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RESEARCH ARTICLE

A RARE CASE REPORT OF OHVIRA SYNDROME IN AMENTALLY RETARDED GIRL

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Abstract

Obstructed Hemi vagina & Ipsilateral renal agenesis or Herlyn werner Wunderlitch syndrome is a rare Mullerian duct anomaly with uterus didelphys, Unilateral Obstructed Hemi vagina & Ipsilateral renal agenesis. Patients can usually presents soon after menarche and mayshow non - specific and variable symptoms with resultant delay in diagnosis. The most common presentation isdysmenorrhoea , abdominal mass in the lower abdomen secondary to hematocolpos and or haematometra. Here presenting a case of 14 years old mentally retarded female presented with pain abdomen & mass per abdomen and was diagnosed as **OHVIRA** syndrome based on Ultrasonography & MRI pelvis. She was treated with hemi vaginal septal resection and was under follow up.

Introduction:-

OHVIRA syndrome describes a congenital malformation consisting ofa uterusdidelphys with obstructed hemivagina and an associated Ipsilateral renal anomaly , most likely as a result of defective fusion of the paramesonephric ducts during embryonic development.

Prevalence varies between 0.1% to 3.8% . It is generally observed inpostmenarche adolescents with dysmenorrhoea, irregular menses, abdominal pain & pelvic mass. Diagnosis is often difficult due to theinfrequency of the syndrome & a high index of suspicion is required.

OHVIRA syndrome diagnosis requires a multimodal approach, integrating thepatient's history, clinical presentation, imaging studies including ultrasonography, computed tomography, MRI & direct visualization via hysteroscopy or laproscopy. A high index of suspicion in patients with renal anomalies & endometriosis is neededto detect the **OHVIRA** syndrome for timely diagnosis to avoid complications form the syndrome. Delay in diagnosis increases the risk of complications such as endometriosis & infertility.

Case Report:

A 14 years old mentally retarded girl presented with continuous painin lowerabdomen for the last 3 weeks. She had menarche at 13 years & a normal menstrual cycle of 5/28 days. Physical Examination revealed normal external genitalia. Clinical abdominal examination was tenderness in right iliac & suprapubic region. Hemogram & otherbiochemical parameters were within normal range. Ultrasound examination at outside shows:- 1) right kidney not visualised. 2) bicornuate uterus with gross hematometra & hematocolpos in left horn reaching up to level of umbilicus & right hematosalpinx. 3) mild hydroureteronephrosis of left kidney.

