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HERLYN-WERNER-WUNDERLICH SYNDROME --AN INCIDENTALLY DETECTED RARE CASE OF PAIN IN ABDOMEN IN 20 YRS MARRIED FEMALE

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ABSTRACT Herlyn- Werner- Wunderlich syndrome (HWWS) is a very rare congenital developmental anomaly of Mullerian ducts and associated with unilateral renal agenesis (1).

It is a very rare type of syndrome with few cases has been reported so far.

In our case report, a 20 years muslim nulliparous lady with history of 2 years married life came to opd, with clinical presentation as pain in left flank of abdomen off and on, of mild to moderate in nature ,usually subsides after taking oral and injectable medicines.

The diagnosis of HWWS was suspected on ultrasound examination and confirmed by gynec examination and c.e.c.t abdomen scan with renal angiography.

The detailed examination and confirmation of syndrome is esential in order to avoid any complication and proper treatment in time.

KEYWORDS : Bicornuate uterus, vaginal septum, renal agenesis, Herlyn –Werner-Wunderlich syndrome, hemivertebrae, malrotated kidney

HISTORY / BACKGROUND

As per medical literature, the incidence of Mullerian duct anomalies is found to be ranging from 0.5 to 5 % in general population. the exact etiology of HWWS is still unknown, but it may be cause by the abnormal development of Mullerian and Wolffian ducts. it's estimated occurrence is 0.1% to 3.8% (2).

Crosby and hilli in 1962 firstly introduced the theory of the developments of Mullerian ducts (3).

EMBRYOLOGY

The Mullerian ducts (paramesonephric) develop from the coelomic epithelium and develop caudally along the Wolffian ducts (mesonephric) towards the urogenital sinus forming the two uterovaginal canal.

At the 11 weeks of gestation, the Mullerian ducts fused laterally to form a solitary canal which later on develops as uterus and proximal two-third of the vagina (4).

In the mean time, the sino-vaginal bulbs invaginate from the urogenital sinus and join the caudal end of the fused mullerian ducts to form the vaginal plate.

Ultimately, the vaginal plate is reabsorbed and become canalized to develop the lower 1/3rd of the vagina. the whole process os reabsorption is completed by 24 weeks of gestational period.

Any defect of fusion or failure of reabsorption of lower portion of the mullerian ducts during early embryological life may result in forming bicornuate, septate or uterus didelphys.

There may or may not be septation of vagina as well.

The coincidence of unilateral renal agenesis on the same side of obstructed vagina can be explained by an embryologic arrest at 8 weeks of pregnancy, simultaneously involving renal (wolffian) and mullerian duct (metanephric) anomalies.

INTRODUCTION

In the year 1922, Purslow ce firstly describe a case of a young woman with regular menstruation had gradually increasing pelvic pain with presence of a pelvic mass after menarche.

Mostyn p and embrey b in 1950 described a case of obstructed hemi vagina and a uterus didelphys as well as an ipsilateral renal anomaly (5).

In the year 1971, Herlyn and Werner initially explained the syndrome as blind hemivagina with ipsilateral renal agenesis (6).

In 1976 Wunderlich described an association of right renal aplasia with bicornuate uterus & simple vagina in the presence of an isolated hematocervix (7).

Now a day, Herlyn-Werner-Wunderlich syndrome (HWWS) represents a complex female genital developmental defect like uterus didelphys / bi cornuate uterus or septate uterus with ipsilateral renal agenesis, with or without associated unilateral low vaginal obstruction (8).

DESCRIPTION OF CASE ----

A nulliparous muslim lady of 20 years age with 2 years married life presented in obs & gyn opd with complained of mild to moderate off and on pain in abdomen since last 2 to 3 months.

The pain in abdomen was not assocaited with nausea, vomitting, fever and abdominal distension. no past history of t.b., hypertension, and menorrhagia is noted.

As per history given to gynecologist , her menses were regular and menstrual bleeding usually lasted for 3 to 4 days .she had no history of getting pregnant / or abortion during 2 years of married life .

Her physical examination revealed normal vitals , thin built , average health and height, general conditions were stable.,pulse 7o/ m, b. p. 110/ 66 mm of hg,rr 22/mt.

Her abdominal examination did not reveal any mass on palpation.

Pelvic examination findings –
P/v uterus is normal in size & retroverted in position.
Bilateral fornices are free.
Mild tenderness in right fornix.
P/S cervix appears healthy.
Vaginal walls are normal. no vaginal septum is seen.

P/R

Retal examination revealed no abnormal findings.

The routine hematological and urine examination were normal except mild anaemia.

Abdominal sonography findings are 'malrotated right kidney with laterally directed renal hilum'. right kidney is showing duplex collecting sysytem. left kidney is not visualized—may be renal agenesis.

Urinary bladder is normal in outline.

