Obstructive hemivagina and ipsilateral renal agenesis

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ABSTRACT
Introduction: OHVIRA syndrome is a rare Mullerian anomaly with triad of uterine didelphys, obstructive hemivagina and absent ipsilateral kidney presenting with different symptoms and ages. We report an unusual presentation of the syndrome and discuss variation of its clinical symptoms and management. Case Description: A 14-year-old girl, presented with difficulty in emptying bladder intermittently for over a year along with urinary hesitancy, unassociated with menstruation. There was no other significant history, and general physical and abdominal examinations were unremarkable. Vulva appeared normal. Digital examination revealed a soft bulge on right side in vagina. CT abdomen and pelvis showed bicornuate uterus with hematocolpos, vaginal septum and absent right kidney. MRI pelvis and abdomen showed two separate uterine horns representing uterine didelphys, right sided hematometrocolpos and absent right kidney confirming OHVIRA. Resection of vaginal septum with cystoscopy and vaginoscopy was performed. A twenty-four-gauge Foley's catheter tamponade was inserted into the vagina and vagina was packed with gel gauze. Recovery was uneventful. 6 months later, she came to clinic with urinary hesitancy but regular cycles. Ultrasound was normal with nil post residual volume hence patient was reassured. One year follow-up in clinic was unremarkable and she had regular cycles.

Discussion: OHVIRA has a variety of clinical presentations thus can be missed easily. Recurrent urinary symptoms in a young adolescent female should elicit a high suspicion of OHVIRA. MRI is the gold standard investigation. Surgical correction at its earliest should be performed to relieve the symptoms and prevent long term complications.