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CASE REPORT

A Rare Case of OHVIRA Syndrome with Full Term Pregnancy – A Case Report

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ABSTRACT

Herlyn-Werner-Wunderlich syndrome or OHVIRA syndrome refers to obstructed hemivagina/hemicervix with ipsilateral renal anomaly. The incidence of Mullerian malformation is 2-3% amongst which OHVIRA syndrome is the least common type of Mullerian malformations which results from defective fusion of Mullerian ducts.

Here we report a rare case of such syndrome who presented with full term pregnancy in active phase of labor. A young primi of 19 years old, 39 weeks pog with breech was referred from Primary health care centre for breech presentation. On examination, obstructed hemi vagina was noted. Emergency cesarean section was done and intraoperatively, uterine diadelphy was noted. The diagnosis of OHVIRA syndrome is usually made during the period of menarche where patients present with cyclic abdominal pain, mass on pelvis and infertility. However, they may also present rarely with malpresentation during pregnancy and complications during labor.

Early diagnosis of this syndrome with appropriate surgical management of resection of septum can aid in early management of obstetric cases with significant decrease in maternal and neonatal complications by early referral and management.

Key-Words: *Breech presentation, hemivagina, herlyn-werner-wunderlich syndrome, mullerian anomaly, obstructed renal agenesis, uterine didelphys*

INTRODUCTION

Development of the female reproductive tract - uterus, fallopian tubes, and the upper one-third of the vagina is from the müllerian/paramesonephric duct, and the lower two-thirds of the vagina from the urogenital sinus.[3] Mullerian duct anomalies can occur in any stages of developmental failure such as agenesis, hypoplasia, or fusion defect in one or both. Mullerian abnormalities can be classified according to American Fertility Society into the following types:

1. Hypoplasia/agenesis
2. Unicornuate
   -Non-communicating, cavitary horn
   -Non-communicating, non-cavitary horn
   -Communicating, cavitary horn
   -No horn
3. Didelphys
4. Bicornuate
5. Septate (Müllerian duct anomalies: From diagnosis to intervention)[4]

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Herlyn-Werner-Wunderlich syndrome or OHVIRA syndrome refers to obstructed hemivagina/hemicervix with ipsilateral renal anomaly which falls on class III of AFS classification.

CASE

A 19 Years old young primigravidae presented to our obstetric emergency room with the chief complaints of pregnancy of 39 weeks and pain abdomen which was severe and intermittent in nature over the lower abdomen and back radiating to the inner thighs, occurring at regular intervals and a sense of bearing down for two hours. She was a booked case of primary health centre and was referred for breech presentation. On examination, her general condition was fair, her vitals were stable, on per abdomen examination, uterus was term size, longitudinal lie, breech and the fetal heart sound was 130 bpm and regular. On per speculum examination – cervix was not visualized, there was a fleshy tissue which obliterated the vagina over right side. On per vaginal examination there was a band like fleshy tissue extending across the right side obliterating the vagina and on further manipulation by pushing the tissue aside, cervical opening of 7 cm dilatation was found on the left side through which the presenting part – rump was palpable. Cervix was soft, effacement was 70%, station was 0, bag of membrane was absent and liquor was clear. Emergency Cesarean section was done, intraoperatively frank breech was noted and baby was delivered by breech extraction. A healthy live female baby of 2.9 kg with AS1 = 9/10 and AS5 =10/10 was delivered. Intraoperatively two uterine fundus was noted with baby in the right horn with fallopian tubes and ovaries present with each uterine fundus. Post operative recovery was uneventful and patient was discharged on post operative day 4. Ultrasonography of abdomen and pelvis was done on her post – operative day 4 and absence of left kidney was noted with mild hydronephrosis of right kidney.

