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Case Report

Fetal magnetic resonance imaging is a useful modality for prenatal diagnosis of obstructed hemivagina and ipsilateral renal anomaly syndrome

M. Mayumi[1], M. Obata-Yasuoka[1], H. Hamada[1], T. Satoh[1], Y.O. Tanaka[2], K. Masumoto[3]

[1]Department of Obstetrics and Gynecology, University of Tsukuba, Tsukuba
[2]Diagnostic Imaging Center, The Cancer Institute Hospital, Tokyo
[3]Department of Pediatric Surgery, University of Tsukuba, Tsukuba (Japan)

Summary

The authors report the first case of prenatal fetal MRI diagnosis of unilateral hydrocolpos, uterus didelphys, and ipsilateral renal agenesis in a case of obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome. A 33-year-old woman was referred to the present institution because of a fetal pelvic cyst. Detailed ultrasound revealed a female fetus with a retrovesical cyst and right renal agenesis. Fetal MRI confirmed unilateral hydrocolpos, uterus didelphys, and normal intestinal tract of the fetus. Fetal MRI is a useful modality for prenatal diagnosis of OHVIRA as a method of evaluating the pelvic structures and excluding other cloacal anomalies.

Key words: Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA); Herlyn-Werner-Wunderlich syndrome (HWWS); Unilateral hydrocolpos; Cloacal anomaly; Fetal MRI.

Introduction

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome, also known as Herlyn-Werner-Wunderlich syndrome, is a rare syndrome characterized by the triad of uterus didelphys, obstructed hemivagina, and ipsilateral renal anomaly [1-3]. There are three reported cases of OHVIRA diagnosed in the neonatal period [4-6]. In all three cases, renal agenesis and/or retrovesical cyst were detected by prenatal ultrasound and MRI was performed after birth. Here the authors report the first case of OHVIRA diagnosed prenatally by fetal MRI.

Case Report

A 33-year-old woman, gravida 0, para 0, was referred to the present institution at 28 weeks' gestation because of a fetal pelvic cystic mass. Detailed ultrasonographic examination revealed a female fetus with a retrovesical cyst and right renal agenesis. At 35+4 weeks' gestation, MRI was performed for detailed evaluation of the fetal pelvis. MRI confirmed uterus didelphys, retrovesical cyst measuring 23×17×33 mm, and right renal agenesis (Figure 1). Left renal length was 36×20 mm. The cyst was located between the bladder and rectum on T1-weighted image (T1WI), and was connected to the cervix of the uterus didelphys on steady-state free precession (SSFP) image, of which contrast reflected the T2/T1 ratio. Based on these findings, the cyst was thought to originate from the genital tract. In addition, the cyst was low intensity on T1WI and high intensity on SSFP, similar to amniotic fluid, suggesting the cyst contained serous fluid. Therefore the cyst was diagnosed as hydrocolpos: an expanded, fluid filled vagina. No anorectal anomalies were visualised. Thus the prenatal diagnosis by fetal ultrasonographic examination and MRI was OHVIRA syndrome.

The female infant weighing 2,645 grams with Apgar scores of 8 and 9 after one and five minutes, respectively, and umbilical ar-

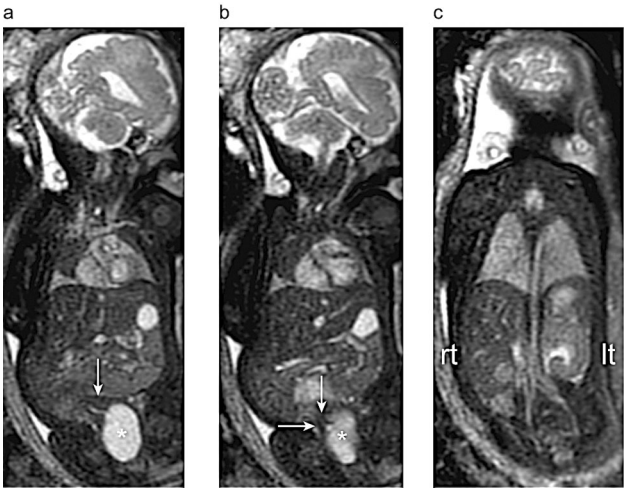

Figure 1. — MRI of the fetal pelvis at 35 weeks of gestation. Steady-state free precession image in the coronal view. (a,b)The uterus didelphys (arrow) is located next to the hydrocolpos (*). (c) The right kidney was not detectable.