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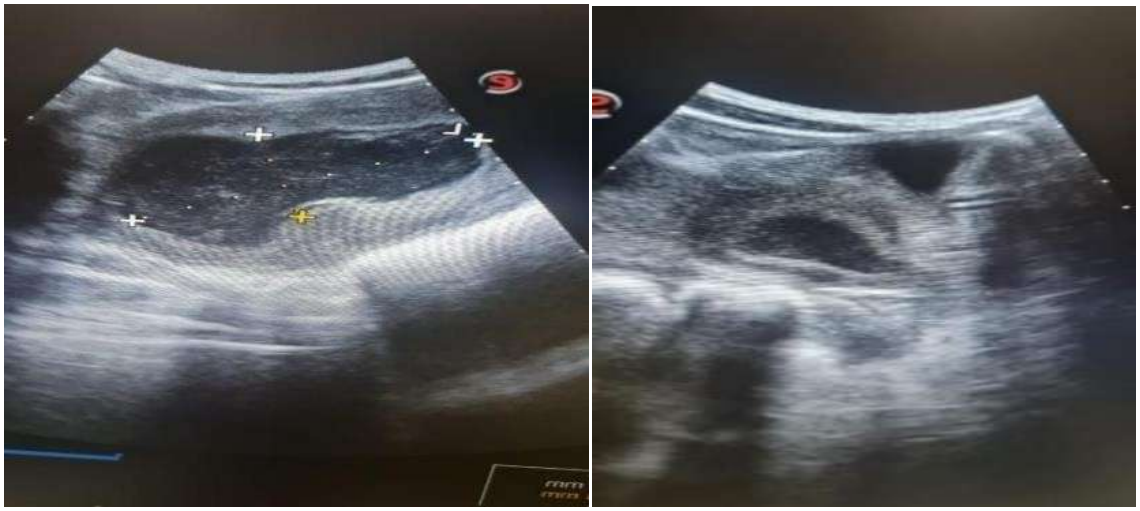
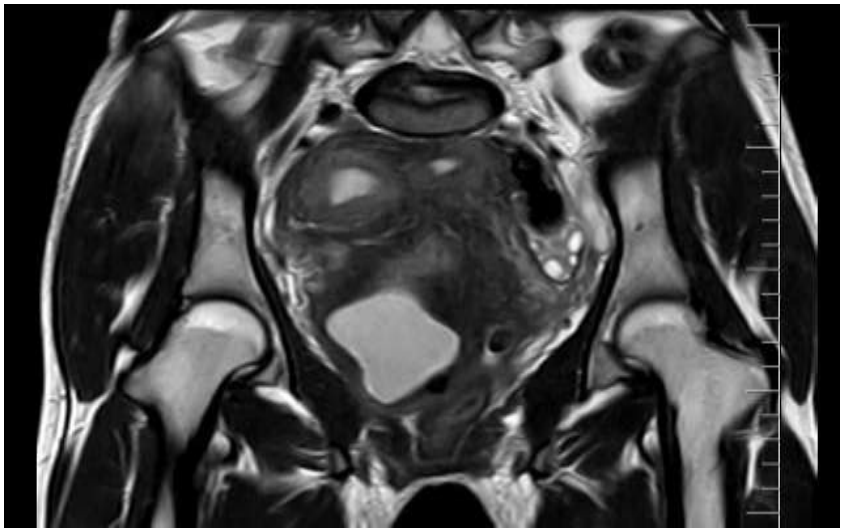
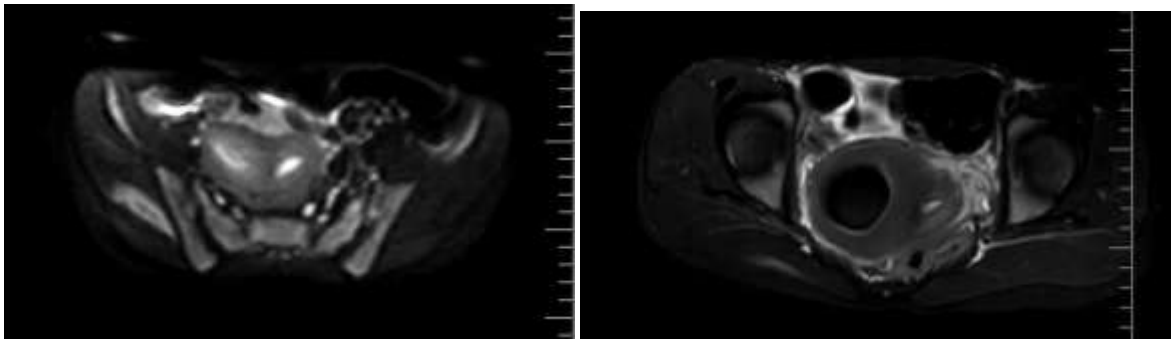
Ultrasound images of OHVIRA syndrome



Transabdominal ultrasound of the pelvis at our institute revealed 2uterine horns & uterus of size 15*9 cms & evidence of echogenic collection noted in uterine cavity which is extending into bilateral fallopian tubes (right > left) & evidence of 2.2*1.6cms collection noted at right side of uterus with thin ET ie.,rudimentary horn of uterus. Right kidney not visualised ie., ectopic or agenesis & moderate hydroureteronephrosis of left kidney:- Suggestive of biocornuate uterus with hematometra in left cornua with bilateral hemato salpinx & right ectopic/agenesis of kidney & advised MRI pelvis to rule out Mullerian anomalies. MRI done on next day revealed 2 vaginal cavities.

