Case Report

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OHVIRA syndrome obstetric patient for lower segment caesarean section: peri-operative considerations

Abstract

OHVIRA syndrome is a rare congenital developmental condition manifested by uterus didelphys, lower genital tract obstruction and unilateral renal anomaly. We report a rare case of a full term obstetric patient with OHVIRA syndrome, posted for elective lower segment caesarean section. Peri-operative anaesthetic considerations in a patient with this syndrome have been described. The importances of systemic preoperative assessment and investigations to rule out associated anomalies have also been highlighted.

Keywords: OHVIRA Syndrome, LSCS, Uterus didelphys, Mullerian and Wolfian ducts anomalies; Renal agenesis.

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Introduction

Mullerian duct anomalies (MDA) are developmental defects involving female genital system with varying presentations ranging from undiagnosed very mild segmental anomalies to unicornuate, bicornuate, didelphic uterus and to even absence of uterus and vagina. The incidence is 2-3%.[1] Renal agenesis occurs in these patients as a result of simultaneous arrest of mullerian and metanephric ducts at 8 weeks of gestational age. These patients usually presents at menarche with haematometra and haematosalphinx. We report a rare case of full term pregnancy with OHVIRA syndrome (Obstructed Hemi Vagina with Ipsilateral Renal Anomaly) with congenital uterine didelphys posted for elective lower segment caesarean section (LSCS).

Case report

A 24-year old primigravida female presented to our institute with 37weeks pregnancy in the right horn of a didelphys uterus, posted for LSCS. The patient had history of gestational hypothyroidism and gestational hypertension controlled on oral thyroxine 25mcg once daily and labetalol 100mg twice daily respectively. She was a known case of OHVIRA syndrome. She had previous history of laparotomy and drainage of haematosalphinx and hamaetometra followed two days later by fimbrioplasty and vaginal septal excision under general anaesthesia. Patient conceived spontaneously. Pre-operative evaluation and investigations including 2D echocardiography were within normal limits. Magnetic resonance imaging (MRI) of the abdomen revealed absent left kidney (**Figure 1**: MRI abdomen of the patient). Adequate packed cells were arranged before shifting patient to the operating room. All standard American society of Anesthesiology recommended monitoring were initiated along with invasive radial arterial line, pre-induction. Subarachnoid block using 25G Quincke's needle with patient in sitting position, at L3-L4 Inter-space (1.8ml of 0.5% bupivacaine (Heavy) and 10mcg fentanyl) was given and sensory block upto T6 level achieved. A healthy (2.975 kg) baby with APGAR score of 10 was delivered. Intra-operative course was uneventful. The uterus contracted well with oxytocin infusion post-placental delivery (Figure 2: Photo showing right horn of the uterus). Post operatively, patient was hemodynamically stable and later shifted to ward in a stable condition (Figure 1&2).

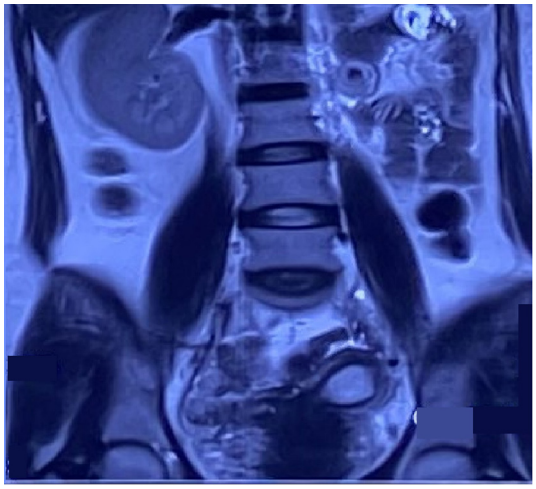

Figure 1 MRI image of abdomen showing single right kidney with empty left renal fossa.

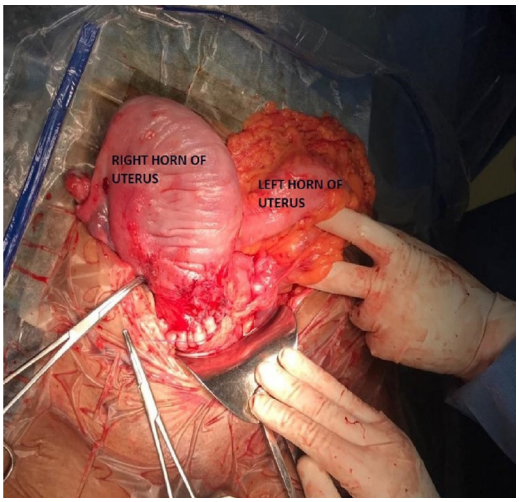

Figure 2 Showing left and right horns of uterus. Uterotomy incision on right hemiuterus can be seen. Non communicating left hemiuterus can also be seen.

