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HIGH OHVIRA SYNDROME WITH CERVICAL STENOSIS – ATYPICAL PRESENTATION OF A RARE ENTITY A CASE REPORT

Gynaecology

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ABSTRACT

Background-We present an unusual presentation of a rare case of OHVIRA syndrome with high vaginal septum, cervical stenosis and no hematocolpos. **Case report-** The patient is 13year old girl who presented with progressive worsening pelvic pain and pelvic mass but regular menstrual cycles. Radiological investigations detected high vaginal septum, cervical stenosis along with renal agenesis and endometrioma on the right side. Use of Foley's catheter and 3 months continuous oral contraceptive pills course was an addition to the standard treatment of septal resection in this case. **Summary and conclusion-** There is paucity of data on high OHVIRA syndrome. Our goal is to create awareness among clinicians to suspect cervical anomalies with OHVIRA syndrome in the absence of hematocolpos. This will help in successful management of the disease.

KEYWORDS

High OHVIRA, hematocolpos, MRI, Foley's catheter, Oral contraceptive pills

INTRODUCTION

Reproductive tract anomalies are present in 7% of young women.[1] OHVIRA (obstructed hemivagina with ipsilateral renal agenesis) syndrome is a constellation of uterine anomalies with obstructed hemivagina and renal agenesis or anomalies. Most common uterine anomaly is uterus didelphys in OHVIRA syndrome but cases of septate uterus have also been seen.The prevalence of this syndrome is 0.4-6.7 % among mullerian anomalies as per various researches.[2] It belongs to class III of mullerian anomalies according to American Fertility Society classification and U3bC2V3 according to ESHRE classification.[3]This case is amongst the very few variants of OHVIRA syndrome that present with cervical stenosis.

CASE REPORT-

We discuss a case of 13year old girl who presented with complaint of severe colicky pelvic pain on the right side for 1 year, more during menses. Patient attained menarche 6 months back and had regular menstrual cycles. The girl was sexually inactive with no history of hormonal treatment. Birth history was suggestive of a term vaginal delivery with normal milestones. Body mass index was14 kg/m2 making her underweight. Tanner staging was grade 4 for both breast and pubic hair with normal external genitalia. Patient had a tender, vague mass with ill defined margins and restricted mobility in right iliac region.

USG whole abdomen and pelvis was suggestive of absent right kidney with two uterine cavities and two cervices. Right horn was bulky with hematometra but no hematocolpos and right sided cervical stenosis. MRI pelvis was done which confirmed the above findings in addition to enlarged right ovary with 6 cm X 2.6 cm chocolate cyst. (figure 1 a,b,c). On the basis of abdominopelvic MRI differential diagnosis of OHVIRA syndrome was made.

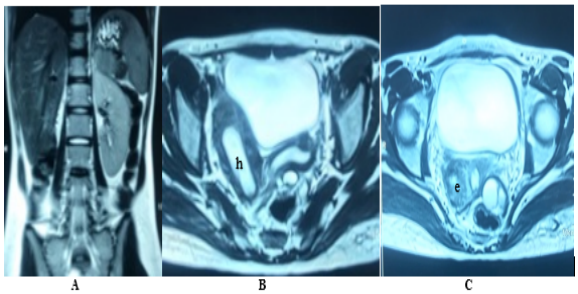

Figure 1 : MRI abdomen and pelvis with contrast

A- right renal agenesis(sagittal)
B- uterus didelphys with right hematometra (h) and right vaginal septum(transverse)
C- right sided endometrioma(e)(transverse)

Examination under anaesthesia was done and needle aspiration was tried per vaginally, but no fluid was aspirated confirming the absence of hematocolpos. Patient was re-evaluated during menses and vaginal septal resection along with hematometra drainage was planned. Considering the possibility of high vaginal septum, cervical stenosis/ cervical atresia/ rudimentary horn consent for laparoscopy, laparotomy and hemi- hysterectomy was also taken.

Patient was operated under general anaesthesia. On per vaginal examination 12 weeks oblong mass was felt on the right side.16 gauge needle was inserted and after 3 cm of negotiation with difficulty some old charry fluid (haematometra) was drained (fig 2) suggestive of obstruction at the level of both cervix and vagina. On expanding the space with artery forceps free flow of charry blood seen and around 30 ml drained. The level of obstruction was in the upper third of vagina above the level of cervix on the left side, making it a case of high OHVIRA. Hysteroscope was introduced and canal was visualized but clarity was restricted due to bloody field. As external os of cervix was not visualized, there was suspicion of excising a part of cervix at the site of obstruction with septal resection and so conservative approach was taken.

