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Case Report

HERLYN-WERNER-WUNDERLICH SYNDROME- ONE OF THE RARE CAUSES OF LOWER ABDOMEN PAIN IN YOUNG FEMALE

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ABSTRACT

Herlyn-Werner-Wunderlich syndrome (HWWS), characterized by uterus didelphys, obstructed hemivagina, and associated with ipsilateral renal agenesis, is an uncommon combined Mullerian and mesonephric duct anomaly. We are reporting a case of 12 year old Female presented with lower abdominal pain, clinically as suspected to have lower abdominal mass. On subsequent radiological investigation patient was diagnosed to have uterus didelphys, obstructed left hemivagina, and ipsilateral renal agenesis. Patient was referred for further surgical management.

Keywords: *Uterus Didelphys, Hematometra Hematosalphinx*

INTRODUCTION

Herlyn-Werner-Wunderlich syndrome (HWWS) is often a misdiagnosed condition and need a high index of suspicion is required in patients with Mullerian and mesonephric duct anomalies. It usually presents at puberty with pelvic pain, the form of pyometra, ischiorectal swelling, urinary obstruction, and primary infertility. Since this condition can be treated by vaginal septum excision, and delay in diagnosis may worsen the associated endometriosis, early diagnosis is beneficial (1). This entity is also known as obstructed hemivagina and ipsilateral renal anomaly (OHVIRA). It is not to be confused with the Wunderlich syndrome.Wunderlich syndrome is a rare condition, in which spontaneous nontraumatic renal haemorrhage occurs into the subcapsular and perirenal spaces. Wunderlich syndrome is clinically characterized by the Lenk's triad viz 1**.**acute flank pain, 2. flank mass 3. hypovolemic shock. Magnetic resonance imaging (MRI) is the modality of choice for the diagnosis of HWWS and other such anomalies because of better anatomic delineation of pelvic structures and higher sensitivity for blood products.

CASES

A young adolescent girl aged about 12 years presented with lower abdominal pain for the past one month. Pain was more towards the left iliac fossa. It was more of a dragging pain and radiating to the back. Patient noticed a swelling in the lower abdomen towards the left side. Pain was not relieved with anti-inflammatory drugs. The patient had no history of fever, no vaginal discharge. She had attained menarche 2 months ago and has regular cycles of 28 days with flow for 3-5 days. Past and family history was not of significance. No other significant history was noted.

On clinical evaluation it was found that patient was moderately built and nourished and had normal vital signs. There was firm tender mass of size 4 x 4 cms with palpable in the left iliac fossa. Per vaginal examination was not done as patient was unmarried. Per rectal examination was done and Uterus could not be felt separately and external genital organs were normal.

Patient was then referred for USG. USG of the liver, pancreas, GB, right kidney and spleen were normal Left kidney is not seen in left renal fossa, LIF and in RIF. USG of bladder was normal

USG of the pelvis shows well defined cystic lesion with mobile internal echoes noted within the left ovary and two separate uterine corpus and fundus noted. Left uterus horn shows distended endometrial

cavity filled with fluid containing mobile internal echoes. Left adnexa show well-defined saccular elongated lobulated hypo echoic structure with incomplete septation and mobile internal echoes within it. Minimal free fluid in POD noted. Diagnosis was made as two separate uterine corpus and fundus – uterus didelphyus with obstructed left uterine horn causing left hematometra and left hematosalphinx with left ovarian endometrioma/ hemorrhagic cyst with absent left kidney.

