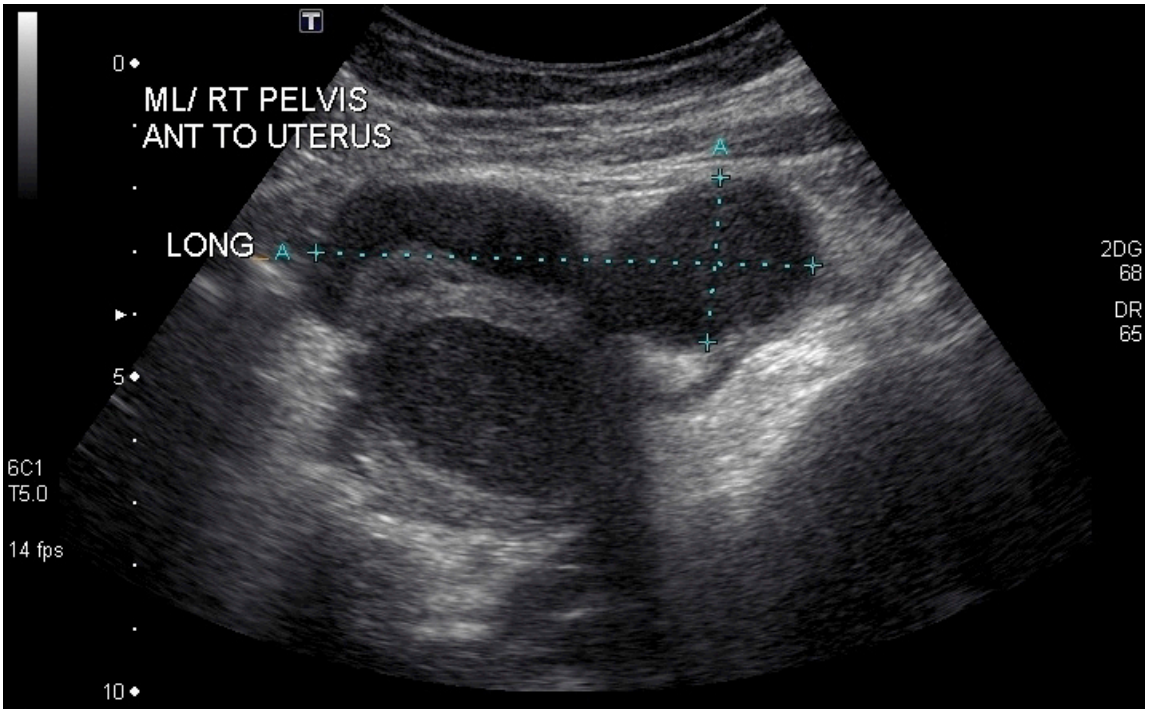

Figure 2: Sagittal scan showing a tubulocystic collection containing echogenic material arising from the cornu of the right hemiuterus.

