Complete septate uterus, obstructed hemivagina, and ipsilateral adnexal and renal agenesis in pregnancy

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Most cases of double uterus with obstructed hemivagina and ipsilateral renal agenesis were diagnosed at adolescents after menarche. This is the first reported case of complete septate uterus with obstructed hemivagina and ipsilateral renal agenesis in addition to ipsilateral agenesis of fallopian tube and ovary in which the diagnosis was delayed until pregnancy. The pregnancy was uneventful in spite of intermittent vaginal spotting. During the cesarean section, the septum of the uterus was resected and about a 3-cm×3-cm window was made on the vaginal septum to allow an opening for the obstructed vaginal discharge. We followed the patient up for one and half years, and she has not had symptoms such as dysmenorrhea or abnormal vaginal bleeding.

Keywords: Hemivagina; Ovarian agenesis; Pregnancy; Renal agenesis; Septate uterus

Case report

A 30-year-old nulligravida was referred to our hospital because of an uterine anomaly in pregnancy. She presented at 5+4 weeks of pregnancy without abdominal pain or vaginal spotting. The patient attained menarche at 14 years of age, and had regular periods and a history of severe dysmenorrhea. She had been given a diagnosis of left renal agenesis at birth. However, she had not visited any hospital for evaluation of her dysmenorrhea with unilateral renal agenesis. There was no other past history. A systemic examination was normal. The abdomen was...
soft and nontender on examination. Speculum examination revealed a blind bulging pouch at the left side of the vagina. A small cervix in the right was seen lateral to the bulging pouch.

Two-dimensional (2D) ultrasonography (US) revealed a complete septate double uterus with diverging cornua. An 8-mm sized gestational sac was seen in the right horn. A dilated hypoechoic left vagina was evident (Fig. 1). The right ovary was visualized normally, but the left ovary was not visualized. Three-dimensional (3D) US revealed the complete septate uterus with a tiny gestational sac in the right uterus and decidual reaction in the left uterus. The markedly dilated left vagina bulged into the right vagina (Fig. 2). The left maternal kidney was absent and the right kidney was normal. Our diagnosis was a complete septate uterus with obstructing hemivagina and ipsilateral adnexal and renal agenesis. Intermitting vaginal spotting had been present since about 14 weeks of gestation. However, the patient and her fetus were otherwise normal.

She visited the St. Vincent’s Hospital for spontaneous rupture of membrane at 38\(\frac{1}{2}\) weeks of gestation. She gave birth by cesarean section because the left bulging vaginal pouch made a vaginal birth difficult. During the cesarean section, we confirmed that the uterine anomaly was a complete septation that continued to the vagina. There was thickening and obliteration of the cul-de sac and adhesion between the posterior wall of the uterus and descending colon. The right ovary and tube were normal, but the left ovary and tube were absent. The septum of the uterus was resected and the left cervix was able to be palpated and visualized. Through a vaginal

approach, the vaginal septum was incised and a window of about 3 cm×3 cm in size was made by suturing the incision site. About 200 mL of chocolate colored sticky fluid was drained.

The baby’s weight was 2.98 kg and the 1- and 5-minute Apgar score was 8 and 9, respectively. The patient was discharged after 4 days without complications. She was followed-up for 1.5 years, with no complaints of dysmenorrhea and abnormal vaginal bleeding after the first postpartum menstruation. The vaginal septum window has remained intact with no fluid collection in the left vagina.