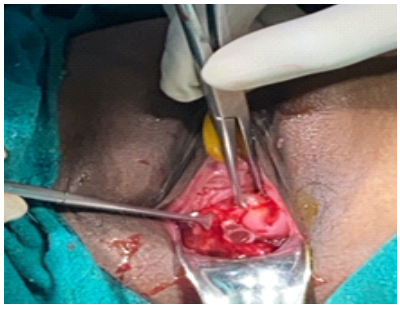

Figure 2- Hematometra drainage using 16 gauge needle c-cervix

Tissue obtained was sent for histopathological examination which was later reported to be vaginal septum. Vaginal reconstruction was done. Foley catheter no. 12 was inserted into right side cervix and uterus. (Fig-3 a and b)

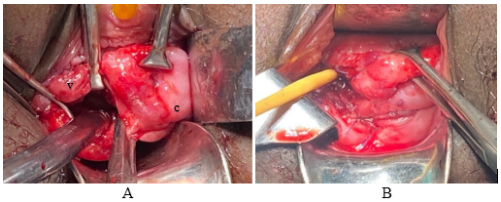

Fig 3 - A- vaginal septum (v) B- foley insertion post septal resection

Patient was discharged on post- operative day 7 under stable condition with foley's catheter in situ and continuous oral contraceptive pills were advised for 3 months. Patient came for follow up on postoperative day-14 when repeat USG was done which showed no collection and foley's catheter was removed. Patient has been followed up for 6 months and she is totally symptom free.

DISCUSSION AND CONCLUSION

Management of OHVIRA syndrome also called Herlyn Werner Wunderlich syndrome is complex and requires excellent knowledge of pelvic anatomy. The vaginal obstruction is mostly seen on the right side in 65% of the cases.[4] Renal anomalies are present in 43% of the cases as renal agenesis, renal dysplasia, duplication of kidneys and IVC duplication.[5] Girls with this anomaly mostly present around puberty with nonspecific pain, paravaginal mass and regular menstrual cycles making diagnosis difficult. Rarely they may present with urinary retention, UTI, chronic vaginal discharge, pelvic inflammatory disease and even sepsis. Usual presentation is with large hematocolpos, but in the present case due to the presence of high vaginal septum with cervical stenosis, there was no hematocolpos causing a diagnostic dilemma.

According to Acien hypothesis, origin of vagina is from mesonephric ducts and sinovaginal bulbs. Agenesis of distal part of mesonephric duct most likely results in upper vaginal agenesis (high septum) and distal cervical stenosis. Abnormal wolffian duct that fails to open into urogenital sinus, leads to absence of ureteric bud and right ureter and further right kidney development. [6] In the study by Wang et al, 3 cases of hemicervical atresia have been reported. This is an extremely rare variant of OHVIRA syndrome. However in our case cervix was well developed but stenosed. [7]

MRI has been proved to be gold standard modality and helps in diagnosing the level of obstruction, thickness of vaginal septum, renal anomaly, uterine anomaly and also the presence of endometriosis. It is useful in avoiding unnecessary laparoscopy and laparotomy in patients with OHVIRA syndrome. The treatment of choice is hematocolpos and hematometra drainage with vaginal septal resection as done in this case in one sitting only. Two stage treatment is advocated in cases where infection is suspected such that in first sitting drainage is done while in the next sitting septal resection or hysterectomy is done. Unilateral hysterectomy can also be offered once vaginal restenosis is seen or in cases of high OHVIRA.[8] As cervix was well developed in this patient cervicoplasty was needed and patient responded well to vaginal septal resection.

In our case due to high obstruction and no hematocolpos, there was early formation of endometriosis. In patients of high septum/ cervical atresia, early surgical management is all the more important to prevent endometriosis and preserve fertility. In this case we used foley's catheter no 12 for 2 weeks in the obstructed hemi uterus.It proved to have favourable outcome especially in the presence of amenorrhea induced by continuous use of OCPs as in this case. OCP's further helped in healing at the vaginal surgery site after septal resection. Reproductive outcome is good post surgery with 87% of women being able to conceive spontaneously.

Suspicion of high OHVIRA with cervical anomalies should be kept in mind in the absence of hematocolpos. Paediatric surgeons and gynaecologists should become vigilant and suspect for renal anomaly once patient presents with uterine anomaly and vice versa. Early detection relieves symptoms as well as prevents complications.

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