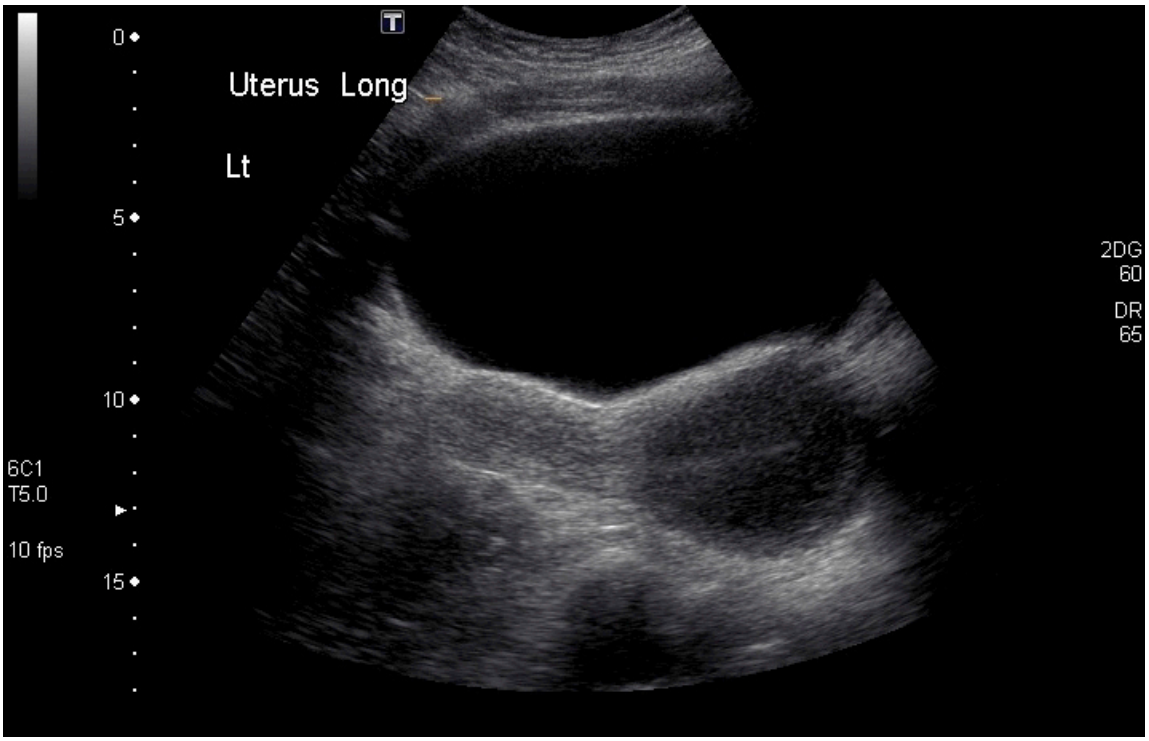

Figure 3: Sagittal scan showing normal left hemiuterus.

Cases

1) A 13-year-old girl presented to the Emergency Department with worsening lower abdominal pain. This was her third such presentation in nine months. She was known to have an absent right kidney. She had menarche at the age of 11 years with menses lasting 5 to 6 days and irregular 30 to 60 day cycle length. She had worsening dysmenorrhoea requiring treatment. At her first presentation she was admitted with suspected appendicitis. The appendix was found to be normal at surgery and on histopathology. At her second presentation she was treated for a suspected urinary tract infection. Pelvic ultrasound performed in the Medical Imaging Department at her third presentation showed uterine didelphys, right hematometrocolpos (Figure 1) and probable right haematosalpinx (Figure 2). The left hemiuterus appeared normal (Figure 3). The right kidney was absent (Figure 4). Laparoscopy and hysteroscopy revealed normal ovaries and left tube, normal left hemiuterus, enlarged right hemiuterus, right haematosalpinx and a thick longitudinal vaginal septum with right-sided haematocolpos. The vaginal septum was excised with drainage of 400 milliliters of old blood.