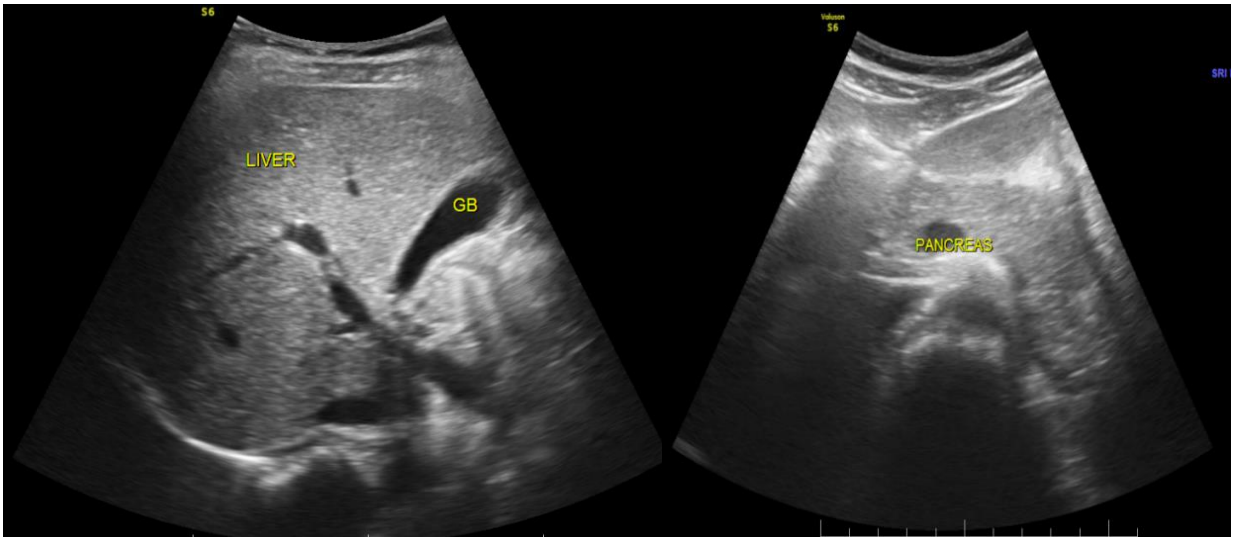

Figure1

Figure 2

Figure 1and 2: USG image showing normal liver, GB and Pancreas.

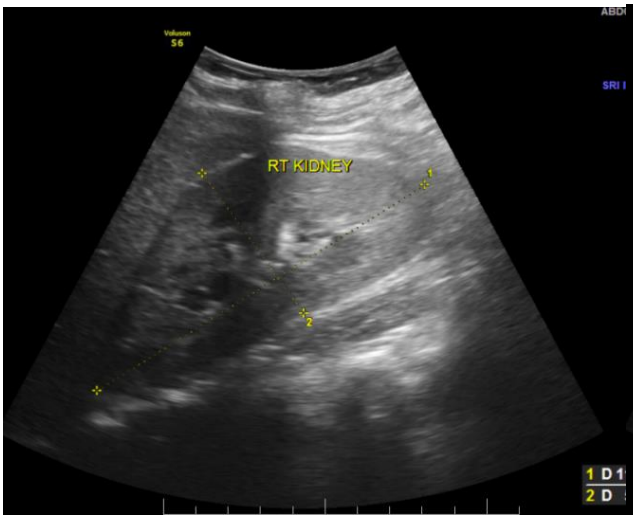

Figure 3

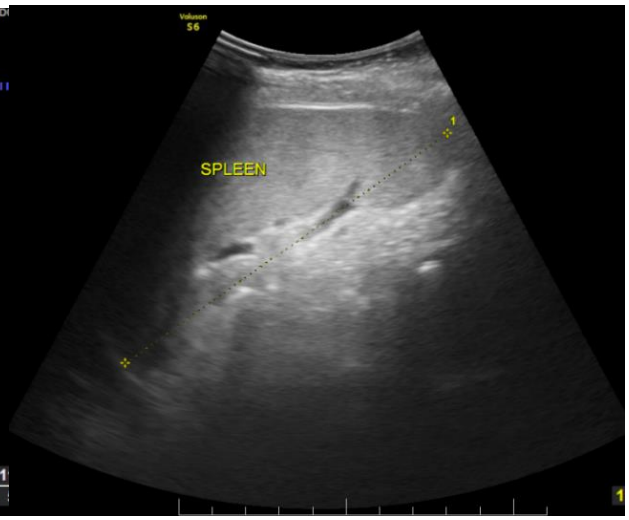

Figure 4

Figure 3 and 4: Normal right kidney in the right renal fossa, with non-visualization of left kidney into the left renal fossa, LIF, RIF and thorax.

