

A Case Report on Ohvira Syndrome

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Introduction

A 24 years old nullipara women was admitted in gynaecological ward with a history of whitish vaginal discharge for 6 months. On assessment her BP was 120/70 mm hg ,and she was conscious and oriented. Then she undergone investigations like USG and confirmed OHIVRA SYNDROME. The findings are complete septate uterus, OHVIRA syndrome, posterior vaginal wall cyst, endometrial left ovarian cyst, and bilateral hydrosalphous hematocolpos left side. and Hb is 12.9 g/dl, on 4-6-2024. she already undergone a anal transposition (10th day of birth and having history of spina bifida occulta with mid scoliosis) and in 2023 under gone laproscopic left ovarian cystectomy and appendectomy. At present she undergone septal resection under general anesthesia on 6//2024.

Investigations

Blood grouping is B positive, urea is 19, creatinine-0.6, potassium 3.8 mEq/L, sodium-138 mEq/l, random blood sugar is 81 mg/dl, TSH – 3.49mIU/l. At the time of admission the examination findings is – Cervix on right, bulge on left side of vaginal. Per vaginally the exact uterine size not made out.

Preoperative care

NPO from 12am, administered Tab.dulcolax (2tabs) @ 10pm if not passes stools suppository and Inj.xylocaine 0.1 cc ID, Inj.DT 0.5ml IM, Inj.suppacel 1.5gram IV stat before surgery, Inj.metrogel IV 400 mg before surgery and 1pint PRBC cross match, ensured the consent forms, parts preparation was done.

Intraoperative care

Position – lithotomy, Per speculum examination done. bulge was noted in the left side. Needle inserted, purulent material aspirated. Incision was made on the same area,20ml of purulent material left out. septum identified and resected. Upper and lower edges of septum sutured.

Postoperative care

Patient is on NPO till 3pm given clear fluids. TPR chart maintained BP chart Q ½ hourly for 4 hours. I/O chart Q1 hourly maintained, Continuous bladder drainage removal @ 6am tomorrow, Watch for bleeding per vagina.

DVT prophylaxis was done.

Heparin 10,000, intravenously, DVT stockings, sequential compressive device.

Herlyn–Werner–Wunderlich syndrome, also known as **OHVIRA** (obstructed hemivagina and ipsilateral renal anomaly) is an extremely rare syndrome characterized by a congenital birth defect of the lower abdominal and pelvic organs. It is a type of abnormality of the Mullerian duct. In most cases, OHVIRA presents as a double uterus with unilateral obstructed (or blind) hemivagina and ipsilateral renal agenesis (or renal anomalies). It can also affect the urethra, urethral sphincter, ureters, bladder and spleen.

Although the true incidence is unknown, it has been reported to be between 0.1% and 3%.

Symptoms

Although there are no specific symptoms for this condition, common complains include progressively increasing pelvic pain during menstruation and hematocolpos due to the buildup of blood in the body. Other symptoms may include swelling of the abdomen, nausea and vomiting during menstruation, and pelvic pain. Fertility may also be affected.

The condition often leads to an increased need for C-sections due to the smaller uteruses.

Diagnosis

The condition is often diagnosed through an **MRI** or ultrasound. Consulting a specialist (in this case a gynecologist) is recommended.

Treatment

Symptoms can be ameliorated with medication and surgery. In most cases, the blind hemi-vagina is opened, and the fluid drained. In adolescents particularly, vaginoscopic incision of the oblique vaginal septum is a viable option.

Pregnancies in women with OHVIRA are categorized as high risk due to the size and shape of the uteri and cervixes as well as the reduced kidney function. In addition, women with undiagnosed OHVIRA can experience genital bleeding during pregnancy and may require hospitalization. Expectant mothers are often managed with cervical sutures and C-sections to prevent fetal distress during labour.

Fertility sparing surgery

Early diagnosis in OHVIRA syndrome is very important to avoid delay in management, and associated complications (i.e. pelvic infection, endometriosis, and infertility). The mainstay treatment is surgery, using a vaginal approach to remove the septum. However, OHVIRA presents mostly early during adolescence, when most patients are still virgo, precluding the vaginal approach. In such patients, minimally invasive approaches are preferred for symptomatic relief. Traditionally, OHVIRA syndrome was best treated with transvaginal resection of the obstructed septum, followed by anastomosis of the vaginal mucosa, which can be extremely difficult to do in virgin girls due to the tight vaginal opening. Moreover, this requires hymen disruption, limiting this approach in patients refusing to lose their hymen integrity.

In the case of virgin patients who wish to maintain their virginity, hysteroscopic resection of the septum under transabdominal guidance has been reported. Dissecting the vaginal septum made the space wider and the ipsilateral cervix visible. Hysteroscopic resection of the obstructed septum is a fast and simple way to preserve the hymen. Accuracy is improved with the use of the transabdominal US, facilitating the procedure in case of small hematocolpos.