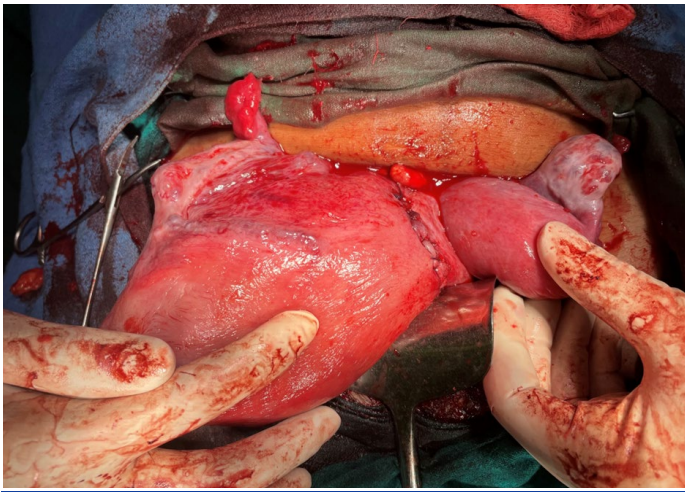

Fig.1 : Uterine diadelphy with incision mark on lower uterine segment on the right side. There are two ovaries and two fallopian tube each on both side of the uterus .

Fig.2: Uterine diadelphy with incision mark on lower uterine segment on the right side. There are two ovaries and two fallopian tube each on both side of the uterus.

DISCUSSION

The incidence of OHVIRA is 0.1%–3.8%.[5] Diagnosis of this syndrome is usually made at puberty with nonspecific symptoms such as acute or chronic pelvic pain, dysmenorrhea, and palpable mass due to the associated hematometra or hematocolpos, which results from retention of menstrual blood behind the obstructed vagina. Radiological diagnosis is by ultrasonography which is the first line of investigation and a 3D USG has 93% sensitivity and 100% specificity in the assessment of Mullerian duct anomalies. The gold standard investigation is MRI with 100% accuracy.[6]

Generally, women with Mullerian anomalies tend to have higher rate of fertility issues and obstetric complications. Obstetric complications such as spontaneous miscarriage, preterm birth, Intra uterine growth retardation and malpresentation are anticipated in these patient groups. Despite that, OHVIRA syndrome has good obstetric prognosis with 87% of pregnancy rate and the live birth rate of 77%.[7]

Our patient presented with all three signs of OHVIRA syndrome. She had obstructed hemi vagina, with absence of left kidney with hydronephrosis of the right kidney. She presented with classic breech malpresentation of fetus in active stage of labour. Although right renal agenesis is more common, left renal anomaly has been noted in a good number of cases. Since, she had renal agenesis on left side the rarity of this case is justified.

Usually, presentation of this syndrome in at an early age which directs the management towards the symptoms presented such as hematocolpos and hematometra for which resection of the transverse vaginal septum is done along with drainage of the collected blood. In the past, hemihysterectomy was also done for obstructed hemiuterus which is now not preferred owing to the fact that the incidence of conceiving has been found to be equal for both the hemi uteruses with good perinatal outcome.[8]

Malpresentation is a fairly common complication of Mullerian anomalies. Acc to Ma S, the occurrence of malpresentation in Mullerian anomalies is 28.4%[9] which is much higher than the general population of 2.5 % in a study done by Maskey S.[10] The prevalence of breech presentation is 3-4 % in general population.[11] The incidence of malpresentation in Mullerian abnormality itself

has yet to be studied. The rate of cesarean delivery in cases of OHVIRA syndrome is 84 %.[6]

Our patient presented with breech presentation and obstructed hemivagina in active stage of labour, we opted for cesarean delivery.

CONCLUSION

The diagnosis of OHVIRA syndrome is very challenging in rural setting as it requires specialized health care services including pelvic examination and radiological diagnosis. Malpresentation is one of the common obstetric complications among patients with Mullerian anomalies. The early diagnosis and treatment can significantly decrease the maternal and perinatal morbidity and mortality.

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