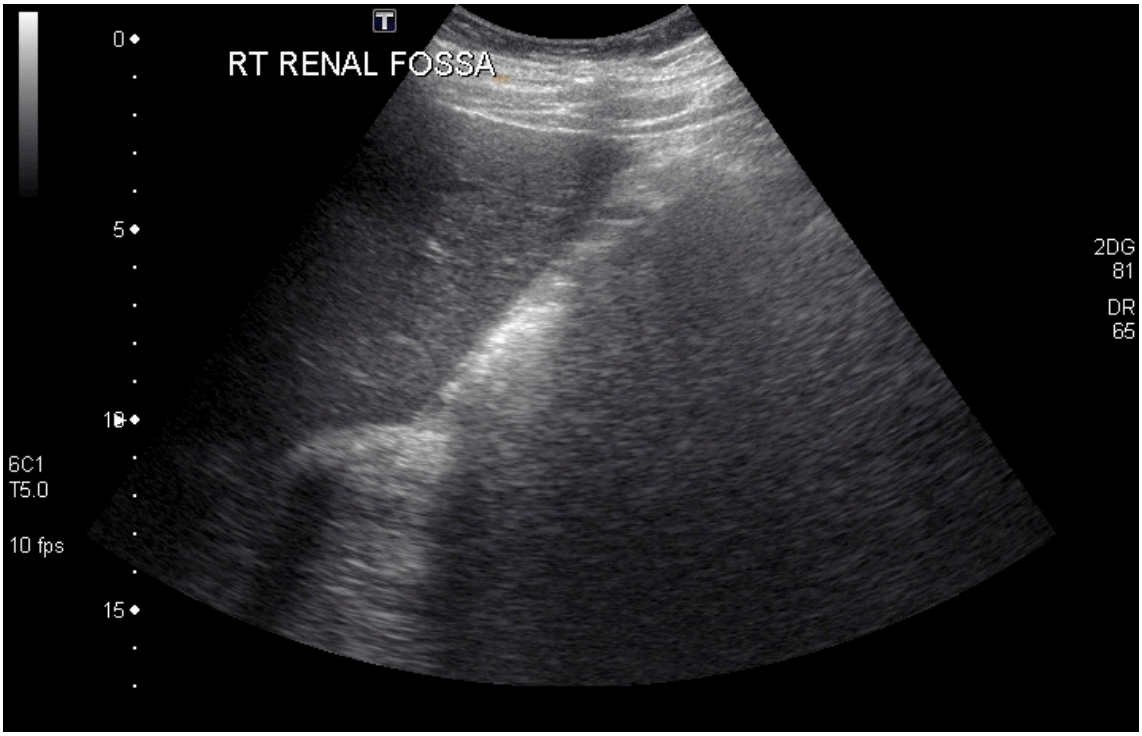

Figure 4: Sagittal scan showing absence of the kidney in the right renal fossa.

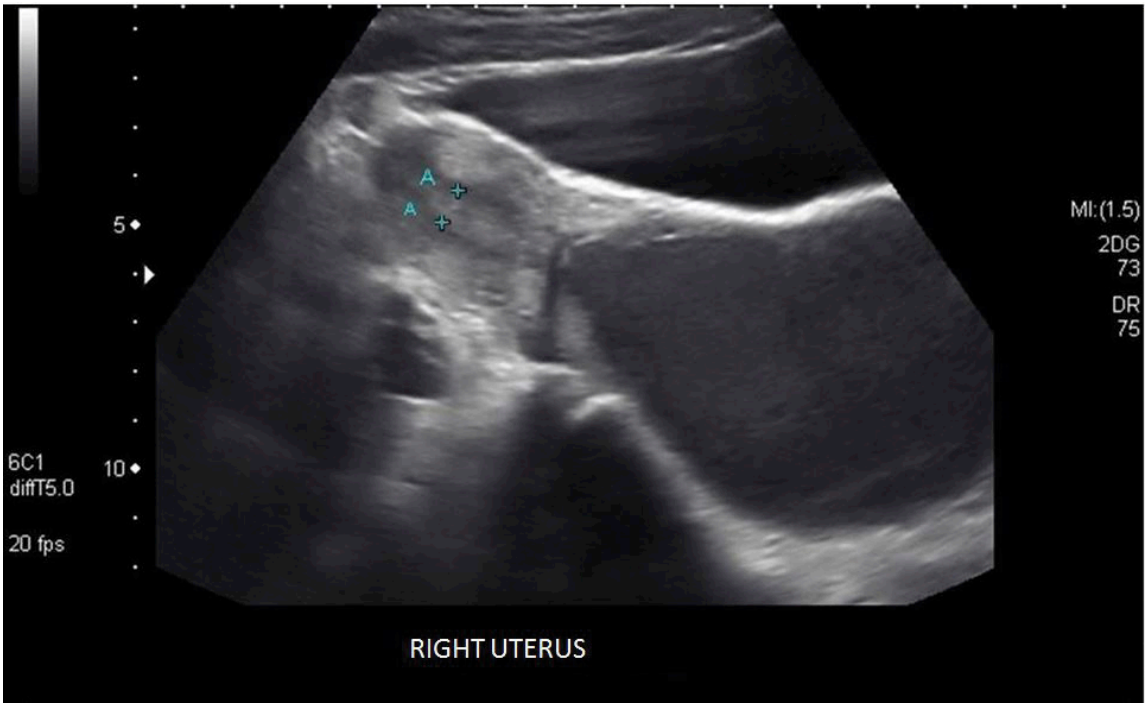

Figure 5: Sagittal scan showing normal right hemiuterus.

The postoperative course was uncomplicated.

