A RARE CASE: OHVIRA SYNDROME (OBSTRUCTED HEMIVAGINA AND IPSILATERAL RENAL ANOMALY) WITH UNICORNUATE UTERUS

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PRESENTATION OF CASE

A 50-year-old unmarried, nulligravida female patient presented with chronic dull aching abdominal pain predominantly in suprapubic region since past 4 months and loose stools since 1 month with history of absence of menarche. There was no history of fever, white discharge per vagina or any significant medical history, and she was a non-diabetic and normotensive. The patient had no history of surgeries in the past. On examination, there was blind ending vagina. She was evaluated initially by radiography, which showed a well-defined, radio-opaque density in the pelvis approximately 4.8 x 4.5 x 2.7 cm in size (TR x AP x CC). Further the patient was evaluated with the computed tomography which showed a large, well defined, oval shaped, lamellated calcification (HU 850-900) measuring 5.1 x 4.8 x 3.1 cm (TR x AP x CC) in the pelvis, posterior to the urinary bladder and inferior to the uterus. The computed tomography of the patient also showed absence of the right kidney. MRI study of the patient showed unicornuate uterus with normal left ovary and the calcification was noted within the upper two – third of the dilated vagina. The right ovary was not visualized on imaging.

CLINICAL DIAGNOSIS

Primary Amenorrhoea

DIFFERENTIAL DIAGNOSIS

- Calcified haematocolpos with unicornuate uterus, absent right ovary and right kidney.
- Calcified foreign body in the vagina with unicornuate uterus, absent right ovary and right kidney.

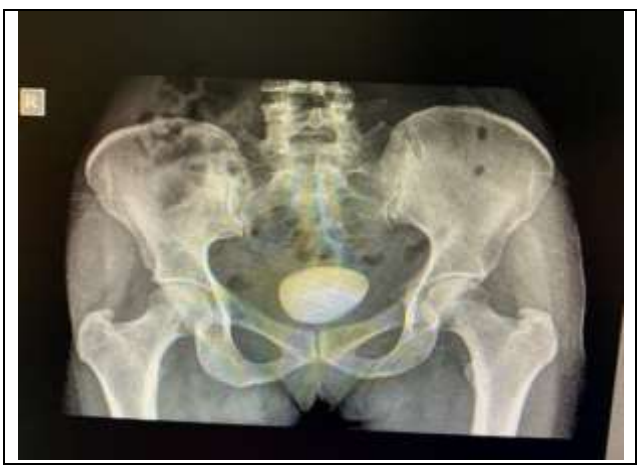

Figure 1. Plain Radiograph Pelvis AP View. Radiograph Showing Oval Shaped, Homogenous, Radiopaque Density Lesion in The Pelvis

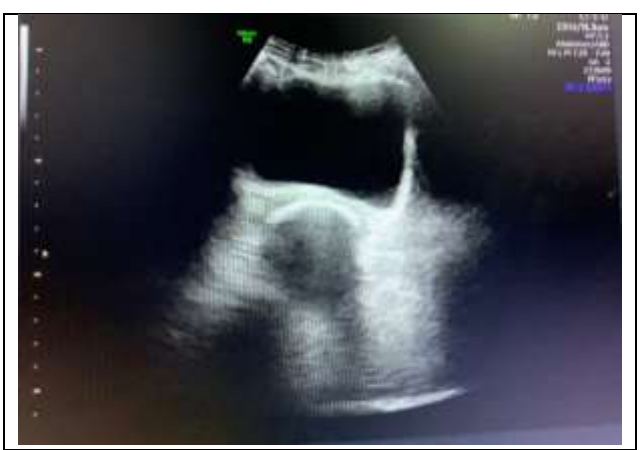

Figure 2. Ultrasound Image showing Calcification in The Pelvis Posterior to The Bladder with Posterior Acoustic Shadowing

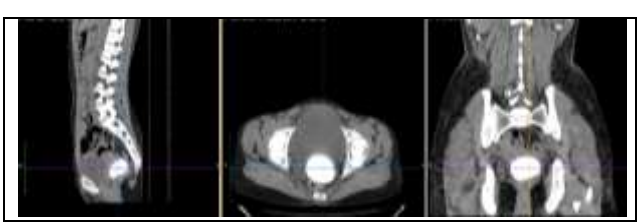

Figure 3. Plain Computed Tomography Image Showing Absence of the Right Kidney with Compensatory Hypertrophy of the Left Kidney and a Well Defined, Oval Shaped, Lamellated Calcification Posterior to the Bladder and Inferior to Uterus

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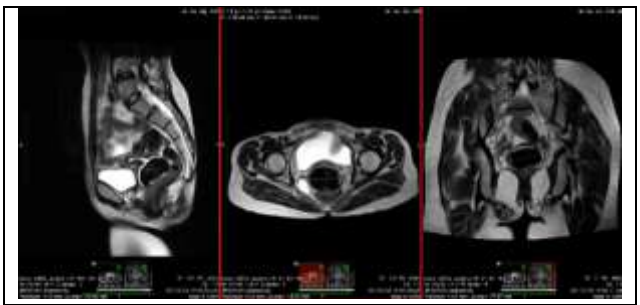