Discussion

This case was a complete septate uterus and obstructed hemivagina with ipsilateral adnexal and renal agenesis, which is a rare variant of the HWWs or obstructed hemivagina with
ipsilateral renal agenesis syndrome (OHVRAs). Mullerian duct anomalies have an incidence of 2% to 3%, and HWWs or OHVRAs constitute 0.16% to 10% of these Mullerian duct anomalies [3]. HWW syndrome is classically a didelphys uterus with obstructed hemivagina. However, variable anatomic structures such as a combination of a bicornuate bicolis uterus or septate uterus with unilateral cervical atresia or septate cervix may be involved in this syndrome. The classic variant of the HWW syndrome is 77% of the variants, bicornuate 11.5%, and septate 11.5% [4]. Ipsilateral renal agenesis is a very frequent accompanying anomaly because these conditions are the result of a disruption in the continuum of embryological development of the urogenital system [4]. It is usually discovered shortly after menarche because of cyclic pelvic pain, a pelvic mass and a foul-smelling discharge. These symptoms or signs are initially easy to misdiagnose as other diseases such as endometriosis, appendicitis or pelvic mass. Such misdiagnoses often delay the correct diagnosis [5,6]. The patient in this case was not diagnosed until she was pregnant in spite of known unilateral renal agenesis. Therefore, awareness of these congenital uterine anomalies is necessary, and an adequate investigation is essential. Adequate radiologic studies such as US and/or magnetic resonance imaging (MRI) are mandatory to assess the anatomic confirmation. US mostly allows the correct diagnosis by showing uterovaginal duplication, hematocolpos or hematometrocolpos, and the absence of the ipsilateral kidney [7]. 3D US can improve diagnostic accuracy. MRI provides more detailed information regarding the uterine morphology, continuity with each vaginal channel, and the bloody nature of the contents [8]. Our case was diagnosed comparatively easily at the first trimester of pregnancy during the first a vaginal ultrasound scan. We obtained information about the known unilateral renal agenesis and dysmenorrhea since menarche. 2D US showed a septated uterus and obstructed hemivagina filled with fluid. 3D US provided a more sophisticated image than 2D US.

The treatment of patients with uterine duplication associated with blind hemivagina is resection of the obstructing vaginal septum as completely as possible using scissors, scalpel, conventional scalpel diathermy or resectoscope [9,10]. We carried out marsupialization during the cesarean section; complete removal of the vaginal septum was very difficult because of the bleeding and very thick vaginal septum. Incision alone of the septum may lead to the development of hematocolpos or pyocolpos after spontaneous closure. Marsupialization was recommended initially for this patient. One month later, the remaining vaginal septum is usually removed, particularly because if the obstructed hemivagina has reached the hymenal ring [11]. We did not carry out the remaining vaginal resection because the patient did not complain of dysmenorrhea or abdominal pain after the operation. Therefore, marsupialization may be an adequate treatment for this case.

Complex anomalies involving the urogenital system and the ovaries occur less frequently than those not involving the ovaries. Whereas the urogenital organs involving the fallopian tubes originate from the Mullerian duct, the ovaries originate from the germ cells. A case of septate uterus with left fallopian tube hypoplasia and ipsilateral ovarian agenesis has been reported [12]. A hypothesis proposed for the ipsilateral agenesis of ovary with Mullerian duct anomalies is that a unilateral defect in the development the urogenital ridge could have profound ramifications on the future gonad on which it would sit [12].

Such a patient who has complex anomalies of the uterus, vagina and ipsilateral kidney in addition to anomalies of the ipsilateral fallopian tube and ovary can safely maintain her pregnancy and deliver a healthy infant by cesarean section. The patient’s dysmenorrhea can be managed by proper treatment of the septate uterus and obstructed hemivagina during the cesarean section.

Mullerian duct anomalies are associated with adverse reproductive outcomes such as recurrent pregnancy loss, malpresentation, intrauterine growth restriction and increased rate of cesarean section, preterm labor, premature rupture of membranes, retained placenta, postpartum hemorrhage, and increased fetal mortality [13,14]. Especially uterine septa have the worst reproductive outcome in terms of pregnancy loss rate and preterm labor rate, and the fetal survival rates without any surgical treatment in the septate uterus is 76% [15]. Poor obstetrical outcomes may result from reduced uterine cavity volume, impaired distension, abnormal myometrial and cervical function, or abnormal endometrial development. Obstructed hemivagina can inhibit the labor progress. However, none of these adverse reproductive outcomes occurred in the present case, which yielded a favourable outcome.

**Conflict of interest**

No potential conflict of interest relevant to this article was reported.
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