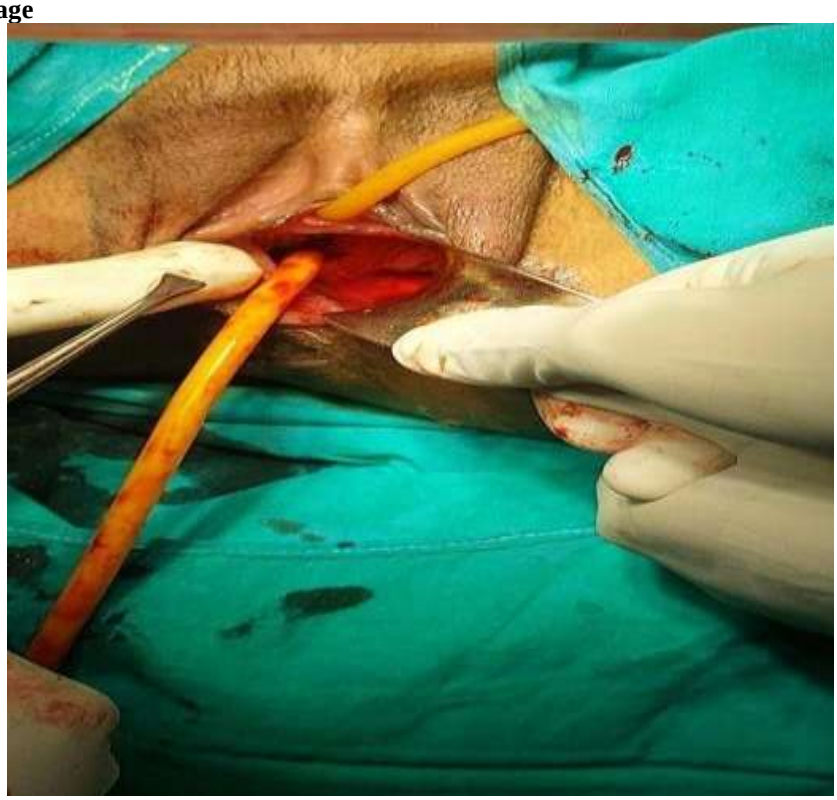




MRI images of OHVIRA Syndrome

The collapsed left hemi vagina was seen to communicate with leftcervix &corresponding uterine horn.

Intraoperative image



The right hemi vagina was markedly distended, & its lower end was convex & placed 1.5cm above the introitus. The upper end communicated with the right uterine horn through the right cervix. Signal intensity of the contentsof distended hemi vagina was consistent with blood products, suggestive of a right hemi – hematocolpos. Impression given was uterus didelphys with hematometra in right horn, right hematosalpinx & hemi – hematocolpos. A diagnosis of **OHVIRA** syndrome was made. Local examination under anesthesia revealed abulge in medial wall of the patent left hemi vagina caused by the distended right hemi vagina. Patient was treated surgically by incising the septum & vaginoplasty was done. After that foley's catheter was introduced & bulb was inflated with 30 cc saline. Leftcervix visualised &dilatation was done serially & surgical outcomewas satisfactory with expectation of near normal fertility & normalsexual function.

Discussion:-

Mullerian duct abnormalities cover a wide range of developmental anomalies, resulting from Non development or defective fusion or defects in regression of the septum during foetal development.

OHVIRA syndrome comes under type 3 mullerian duct anomaly associated with mesonephric duct anomaly. Mesonephric & paramesonephric ducts are the paired urogenital structures fromwhich internal genital organs & lower urinary tract are derived.

Mesonephric ducts are inductor element for paramesonephric ducts.If developmental arrest of one of the mesonephric duct occur, then the Mullerianduct on the side lacking the wolffian duct displaces itself laterally and cannot come into direct contact with urogenital sinus with the resultant formation of ablind sac, imperforate or obstructed vagina. It is again classified into completely obstructed hemi vagina & incompletely obstructed hemi vagina. Incomplete type again classified into 2 types ie., 1) Blind hemi vagina 2) Cervico vaginal atresia without communicating uterus. Here it presents with complaint of dysmenorrhea & pain abdomen. On evaluation, hematometra, hematosalpinx can be found, which if untreated progresses to secondary endometriosis, pelvic adhesions, pyosalpinx,pyocolpos. In incomplete type there are 2 types ie., 1) A small communication exists between two vaginas with resultant incomplete

obstruction. 2) Communication between two duplicated cervices with completely obstructed hemi vagina. Here it presents with complaints of irregular bleeding per vaginum & on evaluation pelvic inflammatory disease canbe found. Treatment requires surgical intervention in the form of excision of vaginal septum to relieve obstruction. Surgery also reduces the chance of pelvic endometriosis due to retrograde menstrual bleeding. Earlier hemi hysterectomy was done, Nomore preferred now as incidence of pregnancy in both horns is almost equal. We have to make everyeffort to preserve the obstructed uterus.

Conclusion:-

OHVIRA syndrome is a rare anomaly with potential short & long term complications. Diagnosis likely to be missed because of normalmenstruation &Non specific abdominal pain. So, early diagnosis & treatment can be prevents the long term complications like pelvic endometriosis & infertility.