Herlyn-Werner - Wunderlich syndrome: a rare case report.

INTRODUCTION: Herlyn-werner-wunderlich syndrome is a rare congenital malformation of the mullerian ducts. It is stipulated to be a developmental abnormality of the wolffian ducts as well as of that of the mullerian ducts. Also known today as uterine didelphys associated with obstructed hemivagina and ipsilateral renal anomaly (ohvira), a broader term, to include other types of renal abnormalities. Significant occurrence is noted commonly on the right side and there is no clear explanation for this fact. The disease is rare and prevalence is undetermined. In the general population, unilateral renal agenesis is estimated to be from 1 in every 600-1200 individuals. In women, the prevalence of genital abnormalities associated with kidney anomalies is estimated between 25-89%. The abnormalities of the urinary tract most frequently associated with developmental abnormalities of the mullerian ducts are renal agenesis, double collecting system, double kidney and horseshoe kidney.

CASE PRESENTATION: Patient is a 14 year old girl presented with history of irregular menses and vague lower abdominal pain. Menarche was at 12yrs. USG performed revealed cystic mass in the pelvis in the midline. Patient referred for MRI Abdomen.
FINDINGS:

- There are widely divergent uterine horns with no communication between them. (Fig 1)
- There is distension of the cervix and proximal two-third part of vaginal canal on right side; with fluid, showing signal intensity of blood degradation products. (Fig 2& 4).
- Left cervical canal and proximal two-third vaginal canal appear to be compressed and stretched over the distended right cervix/vagina. (Fig 3).
- Distal one-third of the vaginal canal appears normal.
- Right kidney is absent. (Fig 5).
- Others: Segmentation anomaly is noted in L3 and L4 vertebrae. (Fig 6).

FINAL DIAGNOSIS: Uterus didelphys with unilateral (right) obstructed hemivagina with hematocolpos with ipsilateral renal agenesis.

DISCUSSION: The Wolffian ducts, in addition to giving rise to the kidneys, are factors that cause adequate fusion of the Mullerian ducts. Therefore, abnormality in the development of the caudal portion of the Wolffian ducts can be the cause of unilateral renal agenesis associated with imperforated hemivagina. The Mullerian duct is laterally displaced on the side in which the Wolffian duct is absent and cannot fuse with the contralateral duct, resulting in a bicornuate uterus with no central urogenital sinus contact. The contralateral Mullerian duct gives rise to the vagina, while the displaced component forms a blind sac, the obstructed or imperforated hemivagina. The vaginal introitus is not involved because it arises from the urogenital sinus.

CONCLUSION: In clinical practice it is important to detect this type of abnormality, despite its rareness, because it is the cause of precocious symptomatology and facilitates early pelvic endometriosis and collection infections (pyocolpos, pyometra, or pyosalpinx).

Regards,

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N.B: This case is authentic and from the archives of Radiance Diagnostics. For any queries/suggestions / feedback write to us at radiance@radiancediagnostics.in. Case of the month can also be accessed anytime online at VIEW BOX at www.radiancediagnostics.in.