ORIGINAL RESEARCH PAPER

Obstetrics & Gynaecology

KEY WORDS: congenital anomaly, hematocolpos, renal agenesis, mullerian duct anomaly.dysmenorrehea.

A RARE CASE OF HERLYN WERNER WUNDERLICH SYNDROME

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ABSTRACT

Herlyn Werner Wunderlich syndrome is a very rare female urogenital tract anomaly involving Mullerian duct anomaly and mesonephric duct malformation It is represented by a - uterus didelphys, ipsilateral renal agenesis, obstructed hemivagina which can be complete or incomplete. Usually presents after menarche with symptoms of abdominal pain, dysmenorrhea .lower abdominal mass secondary to hematometra or hematocolpos .We present a case report of17 year old female unmarried reported with white discharge since 4 months associated with dysmenorrhea . On examination foul smelling discharge was present .On clinical and diagnostic findings of hemivagina, renal genesis and hematocolpos. The diagnosis of HERLYN WERNER WUNDERLICH SYNDROME was confirmed.

INTRODUCTION:

Herlyn–Werner–Wunderlich syndrome, also known as OHVIRA (Obstructed Hemivagina and Ipsilateral Renal anomaly, is an extremely rare congenital condition involving malformations of the lower abdominal and pelvic organs. It is classified as a Müllerian duct anomaly, resulting from abnormal development during embryogenesis.

In most cases, OHVIRA is characterized by a double uterus—typically either uterus didelphys or a bicornuate uterus—accompanied by a unilateral obstructed (or blind) hemivagina and renal agenesis or other renal abnormalities on the same side as the obstruction. Additional anomalies may also be present, affecting the urethra, urethral sphincter, ureters, bladder, or even the spleen.

The underlying cause is a failure of vertical fusion between the paramesonephric (Müllerian) ducts and the urogenital sinus, leading to the formation of a transverse vaginal septum. This septum can obstruct menstrual flow, resulting in retention of menstrual blood in the uterus and vagina, which manifests as primary amenorrhea and cyclic pelvic pain. The exact prevalence of OHVIRA syndrome remains unknown, but it is estimated to occur in 0.1% to 3% of the population based on reported cases.

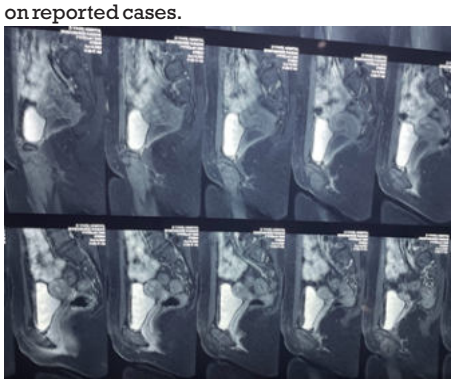

Figure 2: Hematocolpos in utenine cavity

Case Study

A 17 year old female unmarried reported with white discharge since 4 months associated with pain in abdomen during menses. And spotting pv after 4- 5 days of menses. On examination foul smelling discharge was present and usg suggestive of hematocolpos and right sided renal agenesis. MRI showed didelphus uterus and a uterine septum reaching the cervix. Patient was taken for hysteroscopy with unilateral hemi vaginoplasty right hemivaginal transverse septum and hematocolpos was drained.

The diagnosis of herlyn werner wunderlich syndrome was confirmed.

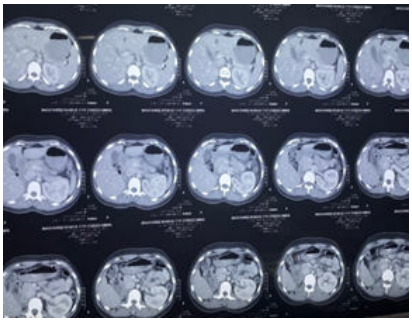

Figure 3 : Unilateral kidney

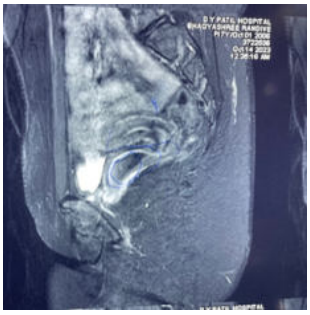

Figure 1: Hematocolpos seen in utrine cavity

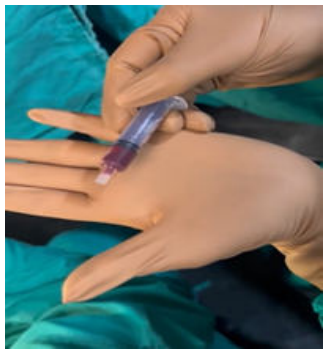

Figure 3: Hematocolpos drained

Usg Findings :

Uterus: Two separate endometrial cavity are seen in region of uterine body and fundus separated by maximum of 1.4cm. ET of right 10.0mm ET of left 9.5mm.

Kidneys: Left kidney normal in shape, position, echotexture. Right kidney not visualised in renal fossa ?ectopic ?agenesis.

MRI Findings: Uterus shows **didelphys morphology** with both horns closely abutting each other and septum reaching internal os.

3.5*3.4*5.3cm sized area/collection seen of altered signal intensity is noted in the vaginal canal 1.5cm from vaginal opening. suggestive of hematocolpos.

Cect Reports: **Right renal agenesis** is noted. Right kidney not visualised in renal fossa. Hematocolpus is noted in vaginal cavity and dilated and filled with blood products. Uterus shows morphology of bicornuate uterus.

CONCLUSIONS

HWW syndrome has variable onset of presentation. In patients with uterine and vaginal abnormalities, work up for associated renal anomalies should be performed. USG ,MRI can be done to know the extent of the syndrome. MRI is gold standard to make the diagnosis. Drainage of hematocolpos and to prevent further blockage the vaginal septum can be resected. Early intervention is needed as it help prevents infertility and endometriosis.

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