Figure 4. MRI Image Showing a Well-defined, Oval Shaped Lesion Which Appears Hypointense on T2 Sequence in The Dilated Vagina

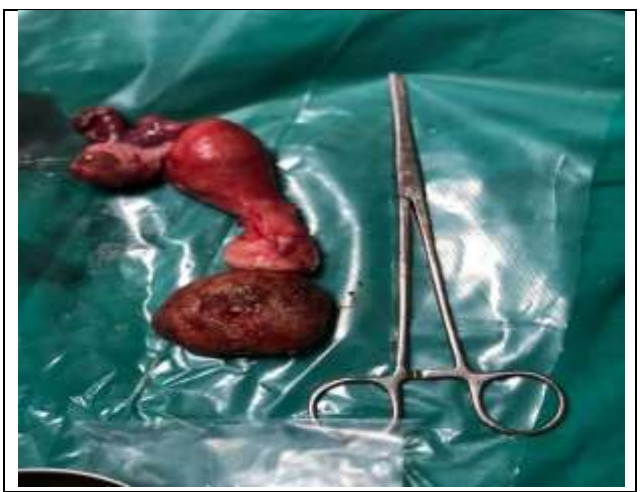

Figure 5. Gross Appearance: Unicornuate Uterus with Normal Left Ovary, Vaginolith and Absent Rudimentary Horn

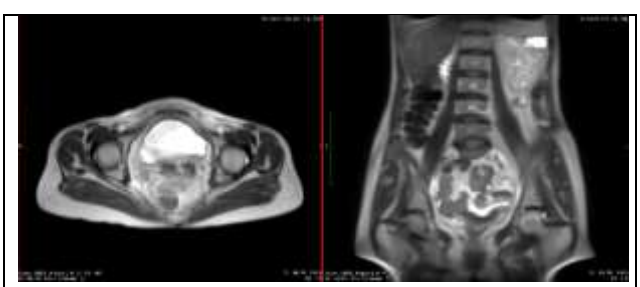

Figure 6. Post-Operative MRI Image of The Patient. Post Hysterectomy Status

DISCUSSION

Herlyn-Werner-Wunderlich syndrome (HWWS) is mostly misdiagnosed and high index of suspicion is required in patients with Mullerian and mesonephric duct anomalies. These patients generally present at puberty with pelvic pain, but rarely can present in neonates or in the adulthood in the form of primary infertility, pyometra, ischiorectal swelling, and urinary obstruction.[1,2,3,4]

Herlyn-Werner-Wunderlich syndrome (HWWS) is a combination of Type III Mullerian anomaly with mesonephric duct anomaly with vaginal septum. The classic renal manifestation of OHVIRA syndrome is ipsilateral renal agenesis, but cases of dysplastic kidneys, duplicated kidneys,[5] recto vesical bands,[6] or crossed fused ectopia[7] have also been reported. Uterus didelphys with obstructed hemivagina is due to lateral non-fusion of the Mullerian ducts with asymmetric obstruction, and it is mostly associated with renal agenesis on the same side to the side of obstruction.[5] Another important point is that the renal agenesis in patients with HWWS is located on the same side (ipsilateral) to the dilated uterine cavity. In case of unicornuate uterus, the renal anomalies are on the same side (ipsilateral) to the rudimentary or absent uterine horn. Magnetic resonance imaging (MRI) is the modality of choice for the diagnosis of HWWS and also for other such anomalies because of better anatomic delineation of pelvic structures and higher sensitivity of the MRI for blood products. Since the vaginal septum can be treated by septal excision, and the delay in the diagnosis may worsen the associated endometriosis,[8] hence early diagnosis and treatment is beneficial.

As per review of literature, our case is the second reported case of OHVIRA syndrome with unicornuate uterus and single ovary.

DISCUSSION OF MANAGEMENT

Treatment of OHVIRA syndrome invariably requires surgical intervention in the form of vaginal reconstruction in young age patients and hysterectomy in elderly patients. In addition to relief of pain due to obstruction, surgery also reduces chances of pelvic endometriosis due to retrograde menstrual seeding. Patients are able to have normal sexual life. Some are even able to conceive and carry pregnancy to term.

FINAL DIAGNOSIS

OHVIRA Syndrome

OHVIRA syndrome is an uncommon congenital anomaly and has clinical significance. It can be treated with simple surgical management. Ultrasound and MRI findings can collectively delineate uterine morphology, absence of the ipsilateral kidney, and the nature of the contents in the uterus and obstructed hemivagina. Hence in a case of obstructed hemivagina in a duplicated genital system and ipsilateral renal agenesis this rare syndrome should be kept in mind to help arrive at an exact and early diagnosis, critical in preventing chronic complications.

Abbreviations:

TR – Transverse.
AP – Antero-posterior.
CC – Cranio-caudal.
cm – Centimeter.
CT – Computed tomography.
MRI – Magnetic Resonance Imaging.

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