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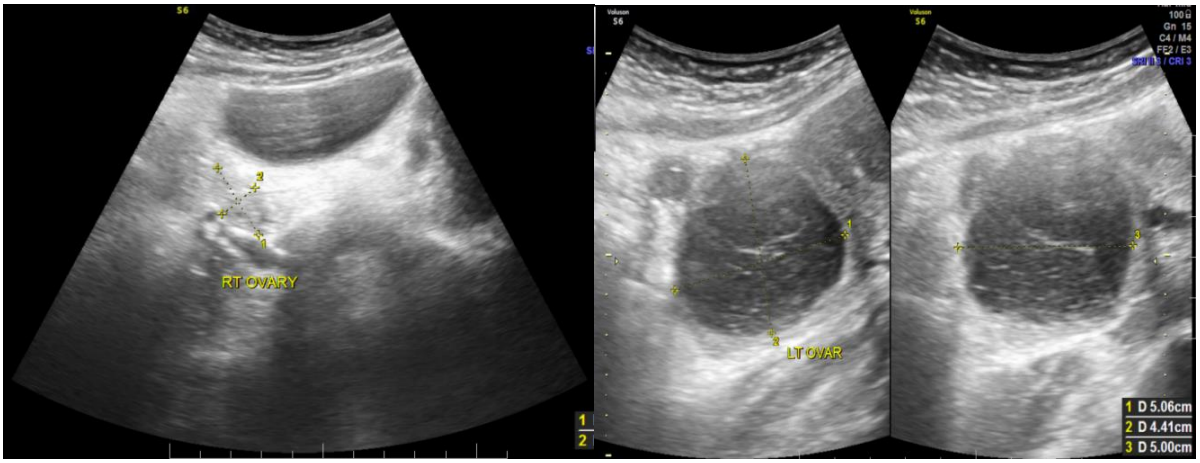

Figure 5 **Figure 6**

Figure 5 and 6: Normal right ovary in the right adnexa. Left ovary shows well defined anechoic cystic lesion of size measuring 5.1x4.4x5.0 cm with fine mobile internal echoes within s/o left endometrioma/ hemorrhagic cyst.

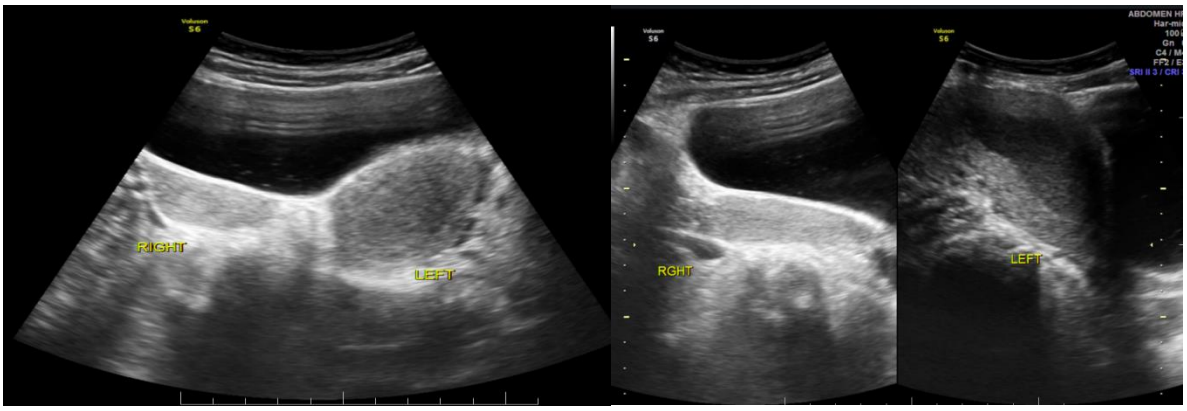

Figure 7 **Figure 8**

Figure 7 and 8: Shows two separate uterine horn with left endometrial cavity is distended with filled with fluid collection with fine mobile internal echoes.

The patient underwent MRI after this to confirm the diagnosis. MRI shows two separate uterine horn with hemorrhagic fluid collection in the left uterine horn, left follapian tube suggestive of left hydrometra and left hematosalpinx and endometriotic cyst in the left ovary.

