A RARE CASE OF HERLYN-WERNER-WUNDERLICH SYNDROME/ OHVIRA SYNDROME

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INTRODUCTION

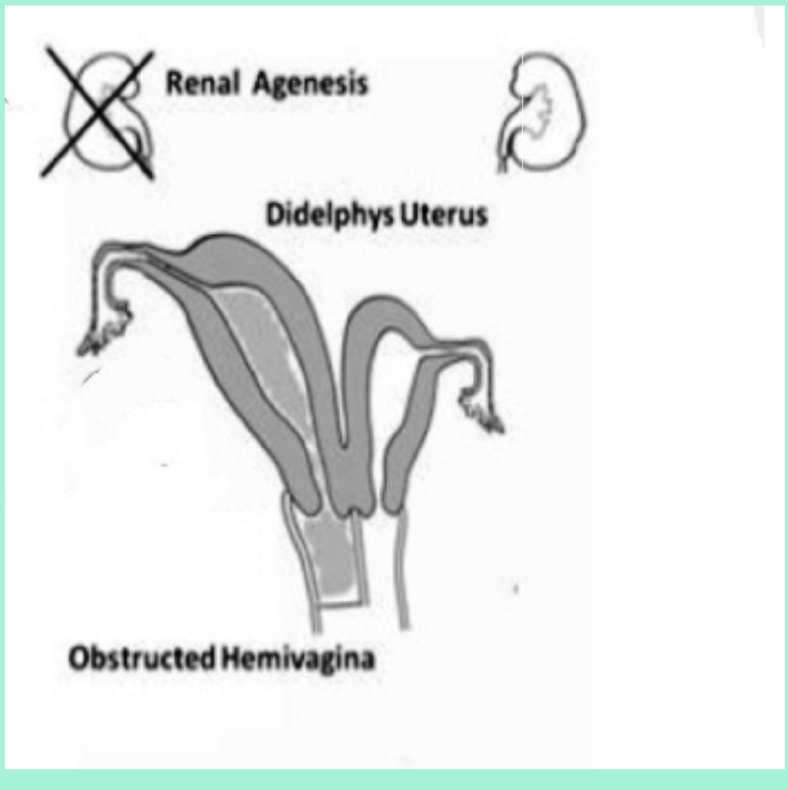
Herlyn-Werner -Wunderlich syndrome, known as OHVIRA syndrome(Obstructed Hemivagina and Ipsilateral Renal Anomaly) a rare complex of structural abnormalities of the female genital tract,characterized by triad of Mesonephric duct induced mullerian anomalies. Its incidence has been reported between 0.1% and 3.8%.It usually presents at puberty with pelvic pain. MRI is the modality of choice for the diagnosis of HWW Syndrome and other such anomalies because of better anatomic delineation of pelvic structure and higher sensitivity of blood collections.