2) A 13-year-old girl was referred to our hospital for definitive surgical management. She was known to have a congenitally absent left kidney. She had menarche at the age of 12 years. Her menses were gradually getting irregular with worsening dysmenorrhoea despite progestin therapy. Pelvic ultrasound organised by her general practitioner demonstrated uterine didelphys, haematometra of the left uterine cavity, haematocolpos and absent left kidney. Examination under anaesthesia by her private gynecologist showed obstructed non-communicating hemivagina. Repeat pelvic ultrasound in the Medical Imaging Department confirmed the above findings (Figures 5–8). Examination under anesthesia and hysteroscopy revealed uterine didelphys with left transverse obstructing vaginal septum and haematocolpos. The vaginal septum was excised with drainage of 450 ml of old blood. The postoperative course was uncomplicated.

Discussion

The pathogenesis of OHVIRA syndrome is related to anomalous development of both the paramesonephric (Mullerian) and

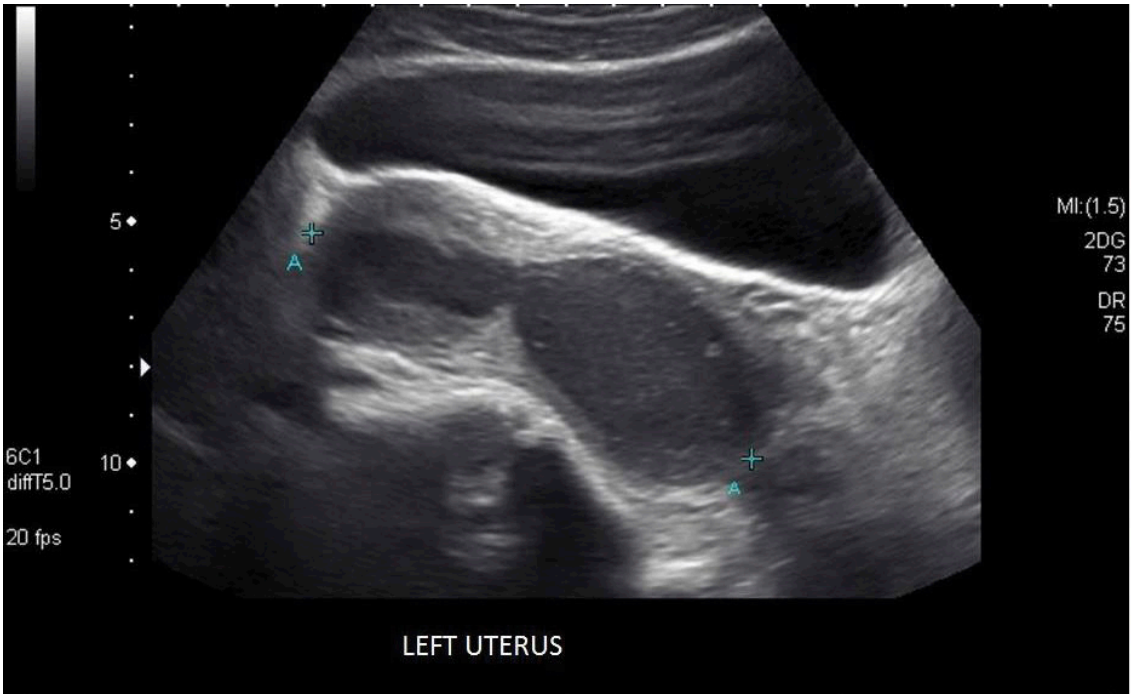

Figure 6: Sagittal scan showing left hemiuterus with distended cavity containing echogenic material.