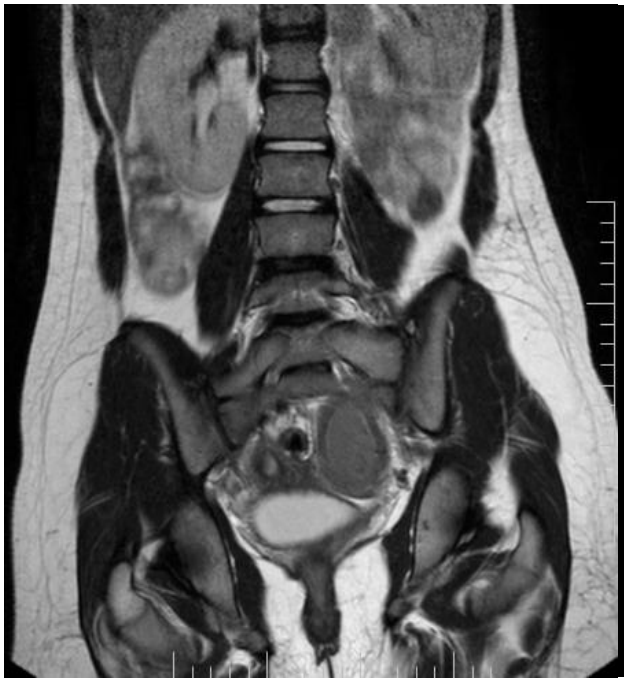

Figure 9: T2 Coronal

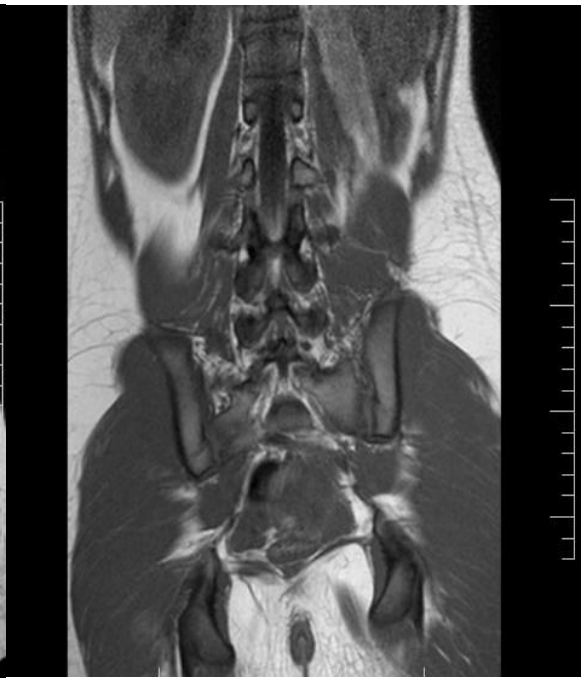

Figure 10: T1 Coronal

Figure 9 and 10: T2 and T1 images shows right kidney in the right renal fossa with absent left kidney.