Another alternative option to the traditional vaginal approach while preserving the hymen is the use of transrectal ultrasound (TRUS)-guided vaginoscopic septoplasty, with the use of a novel ultrasound technique, the three-dimensional saline-solution infusion contrast sonovaginocervicography (3D-SVC).

The standard treatment of OHVIRA syndrome is usually done through the vagina, requiring a two-step or a single-step vaginoplasty approach. A single-step vaginoplasty can be challenging, especially in young girls with narrow vaginas. Thus, the two-step approach can be done as an alternative.

From this overview, a combination of hysteroscopy and laparoscopy as a surgical approach for OHIVRA syndrome management could be considered feasible, except in few cases. Hysteroscopy without laparoscopy should be used if MRI did not reveal any ovarian mass, ectopic ureter, hematosalpinx nor hematometra. Furthermore, the presence of a large bulge which clearly delineates the landmark of the vaginal septum precludes the use of laparoscopy; otherwise, laparoscopy would be needed in cases of small bulge.

Non fertility sparing surgery

The gold standard management for OHIVRA syndrome is septum resection and vaginoplasty. Nevertheless, in some rare cases, a surgeon may consider hemi-hysterectomy or even hysterectomy in difficult cases of proximal vaginal septum and impossibility of performing vaginoplasty, or in cases in which diagnosis is delayed leading to pelvic infections, pelvic inflammatory disease, and pyometra with or without microperforations of the septum.

Strassman procedure

Strassman procedure, which was once used for the treatment of bicornuate uterus, is a procedure during which metroplasty is performed by laparoscopy or laparotomy. A fundal uterine incision is done, followed by dissection layer by layer until reaching the endometrium and then apposition of the two uterine horns followed by uterine suturing in layers

Nursing diagnosis

1. Impaired Comfort related to pelvic pain and discomfort:

Interventions:-Administer prescribed analgesics as needed

- Apply heating pads or cold packs as appropriate.
- Teach relaxation techniques and distraction methods.

2. Impaired Urinary Elimination related to surgical trauma or catheterization:

Interventions:-Monitor urinary output and characteristics.

- Encourage fluid intake unless contraindicated.
- Assist with bladder training if necessary postoperatively

3. Anxiety related to surgical procedures and hospitalization:

Interventions: -Provide emotional support and reassurance.

- Teach relaxation techniques such as deep breathing.
- Encourage expression of feelings and concerns

4. Deficient Knowledge related to condition, treatment, and self-care post-discharge:

Interventions: - Provide clear explanations of the syndrome, surgical procedures, and expected outcomes.

- Offer written materials and resources for further reading.
- Teach wound care, medication management, and signs of complications.

5. Risk for Infection related to surgical wounds or urinary tract:

Interventions: - Monitor vital signs and signs of infection (e.g., fever, increased pain).

- Maintain strict aseptic technique during wound care and catheter management.
- Educate on proper hygiene practices and signs of infection.

6. Risk for Impaired Skin Integrity related to immobilization or surgical incisions:

Interventions:- Turn and reposition the patient regularly.

- Inspect skin frequently, especially around surgical sites.
- Use pressure-relieving devices as appropriate

7. Risk for Constipation related to opioid use and decreased mobility:

Interventions: - Monitor bowel patterns and provide interventions to promote regularity (e.g., stool softeners, fiber).

- Encourage adequate fluid intake and physical activity.
- Educate on dietary measures to prevent constipation.

Conclusion

OVHIRA is a complex syndrome requiring accurate diagnosis and appropriate management. According to the available pieces of evidence collected in this systematic review, minimally invasive approach is a feasible option in most of the cases. Considering the acceptable fertility and obstetric outcomes after surgical management, fertility-sparing surgery should be the first-line option in reproductive-age women.

References

1. M.A. El-Gohary , Uterus didelphys with obstructed hemivagina and ipsilateral renal anomaly (OHVIRA syndrome): a case report. *Journal of Pediatric Surgery Case Reports*, 2 (2014), pp. 410-412
2. C.E. Purslow , A case of unilateral haematokolpos, haematometra and haematosalpinx . *BJOG An Int J Obstet Gynaecol*, 29 (1922), p. 643
3. A. David, S.N. Gudi, R. Shankar Herlyn—werner—wunderlich syndrome: premenarche *Journal of South Asian Federation of Obstetrics and Gynaecology*, 9 (2017), pp. 207-210
4. N.A. Smith, M.R. Laufer ,Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome: management and follow-up. *Fertil Steril*, 87 (2007), pp. 918-922