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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.
Method : 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.
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.

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