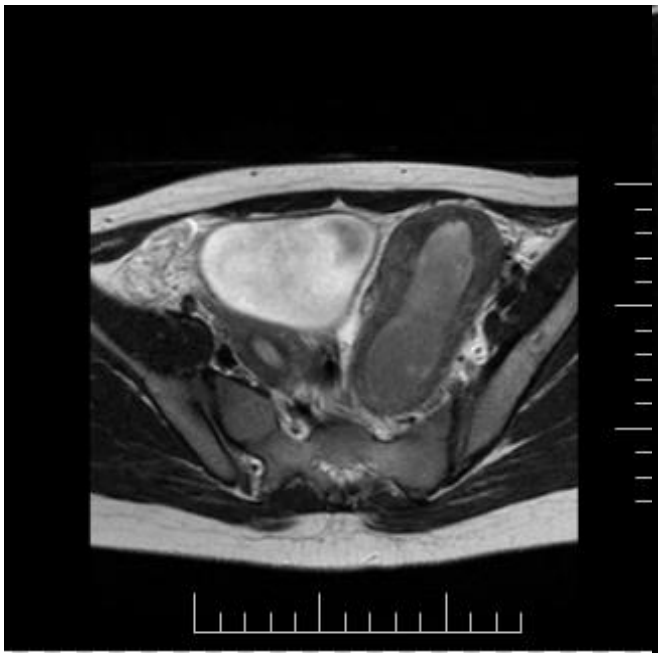

Figure 11: T2 Axial

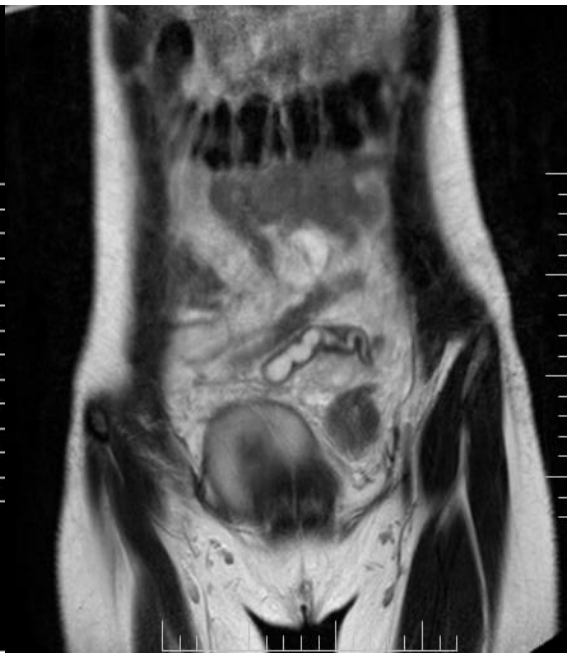

Figure 12: T2 Coronal

Figure 11 and 12: T2 Axial and Coronal images shows bicornuate uterus with fluid collection in the left uterine horn and in the left fallopian tube.

