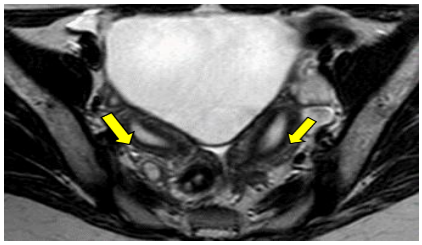
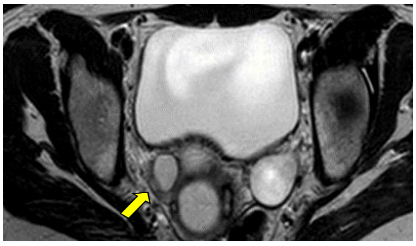
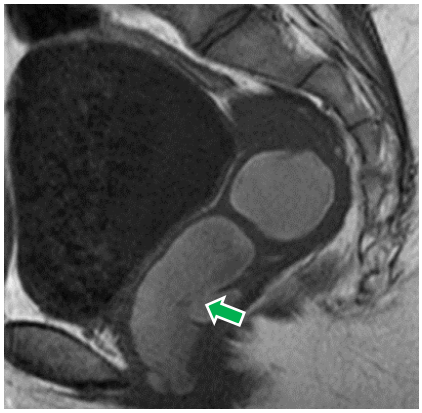
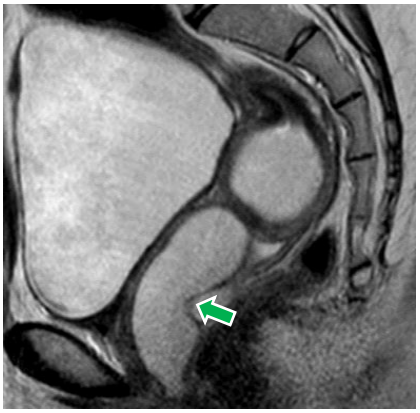
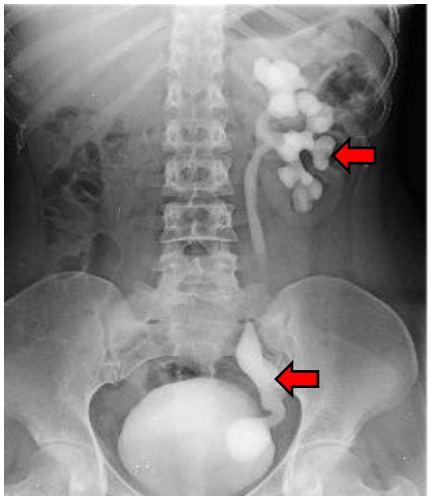
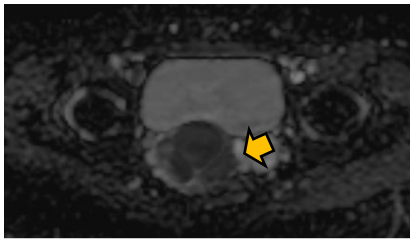
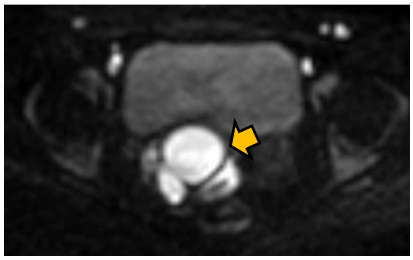
FROM THE CONSOLE ...

CLINICAL BACKGROUND:

A 24-Year-old female presented with complaints of cyclical abdominal pain during menstruation for the last 4 months which aggravated over the past 1 week. She had a normal menstrual cycle 3/28.Physical examination revealed normal external genitalia and clinical examination was unremarkable.

IMAGING FINDINGS:

MRI T2 images at the level of pelvis shows didelphic configuration of uterus (yellow arrow). MRI T1 and T2 sagittal images at the level of pelvis showed distended right hemi cervix and vagina with T1 and T2 hyper-intense content suggestive of hemato-metrocolpos (green arrow). ADC and DWI show (orange) areas of diffusion restriction within the right hemi cervix, suggestive of secondary abscess. Agenesis of right kidney and compensatory hypertrophy of left kidney with mega-ureter, megapolycalycosis and ureterocele which was confirmed with IVP (red arrow).

HERLYN-WERNER-WUNDERLICH SYNDROME

DISCUSSION:

The incidence of mullerian anomaly is estimated about 0.1 to 3.5% among which the incidence of uterine didelphys includes 11%. Renal anomalies are associated with uterine didelphys in ~ 80% of cases. Herlyn-Werner-Wunderlich (HWW) also known as OHVIRA syndrome is a rare complex congenital developmental anomaly is characterized by triad of Uterus didelphys, Unilateral low vaginal obstruction and Ipsilateral renal agenesis [class 3 Müllerian duct anomaly] [1,2].

An early failure of the metanephric diverticulum to develop (around 5 weeks) from the mesonephric duct results in agenesis of the ureteric bud, which leads to agenesis of ipsilateral ureter and kidney. Due to failed positioning of paired paramesonephric duct, the two hemi uterus and hemi cervices fail to unite, resulting in uterus didelphys. Developmental arrest of the distal ipsilateral mesonephric duct results in failure of distal hemivagina to develop, thereby resulting in obstructed hemivagina. Thus in OHVIRA syndrome, all 3 components are secondary to mesonephric duct-induced mullerian anomalies. Cyclical dysmenorrhoea is the most common complaint (1-4).

This syndrome has been reported to be associated with high bifurcation of aorta, IVC duplication, ovarian malposition, and intestinal malrotation. Imaging, particularly an MRI plays a major role in diagnosis, which is often missed clinically (2). Timely surgical intervention and psychological counselling are of utmost importance. Careful consideration must be given to patients with renal malformations to examine the reproductive system given their common embryology (2-4).

An early correct diagnosis is the goal to relieve the symptoms and prevent complications, caused by retrograde menstruation like endometriosis and, also, preserve sexual and conception abilities.

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