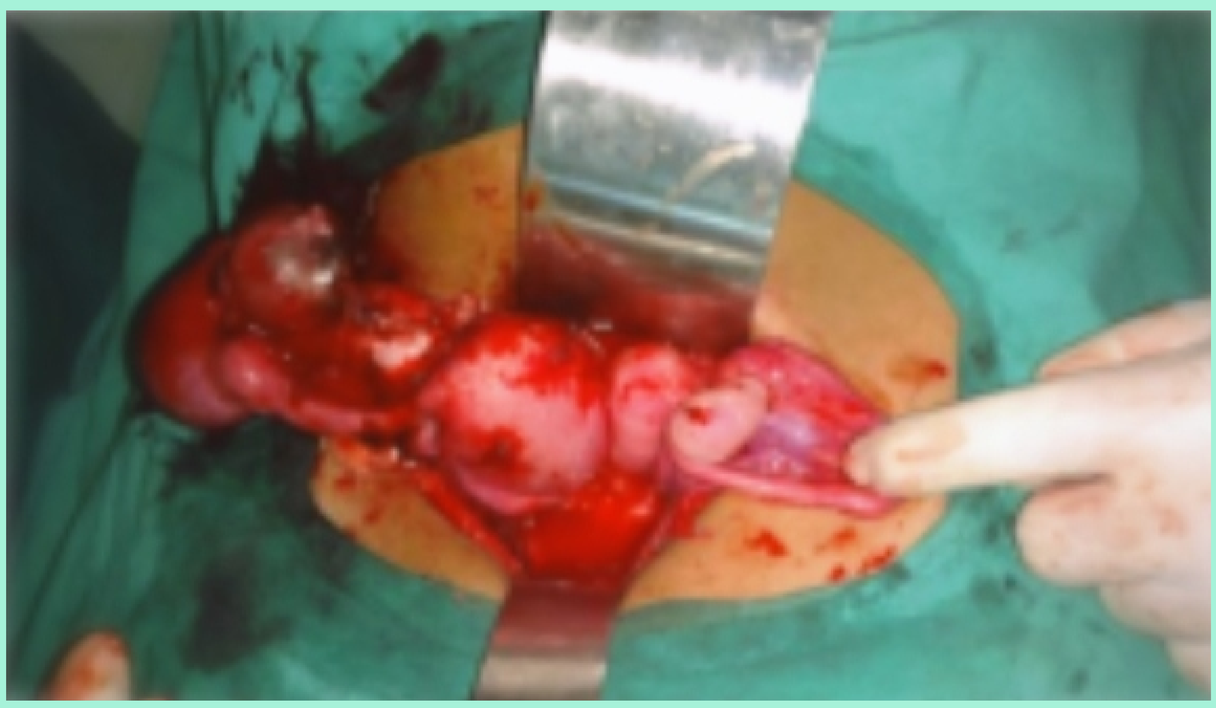
CASE PRESENTATION

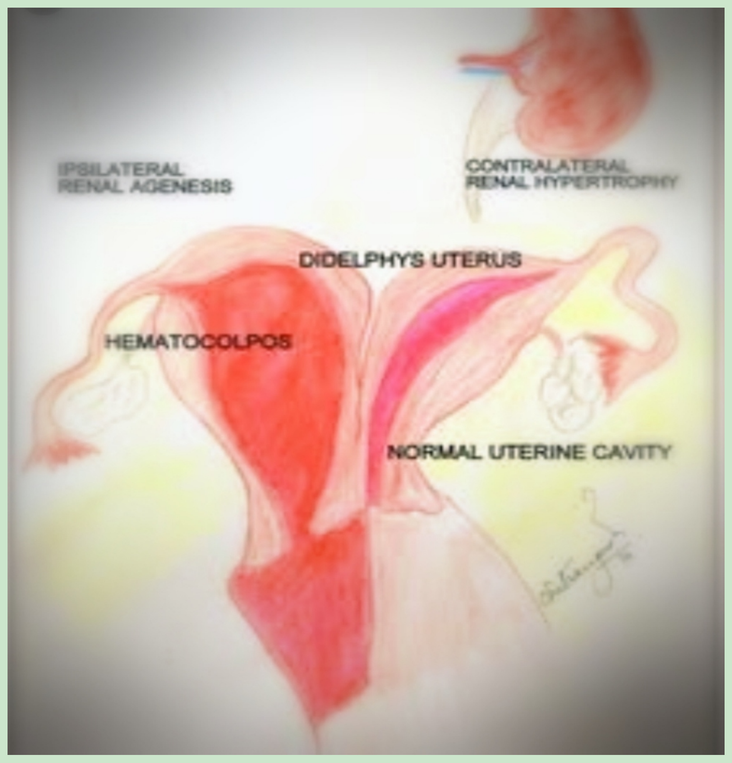
A 15 years girl, presented to our department for right lower quadrant abdominal pain without fever, diarrhoea or urinary symptoms. Menarche occurred 5 months back, cycles were regular and last for 3 to 4 days which was associated with severe dysmenorrhoea. Her general physical examination and vitals were within normal limits. USG findings revealed empty right renal fossa.USG complemented by CECT confirmed uterus didelphus, two uterine bodies, two separate cervices with right sided obstructed hemivagina and right renal agenesis.

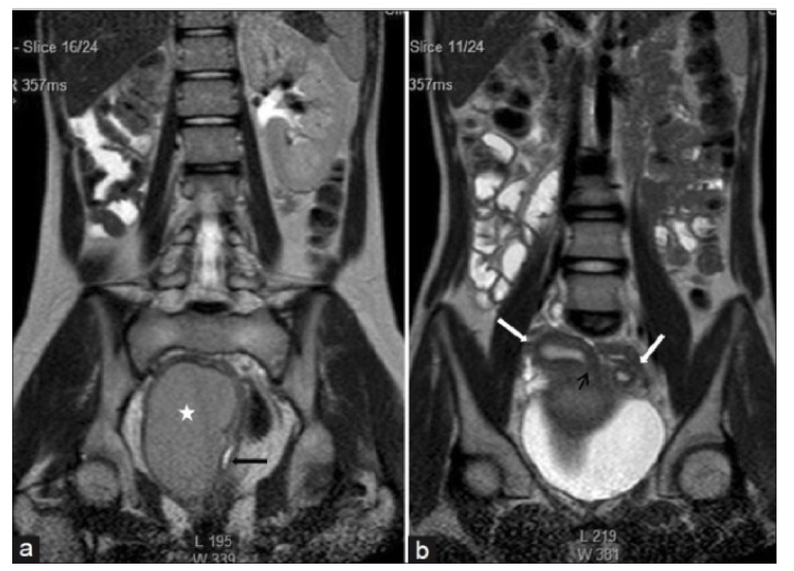
Patient underwent surgery with resection of right uterine horn with hematometra which was not communicating with vagina.

Intraoperative findings- Two horns of uterus seen. Left side was small with normal tubes and ovaries. Right side was large due to collection and with normal tubes and ovaries.









DISCUSSION

Mullerian (paramesonephric) duct anomalies are congenital anomalies of the female genital tract which results from non-development or non-fusion of the mullerian ducts or failed resorption of the uterine septum during sixth to ninth weeks of fetal life. Uterus ,cervix and upper 1/3rd of vagina develops from urogenital sinus. The urinary and genital systems arises from a common ridge of mesoderm arising along with dorsal body wall and rely on normal development of the mesonephric system. Renal agenesis is the most common anomaly although horseshoe or pelvic kidney , duplication of collecting system and ectopic ureters have all been described. HWW syndrome typically presents after menarche with non specific symptoms. The variation in diagnosis makes its diagnosis difficult and hence awareness regarding it needed. CT and ultrasound are most widely used diagnostic tools. However, MRI is considered to be more sensitive for imaging soft-tissue anatomy and delineating subtle findings seen in congenital anomalies.

CONCLUSION

The infrequency of HWW syndrome complicates its diagnosis and hence clinicians should consider mullerian duct anomalies among differential diagnosis in young female patients presenting with abdominal symptoms and menstrual complaints.