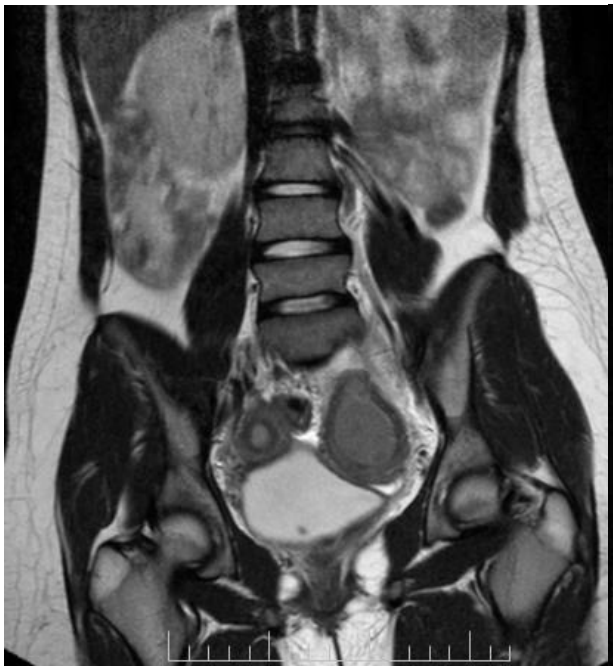

Figure 13: T2 Coronal

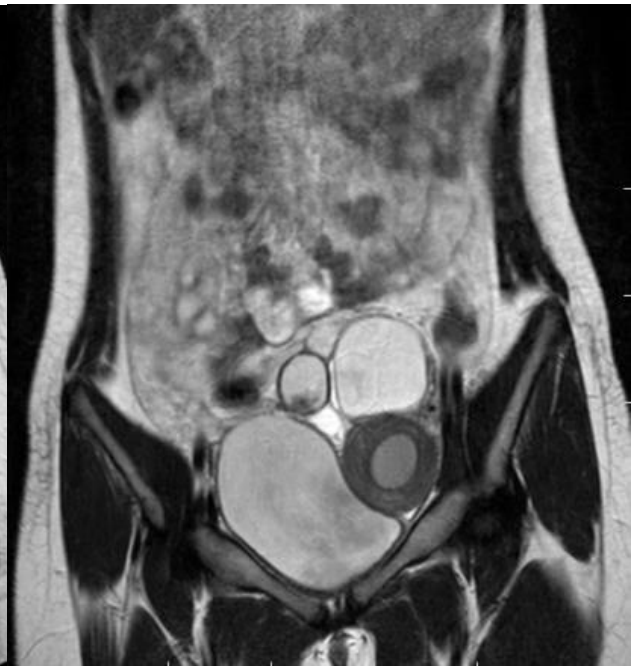

Figure 14: T2 Coronal

Figure 9 and 10: T2 Coronal images shows bicornuate uterus with fluid collection in the left uterine horn and in the left hematosalpinx and left hemorrhagic ovarian cyst

Hence, a diagnosis of uterus didelphys with left hematometra, hematosalpinx, and left hemivaginal obstruction with left ovarian endometriosis was made. Considering that she also had absent left kidney, a final diagnosis of HWWS or obstructed hemivagina ipsilateral renal anomaly (OHVIRA) syndrome was made.

DISCUSSION

Anomalies of the internal female reproductive tract referred to as the Müllerian duct anomalies, account for about 7% of female population. Even though the occurrence is sporadic, there is a strong association of congenital renal and ano-rectal malformations indicating common early developmental etiology.
Mullerian duct anomalies are classified according to the American fertility society criteria as follows:

- **Class I** or uterine hypoplasia or agenesis.
- **Class II**or unicornuate uterus: A banana - shaped uterus with a single fallopian tube. A rudimentary horn (communicating or non-communicating) may be present.
- **Class III**or uterus didelphys. Two complete uteruses, each with its own cervix. A sagittal vaginal septum is seen in the majority of cases.
- **Class IV** or bicornuate uterus. Two uterine cavities with one cervix. MRI shows widely separated uterine horns with an intercornual distance of >4 cm and concavity of the fundal contour or an external fundal cleft of >1 cm in depth.
- **Class V** or septate uterus: A fibrous septum is seen that appear hypointense on T2W images. While the muscular septum appears intermediate in intensity. MRI criteria includes a convex or flat external fundal contour or external fundal cleft of <1 cm in depth.
- **Class VI** or arcuate uterus: it is a normal variant and is characterized by an external convex contour of the fundus with fundal endometrial indentation.
- **Class VII** or diethylstilbestrol - induced: exposure to this synthetic estrogen antenatally can result in a T - shaped, hypoplastic and constricted uterus

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HWWS is a combination of Type III Mullerian anomaly with mesonephric duct anomaly with vaginal septum. The classic renal manifestation of OHVIRA syndrome is ipsilateral renal agenesis, but reports of duplicated kidneys, dysplastic kidneys, [2] rectovesical bands, [3] or crossed fused ectopia [4] have also been described.

As an obstructing vaginal malformation, this condition precludes the outflow of menstruation, resulting in hematocolpos, hematometra, and hematosalpinx. The diagnosis is therefore usually made soon after menarche due to symptoms related to the obstructed hemivagina, such as pelvic pain, dysmenorrhea, and a palpable pelvic mass. Long-term complications of OHVIRA syndrome include endometriosis from retrograde menstruation, and obstetric complications such as recur-rent pregnancy loss, preterm labor, abnormal fetal presentation, and prematurity due to uterine anomalies [5, 6].

CT and ultrasound are the 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. Hence, it should be obtained before any surgical intervention. Laparoscopy is not mandatory but could be helpful in confirming the diagnosis when radiologic imaging is inconclusive, especially in those cases with endometriomas warranting resection.

Current new classification of Herlyn-Werner-Wunderlich Syndrome based upon the morphology of vagina [10]

Classification 1, completely obstructed hemivagina

Classification 1.1, with blind hemivagina- The hemivagina is completely obstructed; the uterus behind the septum is completely isolated from the contralateral uterus, and no communication is present between the duplicated uterus and vagina. Hematocolpos may occur only a few months after menarche. Hematometra and hematosalpinx occurred in some more severely affected patients, as well as bleeding in the periadnexal and peritoneal space.