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tery pH of 7.332 was vaginally delivered at 39+3 weeks' gestation. The urethral orifice, vaginal orifice, and anus were normally positioned. There was no external malformation and no difficulty with urination. Postnatal ultrasonographic examination on the first day of life revealed uterus didelphys, hydrocolpos, and right renal agenesis. When saline was injected into the neonate's opened vagina, the hydrocolpos was found to be located on the right side of the vagina. Vesicoureteral reflux and fistula between the bladder and uterus were ruled out by voiding cystourethrography.

Discussion

Fetal hydrocolpos is detected by ultrasonographic examination as a retrovesical cystic mass, and most cases are detected in the third trimester of pregnancy or after birth [7], although they are in some cases diagnosed as early as the second trimester [8]. Hydrocolpos results from obstruction at different levels of the vagina, and the fluid accumulation consists of cervical and vaginal secretions that develop secondary to circulating maternal estrogens [9, 10]. Hydrocolpos occurs as a result of transverse vaginal septum, vaginal atresia, imperforate hymen, or cloacal anomaly. When a hydrocolpos is detected, it is important to diagnose any associated cloacal anomaly. Because approximately 40% of patients with cloacal anomalies have a double Müllerian system, most patients with hydrocolpos have bilateral hydrocolpos [11]. In the present case, the authors felt that a cloacal anomaly could be preliminarily excluded because hydrocolpos was unilateral on ultrasound, but it was difficult to evaluate the pelvic anatomy in detail by fetal ultrasonographic examination.

Prenatal MRI diagnosis of fetal hydrocolpos secondary to cloacal anomalies has been reported [7, 9, 12]. Fetal MRI can aid in the diagnosis of cloacal anomalies by demonstrating the absence of meconium in the rectum alongside the bladder on T1WI [9]. The most characteristic finding on T1WI was hyperintense signals indicating the presence of meconium in the rectum. Cloacal anomaly was ruled out based on the presence of meconium in the rectum extending down to the perineum. SSFP revealed that the retrovesical cystic mass was a unilateral hydrocolpos: an expanded fluid-filled vagina connecting to the right-sided hemiuterus as its excellent contrast resolution and faster image acquisition time. Thus the authors could distinguish OHVIRA from a fistula between the vagina and rectum.

Capito *et al.* [13] reported that the signal of bladder contents is different than that of hydrocolpos on T2-weighted images in fetuses with isolated genital obstruction. In the present case, the signals of the bladder and hydrocolpos were similar on SSFP imaging, and fetal MRI could not exclude communication between the urinary tract and the vagina. For this reason, the authors performed postnatal voiding cystourethrography to exclude vesicovaginal fistula. Unilateral renal anomaly is one of the main symptoms of OHVIRA. Compensatory hypertrophy of kidney is detectable in 44.4% of the fetuses with unilateral functioning kidney [14]. In the present case, compensatory hypertrophy of left kidney was not observed despite the absence of right kidney.

MRI can demonstrate Müllerian duct anomalies, and lead to the diagnosis of OHVIRA at puberty [15]. MRI evaluation of the genital tract is recommended in all girls with known renal abnormalities before the onset of menstruation [16]. In the present case, the combination of ultrasound findings of retrovesical cyst and unilateral renal agenesis in a female fetus was indicative of OHVIRA, and prenatal MRI confirmed the diagnosis by demonstrating unilateral hydrocolpos, uterus didelphys, and a normal rectum. Thus, fetal MRI is a useful modality for prenatal diagnosis of OHVIRA as a method of evaluating the pelvic structures and excluding other cloacal anomalies.

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Corresponding Author:
M. MAYUMI, M.D.
Department of Obstetrics and Gynecology
Faculty of Medicine, University of Tsukuba
1-1-1 Tennodai,
Tsukuba, Ibaraki 305-8575 (Japan)
e-mail: sekizaka-tuk@umin.ac.jp