Discussion

Obstructed hemivagina and ipsilateral renal agenesis syndrome is a rare congenital anomaly, which is often missed clinically and is difficult to diagnose and a prompt and accurate diagnosis with appropriate surgery aimed at conserving the uterus is required to relieve the symptoms, to prevent complications, and to preserve the sexual and reproductive function.

OHVIRA syndrome or Herlyn-Werner–Wunderlich syndrome (HWWS) is a congenital developmental condition involving Mullerian and Wolffian ducts, manifested by triad of uterus didelphys, lower genital tract obstruction and unilateral renal anomaly. Mullerian fusion defect causes two completely separated uterine cavities, one/two cervices and the presence of a complete/incomplete vaginal septum. It constitutes about 0.16-10% of all mullerian duct anomalies.[2] Since 2007 (other uterine anomalies, such as the septate uterus, being reported), the acronym referring on two features only (the obstructed hemivagina and ipsilateral renal anomaly – OHVIRA) gained a wide acceptance.

During the fusion phase, the Mullerian ducts fuse in their distal portion in order to form the uterus, the cervix and the superior vagina.[3] The abnormalities occurring in this period lead to uterine duplicity, renal agenesis, blind hemi-vagina. Uterus didelphys is the result of a severe fusion defect and it is characterized by two completely separated uterine cavities, one/two cervices and the presence of a complete/incomplete vaginal septum. This process evolves between gestational weeks 6 and 9 of embryogenesis.

Congenital malformations of the female genital system have varied presentations affecting all anatomic levels: the uterus, the cervix and the vagina. Congenital uterine anomalies are associated with unfavorable reproductive/ neonatal outcome such as infertility, recurrent abortion, malpresentations, preterm labor, abruption placenta and cervical incompetence. Yet, there are authors reporting that these uterine malformations are usually asymptomatic.

All non-obstructive OHVIRA cases, including major ones, may have no symptoms during pregnancy. They are at increased risk of small for gestational age fetus, pre-eclampsia, increased incidence of caesarean delivery and low birth weight babies.[4] There is predominance of pregnancy in right hemi-uterus in case of single pregnancy in uterus didelphys, as is seen in our case.[5] To this date, the outcome of pregnancy in women with didelphys uterus is considered jeopardized. There are scarce reports on the association between OHVIRA and near term/term pregnancies.

OHVIRA syndrome is also associated with aortic stenosis. Severity of aortic stenosis and assessment of left ventricular function needs 2D Echocardiography (trans-thoracic echocardiography/ trans esophageal echocardiography).[6]

Also, presence of fused vertebrae which makes neuraxial anaesthesia complicated.[6] Renal functions should be assessed preoperatively as the single kidney in case of renal agenesis may malfunction due to various mechanical/ pathological processes. Multiple renal anomalies like renal duplication, multi-cystic dysplastic kidney are associated with this syndrome, but renal agenesis is the most common.[7] These patients are usually diagnosed on investigating abdominal pain at menarche, due to collection of blood in non-communicating uterus. Magnetic resonance imaging, three-dimensional transvaginal ultrasonography, hysterosalpingography and computer tomography have been used as investigating modalities. Many studies report that MRI is the best diagnostic method, having an accuracy of almost 100%.[8]

These patients may present to anaesthesiologist for emergency or elective caesarean section or for non-obstetric surgeries. OHVIRA syndrome, a rare congenital anomaly, which may be missed clinically. Appropriate systemic preoperative assessment, investigations to rule out the presence of associated anomalies along with proper operation theatre facility, neonatal care unit, preparation for anticipated peri-operative complications and maternal as well as neonatal resuscitation are needed.

Conclusion

OHVIRA syndrome is a rare congenital urogenital anomaly which may present with wide range of clinical presentation requiring surgical intervention. In this case we describe the anaesthetic management of a patient of OHVIRA syndrome with full term pregnancy posted for lower segment caesarean section.

Acknowledgments

None.

Conflicts of Interset

None.

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