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Original Research Paper

Radio-Diagnosis

OHVIRA (OBSTRUCTED HEMIVAGINA WITH IPSILATERAL RENAL AGENESIS) WITH VERTEBRAL FUSION ANOMALY, SCOLIOSIS AND ABSENCE OF COCCYX: AN UNCOMMON MEDLEY!

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ABSTRACT OHVIRA(Obstructed hemi-vagina with ipsilateral renal agenesis),also known as Herlyn-Werner-Wunderlich syndrome(HWW) is a rare case of a complex uterine anomaly.Associationwith congenital vertebral fusion anomalies, curvature abnormality of spine and congenital absence of coccyx together in a single patient has been sparsely documented in literature. We present a case of 15 year old female with varied spectrum of radiographic, ultrasonographic and MRI findings. MRI remains the gold standard investigation for diagnosing this rare congenital uro-genital anomaly.

KEYWORDS :

INTRODUCTION-

HWW syndrome is a complex congenital entity comprising of Müllerian duct anomalies (MDA) associated with mesonephric duct anomalies. MDA are the by products of non-development (agenesia or hypoplasia), defective vertical or lateral fusion, or resorption failure of the Müllerian (paramesonephric)ducts[1].With an estimated overall prevalence of 2% to 3% among all women, and an incidence of 1 per 200 to 600 among fertile women,11% of uterine anomalies are accounted for by Didelphys uterus. 30% of cases show renal tract anomalies[2].

Preliminary stages of this syndrome is clinically masked on account of normal menstrual cycles with sheer symptoms of cyclic dysmenorrhea, treated with anti-inflammatory drugs and oral-contraceptives.[3].

Case Report-

A 15 year old girl presented to Hospital out patientdepartment with complaints of intermittent lower abdominal pain and discomfort from last 5 months with increased intensity of pain since last 4-5 days. She attained menarche at 14 years of age with regular30 day cycles, spanning across 4 dayswith normal menstrual bleeding and no significant dysmenorrhea. Clinical examination revealed a palpable abdominal lump with tenderness in pelvis extending upto right iliac fossa. External genitalia were normal on examination. Hematological parameters were within normal limits and rest of systemic examination showed no other significant abnormalities.

Patient underwent CT scan of abdomen outside our institute, which was reported as complex right adenxal mass lesion with non visualization of right ovary .Transabdominal and translabial USGrevealed two uterine hornswith two vaginas, wherein the right uterine horn -right hemivaginaappeared grossly dilated with anechoic fluid and internal dependent debris, which was causing compression and displacement of left hemi-vagina. Right fallopian tube was alsotortuous and grossly dilated with anechoic fluid with dependent debris.Bilateral ovaries were normal.Evaluation of upper abdomen showedabsence of right kidney in right renal fossa and normal sized left kidney .Rest of solid organs in abdomen were unremarkable.

MRI examination (3 Tesla GE Discovery 750W) .Images were acquired on multiple planes with T1-weighted and T2-weighted Spin Echo sequences, 3D LAVA and GRE images. MRI showeddidelphys uterus with clear demonstration of two vaginal cavities. The interconnected right endometrial cavity and vagina canal were grossly dilated and distended with blood products extending into the right fallopian tube and thereby causing grossly dilated and tortuous right fallopian tube. Bilateral ovaries were normal with anteriorly displaced right ovary which appeared to be compressed by right hematocolpos. The hemotocolpos was seen indenting the posterior wall of the urinary bladder.Right kidney was absent while the left kidney wasnormal in size and located in left renal fossa. Bony abnormalities included scoliosis of dorso-lumbar spine with convexity towards left side, there was congenital absence of coccyx noted with fusion of posterior element of D8 and D9 vertebra which were also documented on frontal dorso-lumbar spine radiograph.

DISCUSSION-

The pathogenesis and etiology of OHVIRA syndrome remains unclear with hypothesis suggesting it to be the result of anamalous development of the Müllerian and Wolffian ducts. Embryologically, two pairs of Müllerian ducts fuse craniocaudally to form the uterus, cervix, and upper two-thirds of the vagina. The fused Müllerian ducts perforate into the urogenital sinus, giving rise to the lower third of the vagina. Wolffian ducts not only develop into the ureter and the kidneys, but also induce adequate Müllerian duct fusion. Maldevelopment of the Wolffian duct therefore causes failure of ureter and kidney differentiation, in addition to the lateral displacement of the ipsilateral Müllerian duct. The displaced Müllerian duct hence cannot fuse with the contralateral duct, causing uterine didelphys, and the resultant failure to contact the urogenital sinus forms a blind sac, leading to an obstructed hemivagina[4-7].

