
Herlyn-Werner Wunderlich Syndrome: A Case Series And Review Of The Literature

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Abstract

Herlyn- Werner Wunderlich syndrome is a complex malformation involving the female urogenital system. It comprises of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis, thought to occur secondary to mesonephric duct induced mullerian anomalies. It classically presents in adolescent girls, soon after menarche. They present with recurrent and progressively worsening pelvic pain and primary dysmenorrhoea. A high index of suspicion is important for an early and accurate diagnosis. Magnetic resonance imaging plays an important role in establishing the diagnosis. Prompt treatment relieves acute symptoms and prevents complications of chronic cryptomenorrhoea, like endometriosis. We report four cases of Herlyn Werner Wunderlich Syndrome, who presented with varying clinical manifestations.

INTRODUCTION

Uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis, referred to as Herlyn-Werner Wunderlich (HWW) Syndrome, is a rare clinical entity. The incidence ranges from 0.1-3.8 %.(1) Most commonly diagnosed in adolescent females, soon after onset of menarche, due to presence of haematometrocolpos/ haematocolpos, secondary to obstructed hemivagina. Early and accurate diagnosis can, not only relieve acute symptoms, but also preserve fertility.

Delay in diagnosis, which is not uncommon, can lead to complications like pelvic adhesions, endometriosis, pyocolpos, pyometra etc. We present a series of four cases of this rare syndrome, who presented to the Gynaecology clinic with varying complaints.

CASE REPORTS

The patient profile, history and clinical examination, Imaging and the surgical management of all the four cases are described in Table1.

Table 1

Patient profile, clinical presentation, evaluation and management of the cases

Patient	Age at menarche (years)	Age at presentation (years)	Clinical Presentation	Clinical Examination	Previous evaluation (done elsewhere)	Imaging MRI § findings	Surgery done
1.	11	13	scanty menstrual flow, worsening dysmenorrhoea	PR * : 3X3 cm bulge felt through anterior rectal wall	USG †: absent right kidney, right haematocolpos	uterine didelphys, obstructed right hemivagina; right renal agenesis with hypertrophy of the left kidney	resection of longitudinal vaginal septum and drainage of haematocolpos
2	13	19	primary infertility, foul smelling vaginal discharge	uterus and cervix felt on right side, 4x4 bulge felt on left with thick vaginal septum	USG †- left ovarian cyst ; absent left kidney	uterus didelphys, transverse vaginal septum on left with ipsilateral haematocolpos and absent kidney. Compensatory hypertrophy of right kidney.	septum excision and marsupialisation of the edges, drainage of pyocolpos
3.	13	27	severe dyspareunia, foul smelling spotting per vaginam	uterus, normal size. Bulge felt on right side extending from fornx to the introitus	nil	uterine didelphys, right haematometroc olpos with transverse vaginal septum. Right renal agenesis, compensatory hypertrophy of the left kidney	excision and marsupialisation of the vaginal septum; drainage of the pyocolpos.
4.	12	13	polymenorrhagia	PR * : 3X3 cm mass felt on right side, around 3 cm from the introitus	USG † uterus didelphys, biscollis, absent right kidney. Left kidney hypertrophied.	uterus bicornuate, bicollis with duplication of vagina. Fluid filled in right hemivagina, with smooth tapering 1.8 cm from the introitus. Absent right kidney and adrenal. Malrotated jejunum.	Septum excision and marsupialisation

PR * : Per rectal examination
 USG †: Ultrasound
 MRI § : Magnetic resonance imaging

DISCUSSION

HWW syndrome is a Wolffian duct induced Mullerian anomaly, comprising of uterus didelphys, vaginal duplication, obstructed hemivagina and ipsilateral absent kidney. The precise etiopathogenesis of HWW syndrome is still unknown.

Embryologically, the internal genitalia and lower urogenital system develops from two pairs of Mullerian ducts and Wolffian (mesonephric) ducts. The two mullerian or paramesonephric ducts fuse to form uterus, oviducts and upper two thirds of vagina. The lower one third of vagina develops from fusion of uterovaginal canal (formed by fused paramesonephric ducts) with urogenital sinus. Arrest in development at 8 weeks’ of gestation leads to developmental abnormality of the mesonephric duct and ureteric bud, resulting in ipsilateral renal agenesis . Abnormality of mesonephric duct causes lateral deviation of ipsilateral mullerian duct, thereby preventing its fusion with contralateral mullerian duct and giving rise to uterus didelphys. Deviated mullerian duct is also unable to meet the

urogenital sinus centrally, hence, obstructed hemivagina develops. (2)

Patients, mostly postpubertal adolescent females, present with progressively worsening pelvic pain and primary dysmenorrhoea .Delay in diagnosis is due to presence of regular menstrual flow from unobstructed system, even in the presence of obstructed hemivagina. This was seen in two of our patients who presented several years after menarche, one with severe dyspareunia and second with primary infertility. Physical examination may reveal a pelvic mass on per rectal examination, but may not be feasible in very young patients.

Ultrasound helps in diagnosis of mullerian anomalies and detection of haematometra /haematocolpos (fluid collection with low level echoes), but it cannot identify the type of anomaly. Magnetic resonance imaging (MRI) with multiplanar image acquisition provides more detailed information of uterine morphology, cervices, patency of each vaginal lumen and associated endometriosis. It can provide information regarding the nature of low echogenicity contents (T1-W fat saturated hyperintensity suggestive of blood). It also reveals the absence of one kidney and the compensatory hypertrophy of the other as well as other associated anomalies (3)

HWW syndrome is, sometimes , associated with ‘thin basement membrane disease’ of the contralateral kidney. Patients may present with microscopic haematuria and recurrent urinary tract infections. Percutaneous needle biopsy of the kidney reveals thinning of the basement membrane of the glomeruli. (4)

Prognosis is good after surgical management. Definitive treatment includes resection of the obstructing vaginal septum, draining the collected blood or pus from the vagina and marsupialisation of the vaginal edges. (5) Marsupialisation is important to prevent recurrence of obstruction. (2) Timely intervention, not only relieves acute symptoms, but also prevents complications of chronic cryptomenorrhoea like endometriosis, pelvic adhesions , pyocolpos, pyometra and pyosalpinx.(1) Reproductive outcome is good after treatment, with 87% women having successful pregnancy. However, 15% may have preterm birth and 23% may have miscarriage.(5)

High index of suspicion in young girls presenting with progressively worsening dysmenorrhoea and pelvic pain, with presence of pelvic mass, will help clinch the diagnosis.

In all patients with mullerian anomalies, it is important to evaluate the urinary tract. Renal anomalies are found in 30% cases with mullerian anomalies due to embryological relationship between the two.⁽³⁾ In a female neonate with diagnosis of renal agenesis or multicystic dysplastic kidneys, it is prudent to look for genital malformations.

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