A case of obstructed hemivagina and ipsilateral renal anomaly syndrome (OHVIRA) with a single uterus, unrecognized before labor and followed by an intrapartal rupture of obstructed hemivagina

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Objective

Müllerian duct anomalies are congenital anomalies of the female genital tract resulting from non-development or non-fusion of the Müllerian ducts, or failed resorption of the uterine septum. The acronym for obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) was created to describe patients with an obstructed hemivagina and ipsilateral renal anomaly and enables inclusion of other uterine anomalies except uterus didelphys. The aim of this article is to present a rare case of OHVIRA syndrome with single uterus, uterine septum (previously resected by hysteroscopy) and renal agenesis, unrecognized before labor and followed by an intrapartal rupture of obstructed hemivagina.

Methods

A 30-year old gravida 1 para 0 was admitted to the maternity ward in the early stage of labor at 41+2 weeks. In her medical history, right kidney agenesis was noticed in her childhood. She had laparoscopy and hysteroscopy procedure in 2012 on account of large uterine septum and primary infertility. A hysteroscopic resection of uterine septum went uneventfully. Magnetic resonance imaging (MRI) was performed preoperatively and renal agenesis was confirmed but without any vaginal deformation. Vaginal examination had not revealed any suspicion of vaginal anomaly. Spontaneous pregnancy was achieved promptly after the procedure. She had regular antenatal visits with normal course of pregnancy. On admission all fetal and maternal finding were normal. The baby was in cephalic position and was at the entrance of pelvis. Cervix was half effaced and 2 cm dilated. The following morning, cervix was fully effaced and 7 cm dilated. Woman opted for amniotomy and augmentation of labor. The fetal findings were normal. On the next exam, the fetal head was below the level of spines, cervix was fully dilated. At the right side, above the birth canal, was some unusual form of vaginal mucosa bulge. Immediately after that, the episiotomy was carried out and the woman gave a birth to a live female newborn, 3610g/51cm, the Apgar score 10/10. After delivery of child and placenta, a right-sided vaginal rupture with intact cervix was observed (Fig. 1). We concluded that there was a rupture of dextrolateral obstructed hemivagina. Then the complete wall of this obstructed hemivagina was resected and the entire injury was sutured in epidural analgesia (Fig. 2). The vaginal and rectovaginal findings after this surgical procedure were satisfactory. Hospital course went orderly. One month later, normal vaginal findings and the regular healing of wound were found. Histopathology confirmed vaginal tissue in a resected sample.

Results

Various symptoms, included in OHVIRA acronym, are a result of different morphologic variants included in this syndrome. We did not find OHVIRA cases in the literature with a single septate uterus/single cervix, associated with obstructed hemivagina and with clinical presentation such in this case. The classic clinical course of this syndrome is an adolescent girl with pelvic pain who, on gynaecologic examination, is found to have a vaginal bulge. Some of women have hematocolpos and typically early clinical presentation due to stasis of menstrual blood, oligomenorrhea and pelvic pain. None of these symptoms was not present our patient. Most likely in our case, the absence of communication between the single cervix/patent vagina and the obstructed hemivagina prevented forming mucocolpos or hematocolpos. The diagnosis of this condition can difficult due to its rarity and heterogeneous presentation. Relevant radiologic studies confirmed that MRI is obligatory when diagnosing OHVIRA patients. We think that non-communicating obstructed hemivagina in our patient, without significant liquid storage, made MRI diagnosis of such a rare variant of OHVIRA syndrome ineffective. In our case, it was necessary to resect complete tissue of obstructed hemivagina and to create a single vagina promptly after delivery due to a huge intrapartal rupture, which totally deformed the whole of preexisting vagina. Favorable condition for us was that this woman bled little.

Conclusion

The presentations and effects of congenital anomalies of the female genitourinary tract are enormously variable. For understanding various presentations of Müllerian anomalies, obstetrics knowledge of basic embryology is needed. In this case report we focused on its rarity in a sense of morphology and intrapartal clinical presentation and treatment. Certain Müllerian anomalies could be easily recognized during standard diagnostic procedures but, on the other side, could represent a very hard diagnostic task as showed in our case, resulting in totally unrecognized obstructed hemivagina within an extremely rare variant of OHVIRA syndrome. This obstacle delayed final diagnosis and therapy of our patient till the beginning of spontaneous labor. A final decision about managing labor in such rare cases of OHVIRA syndrome is difficult, because of lack of guidelines. The most important thing regarding vaginal delivery is the presence of experienced obstetricians in labor wards, because serious risks lay in unrecognized obstructed hemivaginas during labor, including postpartum hemorrhage from a massive vaginal rupture.