Uterine cavity is divided into two with a septum –likely bicornuate uterus .however needs further evaluation to rule out uterus didelphys. both the ovaries are normal.

No free fluid is seen in cul-de-sac. Rest of upper abdominal sonography appears normal.

cect abdomen with renal angiography findings –
Malrotated right kidney with duplex collecting system.

Non-visualisation of left kidney confirmed the finding of left renal agenesis.

Uterine cavity is divided into two chambers by a septa, suggestive of bicornuate uterus.

Vertebral anamoly is also detected as hemivertebrae in distal lumbar vertebrae.

Rest other abdominal organs are normal.

Renal angiography confirms right main and accessory renal artery arising from aorta .non visualisation of left renal artery is demonstrated.

Intravenous pyelograhy revealed contrast filled duplex collecting system in malrotated right kidney. left kidney is not seen.

Fig-1 Usg Abdomen -- Left renal agenesis, malrotated right kidney with duplex collecting system.

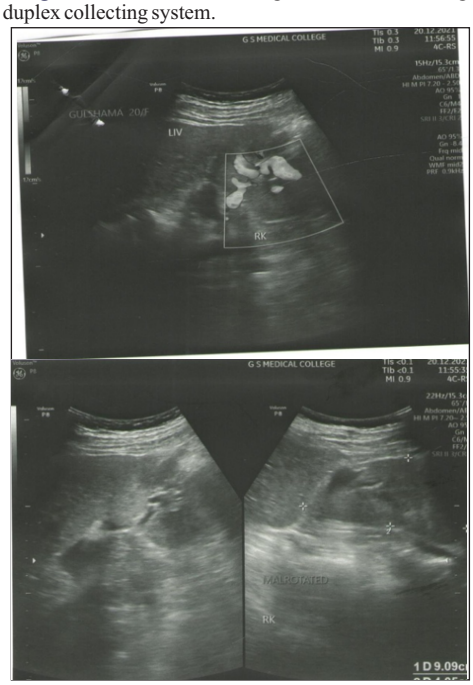


Fig-2 Ivp – Right kidney malrotated with duplex collecting system and left renal agenesis

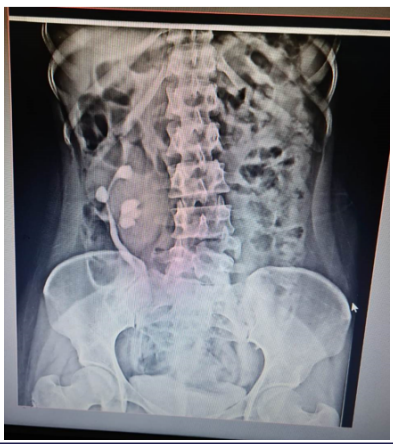


Fig 3 Cect Abdomen -- Malrotated Right Kidney With Left Renal Agenesis

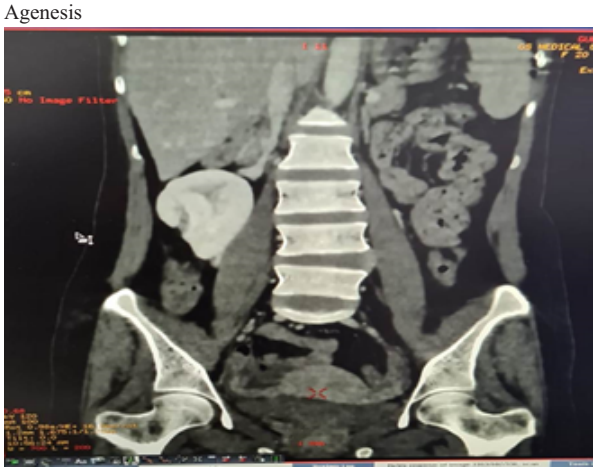


Fig 4 – Cect Pelvis-- Bicornuate Uterus.