The OHVIRA syndrome presumably occursshortly after menarche, with patients presenting with pelvic pain or dysmenorrhea, with or without palpable mass due to hematocolpos or hematometra [4]. In incomplete obstruction, clinical presentation can be delayed as there remains one patent hemivagina, allowing for menstrual blood to exit, while the other side is obstructed [4, 5]. As the incidence of

complications including endometriosis, menstrual disorders, infertility, and obstetric complications increases with time [4-6], accurate and early diagnosis is essential.

In conclusion, MRI is the most efficient tool in establishing the diagnosis of Müllerian duct anomalies and associated conditions, when compared to other imaging modalities. MRI must be performed in young females suspected to have OHVIRA syndrome in order to make an accurate diagnosis and to optimize the early appropriate treatment.

Treatment invariably requires surgical intervention in the form of excision of vaginal septum to relieve obstruction. Surgical intervention aids in relief of pain due to obstruction andreduces chances of pelvic endometriosis due to retrograde menstrual seeding. Patients are able to have normal sexual life with some cases of successful conception and full term pregnancies.

CONCLUSION-

OHVIRA syndrome is an uncommon congenital anomaly with clinical significance and simple surgical management. Imaging, particularly USG is done as a preliminary investigation which is followed by MRI to make a definitive diagnosis. Appropriate surgery is a single stage procedure to either excise or completely divide the obstructing septum. The complex mullerian anomalies have been difficult to diagnose correctly without the aid of radiological imaging. A correct early diagnosis with the essential goal to relieve the symptoms and prevent complications, caused by retrograde menstruation which may result in endometriosis with emphasis on aiding in decent quality of life with preserving sexual and conception abilities makes this an important syndrome to be borne in mind of the clinician and radiologist.

Case Images:

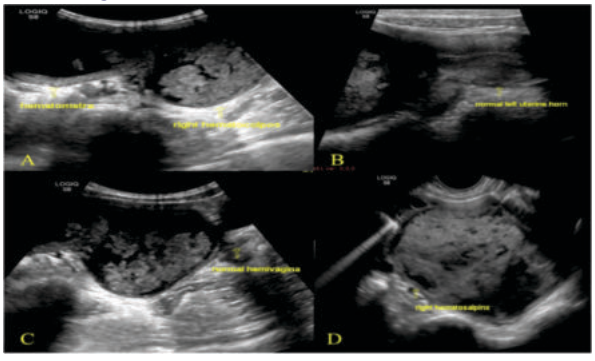

Fig 1: Sagittal Longitudinal USG image (A)shows elongated and dilated right uterine horn with dilated vagina with anechoic fluid with low level echoes. (B)Longitudinal USG image shows normal left uterine horn with normal endometrial cavity. Longitudinal USG image (C)shows normal left hemivagina with adjacent hemotocolpos and hematometra. Transverse USG image (D)shows dilated right fallopian tube with anechoic contents and low level echoes- s/o Hematosalpinx

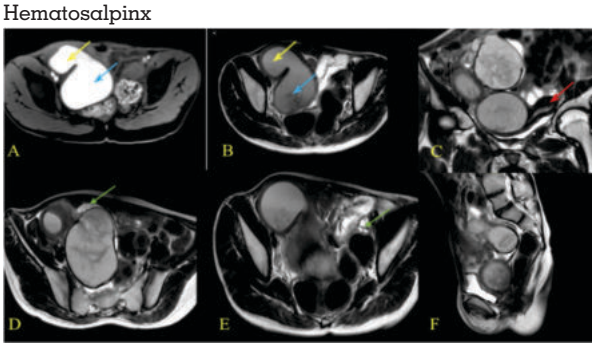

Fig 2. Axial T1W FS image(A) shows hyperintense contents in right endometrial cavity and vaginal canal, corresponding T2W image (B) shows hyperintense signal of contents with hypointense debris independent part of contents-s/o hematometra (yellow arrow)and hematocolpos (blue arrow). Coronal T2W image (C)shows normal left uterine horn with normal left hemivagina (red arrow). Axial T2W images(D) shows normal right and left ovary(E)(green arrows). Sagittal T2W image (F) shows agenesis of coccygeal vertebral segments

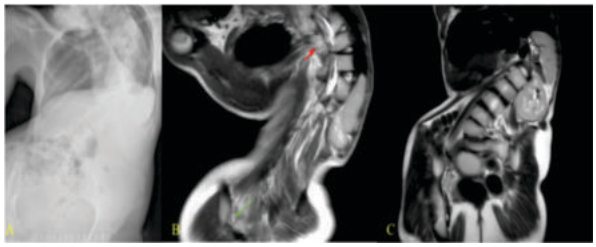

Fig.3 Frontal projection of Dorso-lumbar spine (A) shows severe scoliosis of dorso-lumbar spine with convexity towards left side. Coronal T2W images (B) shows fusion of posterior elements of D8-D9 vertebra (red arrow) with with scoliosis of dorso-lumbar spine and absence of right kidney (C)

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