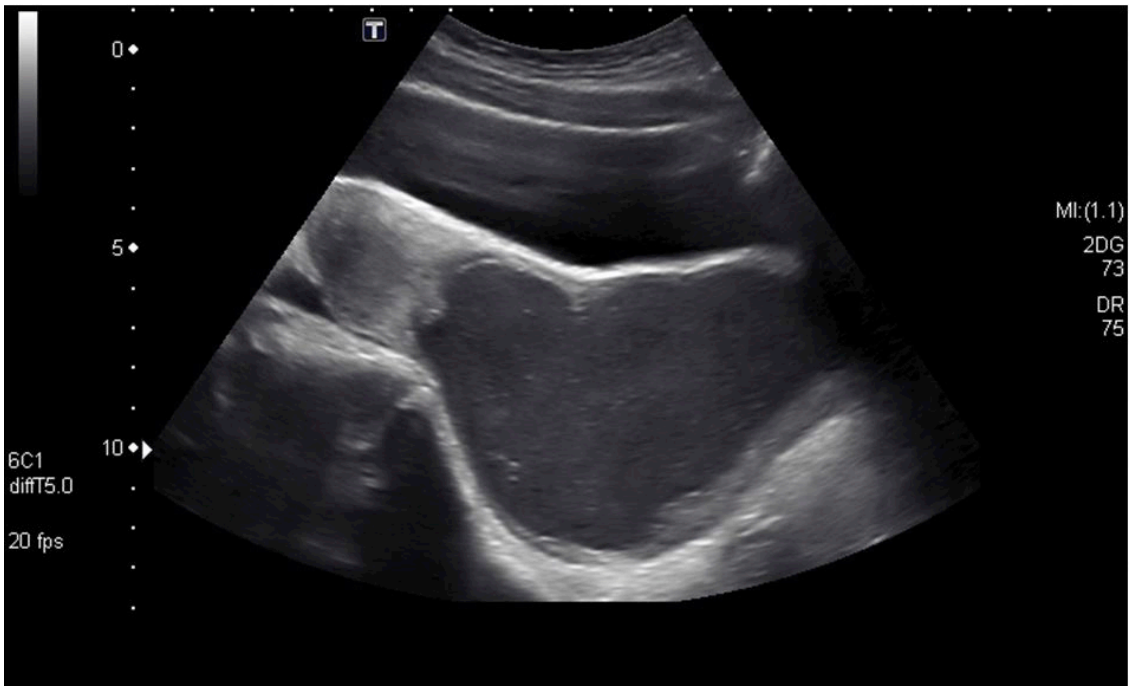

Figure 7: Sagittal scan showing distended vagina containing echogenic material.

mesonephric (Wolffian) ducts.[7] The Wolffian ducts induce normal Mullerian duct fusion and give origin to the kidneys. Abnormal Wolffian duct development leads to unilateral renal agenesis and imperforate hemivagina associated with OHVIRA syndrome.[8] The Mullerian duct on the side where the Wolffian duct is absent is displaced laterally and cannot fuse with the contralateral Mullerian duct, resulting in uterine didelphys. The contralateral Mullerian duct gives rise to a vagina, whereas the displaced Mullerian duct cannot come into contact with the urogenital sinus and centrally forms a blind sac, leading to an imperforate or obstructed hemivagina. There may be an associated longitudinal or transverse vaginal septum and this was seen in the two cases presented. Renal agenesis is the most common associated renal anomaly and is always seen on the same side as the obstructed hemivagina.

OHVIRA syndrome is usually discovered at puberty, shortly after menarche because of increasing pelvic pain, dysmenorrhoea and pelvic mass secondary to haematometrocolpos from retained blood in the obstructed hemivagina.[4] It may remain unrecognised initially as regular menstrual flow from the patent unobstructed hemivagina gives the appearance of normal menses, and dysmenorrhoea, if present, is a common complaint in this age group.[7] Symptom relief of dysmenorrhoea with non-steroidal anti-inflammatory and oral contraceptive medication may also