Classification 1.2, cervicovaginal atresia without communicating uteri- The hemivagina is completely obstructed; the cervix behind the septum is maldeveloped or atresic, and menses from the uterus behind the septum cannot outflow through the atretic cervix. Patients with this classification have similar clinical features as patients with Classification 1.1.

Classification 2, incompletely obstructed hemivagina

Classification 2.1, partial reabsorption of the vaginal septum -- In this type a small communication exists between the two vaginas, which make the vaginal cavity behind the septum incompletely obstructed. The uterus behind the septum is completely isolated from the contralateral uterus. The menses can outflow through the small communication, but the drainage is impeded. These patients have a later age of onset. The attack often comes years after menarche. Purulent or bloody vaginal discharge can be the chief complaints. Patients often have ascending genital system.

Classification 2.2, with communicating uteri- In this type, the hemivagina is completely obstructed, and a small communication exists between the duplicated cervices. Menses from the uterus behind the septum can outflow through the communication to the offside contralateral cervix. Because the communication is small, the drainage is still impeded.

Our case fit in to the Type 1.1. According to the new classification. As obstructive genital lesions may be associated with other anomalies such as coarctation of the aorta, atrial septal defects and abnormalities of the lumbar spine, a complete physical examination and abdominal tests may be indicated. [7] Resection of the vaginal septum is the treatment of choice of obstructed hemivagina[8]. A successful pregnancy is achieved eventually in 87% patients, while 23% have the risk of abortions. [9]

Conclusion

Evaluation of the genital tract by means of MRI scanning is recommended in all young females with known renal abnormalities detected before the onset of menstruation. This enables us to diagnose some patients before menarche and carry out a surgical correction of the vaginal obstruction before any damage has occurred because of Hematocolpos, hematometra and retrograde menstruation.

REFERENCES

Ahmad, Z, Goyal A, Das CJ, Deka D and Sharma R (2013). Herlyn–Werner–Wunderlich syndrome presenting with infertility: Role of MRI in diagnosis. *The Indian journal of radiology and imaging* **23**(3), 243.

Prada Arias M, Muguerza Vellibre R, Montero Sánchez M, Vázquez Castelo JL, Arias González M and Rodríguez Costa A (2005). Uterus didelphys with obstructed hemivagina and multicystic dysplastic kidney. *European Journal of Pediatric Surgery***15** 441-5.

Heinonen PK (2000). Clinical implications of the didelphic uterus: Long-term follow-up of 49 cases. *European Journal of Obstetrics Gynecology and Reproductive Biology* **91** 183-90

Tanaka YO, Kurosaki Y, Kobayashi T, Eguchi N, Mori K and Satoh Y, *et al.* (1998). Uterus didelphys associated with obstructed hemivagina and ipsilateral renal agenesis: MR findings in seven cases. *Abdominal Imaging* **23** 437-41

Rock JA, Jones HW Jr (1980). The double uterus associated with an obstructed hemivagina and ipsilateral renal agenesis. *American Journal of Obstetrics and Gynecology* **138** 339-342

Rock JA, Schlaff WD (1985) The obstetric consequences of uterovaginal anomalies. *Fertility and Sterility* **43** 681-692

Aveiro, Ana Cristina et al. (2011): "Herlyn–Werner–Wunderlich Syndrome: A Rare Cause of Pelvic Pain in Adolescent Girls." *BMJ Case Reports* 2011 bcr0420114147. *PMC*. Web. 28 Dec. 2015

Jindal G, Kachhawa S, Meena GL, et al (2009). Uterus didelphys with unilateral obstructed hemivagina with hematometrocolpos and hematosalpinx with ipsilateral renal agenesis. *Journal of Human Reproductive Sciences* 2 87–9

Candiani GB, Fedele L, Candiani M (1997). Double uterus, blind hemivagina, and ipsilateral renal agenesis: 36 cases and long term follow-up. *Obstetrics and Gynecology* **90** 26–32

Zhu L, Chen N, Tong JL, Wang W, Zhang L, Lang JH (2015). New Classification of Herlyn-Werner-Wunderlich Syndrome. *Chinese Medical Journal* **128** 222-5