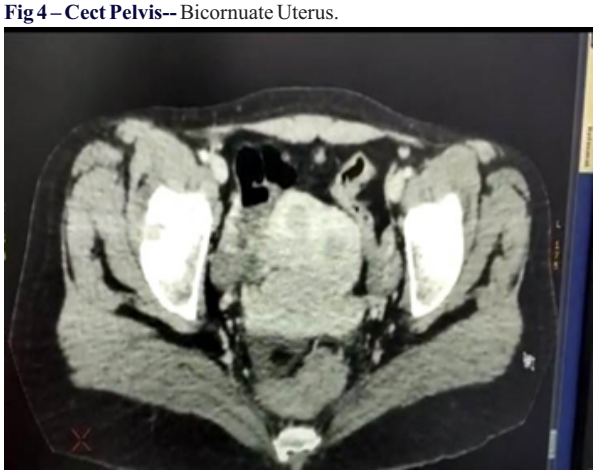
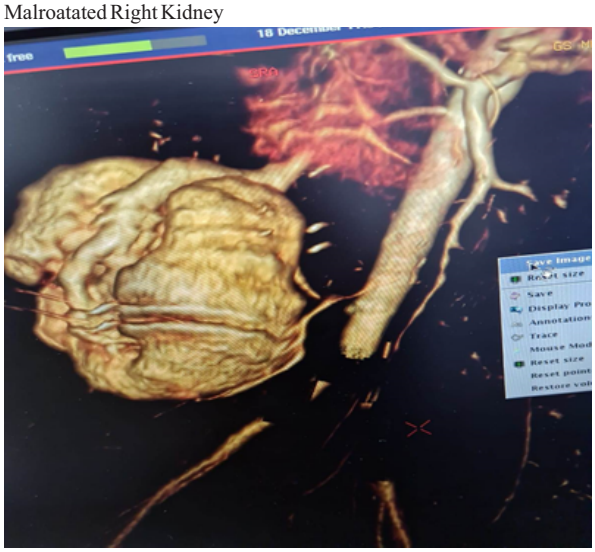


Fig-5 Renal Angiogrpahy Confirm Left Renal Agenesis And Malroatated Right Kidney



Above-mentioned clinical findings, history, physical abdominal, pelvic examinations and supported investigations confirmed that patient is having following findings summarised as follows:--

1. Mullerian developmental defect – as bicornuate uterus without a vaginal septum

2. Left renal agenesis as renal developmental defect

3. Right malrotated kidney with duplex collecting system –also renal developmental anomaly –an additional findings (not seen in other cases)

4. Lower lumber vertebral body defect as hemi vertebrae (not found in previously reported cases)

hence it is concluded that this case is a confirm rarest variant case of Herlyn-Werner-Wunderlich syndrome (type 2).

Lan Zhu,et al proposed new classification of Herlyn-Werner –Wunderlich syndrome based on a complete or incomplete obstructed hemivagina, according to this classification , in our case ,there was no vaginal obstruction owing to absence of vaginal septum (9).

The pros and cons of this syndrome was explained to the family members especially to her husband. gynecologist advised her to undergo further investigation on the line of primary infertility and referred to physician for pain in left flank of abdomen.

CLASSIFICATION:--

Zhu L Chen suggested new classification as HWWS be classified according to the complex or incomplete obstruction of the vagina as follows.

CLASSIFICATION 1, completely obstructed hemivagina.
Classification 1.1, with blind hemivagina.
Classification 1.2, cervicovaginal atresia without communicating uteri.
Classification 2, incompletely obstructed hemivagina.
Classification 2.1, partial reabsorption of the vagina septum.
Classification 2.2, with communicating uteri.

DISCUSSION —

Usually patient of classical HWWS becomes symptomatic after menarche. in cases of obstructed hemivagina with uterine anomaly presented with dysmenorrhoea, hematometra, hematocolpos, endometriosis, retention of urine.

A ten year review of this anomaly showed that 73% of patients presented with dysmenorrhea, 71% with pelvic pain or paravaginal mass.

While in our case the presenting symptoms were mild pain in left flank of abdomen and pelvis resembling mild acute abdomen off and on, this type of presentation was also reported by Aydin r who stated that the acute abdomen presentation is uncommon.

The diagnosis of HWWS is usually confirmed by sonography and cect scan abdomen & magnetic resonance imaging (MRI).

Sonography can detect the pelvic cystic mass and may detect the uterine malformations .however some author now consider MRI as gold standard for the diagnosis, as it provides better and detailed information about uterine contour , endometrial cavity and its extension into vaginal lumen, the type of septum , nature of fluid content ,presence or absence of endometriosis and associated renal anomalies.

Many types of urological malformations have been described with HWWS including renal duplication, Multicystic dysplastic kidney and even renal agenesis.

According of fedelel et all reported in his series that 72.4 % cases were of classic variety & 27.6 % of cases were variants of uterine septum or cervical agenesis (10).

In our case, there was no vaginal septum & absence of any pelvis mass. Mullerian duct anomaly detected is bicornuate uterus with renal agenesis on left side and malrotated right kidney with duplex pelvicalyceal system as renal anomalies.

Wunderlich in 1976 added the bicornuate uterus as a feature of the syndrome.

Hence our case presenting with Mullerian anomaly as bicornuate uterus & associated renal anomaly as renal agenesis of left kidney & malrotated right kidney with duplex pelvicalyceal system is a confirmed case of variant type of Herlyn-Werner-Wunderlich.

CONCLUSION —

Herlyn-Werner-Wunderlich syndrome is a rare Mulleriant duct & urogenital congenital anomaly. The aim of reporting this case is to highlight the entity and the importance of early management to relive the symptoms and to prevent the complications, since timely diagnosis requires a high index of clinical suspicion & radiological confirmation.

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