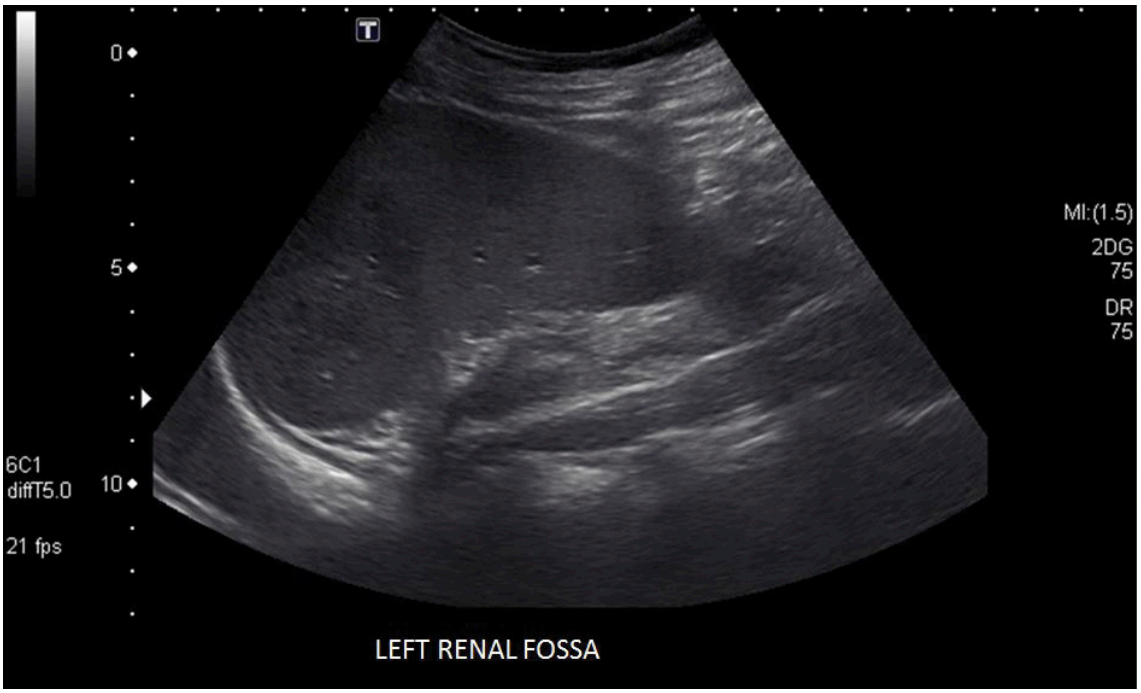

Figure 8: Sagittal scan showing absence of the kidney in the left renal fossa.

lead to a delay in diagnosis.[9] The clinical presentation may be variable and non-specific features such as acute abdominal,[4,10] pelvic[9,11] or vaginal[12] pain, dysuria,[13] urinary retention,[7] vaginal discharge[14,15] and infertility[16] have been reported. The two cases presented were both known to have an absent kidney and this should prompt investigation for Mullerian duct anomalies and vice versa. Associated complications include acute infectious collections and long-term sequelae such as endometriosis, pelvic adhesions, and infertility.[9] Initial misdiagnosis of OHVIRA syndrome and a significant delay from the time of onset of symptoms to diagnosis have been reported with patients undergoing unnecessary and potentially harmful invasive investigations before appropriate surgery was performed.[5,17–19]

Ultrasonography and magnetic resonance imaging (MRI) are used to diagnose OHVIRA syndrome. Ultrasound is quick, cheap, noninvasive and readily available.[7,20] It will generally identify uterine didelphys, haematometrocolpos and unilateral absence of a kidney. The vaginal septum however is difficult to visualise on ultrasound.[7,20] MRI is said to be the imaging modality of choice to correctly diagnose OHVIRA syndrome.[9,10,16,18,21] It is more sensitive in assessing uterine contour, shape of the uterine cavity, thickness and location of the vaginal septum, type of collection in the obstructed hemivagina and coexisting urinary tract malformations.[9] The two cases presented were diagnosed on ultrasound without the benefit of MRI.

Laparoscopy has been advocated as the gold standard for evaluation of OHVIRA syndrome.[2,17] Others suggest that laparoscopy, rather than being performed routinely, be reserved for those cases when the diagnosis is not clear after imaging, when MRI is not available or when there is suspected concurrent intraperitoneal pathology such as endometriosis, adhesions and pelvic infection.[3,9,15] Laparoscopy was performed in one of the two cases presented because of a probable haematosalpinx seen on ultrasound.

The treatment of choice for OHVIRA syndrome is resection of the vaginal septum in order to reestablish the continuity of the obstructed hemivagina.[6,9,10,17,20] This may be performed as a single-stage or two-stage procedure. The single stage procedure involves excising the vaginal septum completely and marsupialising the vaginal cuff. The two-stage procedure involves reducing the haematocolpos first and then excising the remaining septum tissue after a period of wound healing and vaginal remodeling. The latter procedure is considered if there is infection and/or anatomical distortion preventing satisfactory assessment of the vagina for repair.[3] The two cases reported were successfully managed with single stage excision of the vaginal septum.

Conclusion

OHVIRA syndrome should be considered in patients presenting with dysmenorrhoea and a history of renal agenesis. Ultrasound remains a reliable and accurate imaging tool in the diagnosis of this syndrome. Early diagnosis and surgical intervention is important to relieve symptoms, prevent complications and preserve future fertility.

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