HERLYN-WERNER-WUNDERLICH SYNDROME: A LATE PRESENTATION

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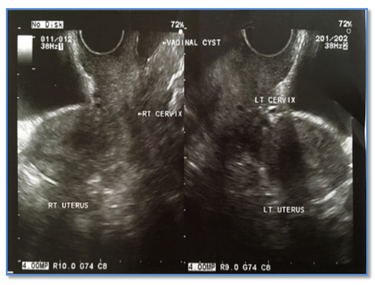
INTRODUCTION

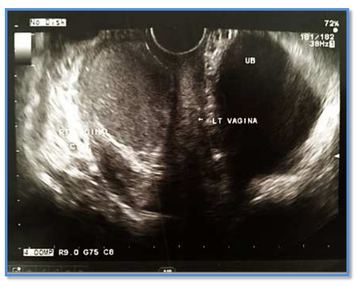
Mullerian duct anomalies are estimated to have an overall prevalence of 2% to 3% among all women. Herlyn-Werner-Wunderlich syndrome (HWWS) is characterized by uterine didelphys, obstructed hemivagina and ipsilateral renal agenesis. It is usually discovered at puberty with non-specific symptoms like increasing pelvic pain, dysmenorrhea and palpable mass due to the associated hematocolpos or hematometra, resulting from retained menstrual flow in the obstructed vagina.

CASE REPORT

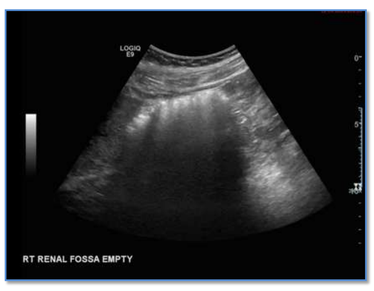
This is a case of a 31-year old, nulligravid, who presented with fever during the first day of her menses. This condition has been persistent for 1 year which was associated with dysmenorrhea, foul-smelling menstrual blood and occasional chills. On the third day of her menses, the fever usually subsides and recurs on the first day of the next cycle. On speculum examination, a cystic vaginal mass was seen occupying the right anterolateral wall of the vagina. On internal examination, the cervix was closed, effaced and deviated to the left. The corpus can not be well delineated. No adnexal mass noted.

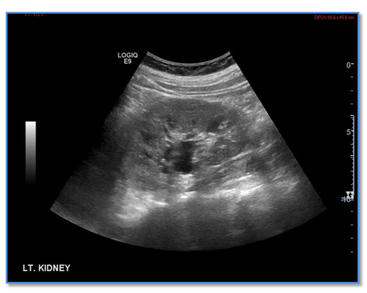
Transvaginal ulltrasound noted two uterine bodies and two cervices. Inferior to the right cervix, a cystic vaginal mass measuring 5.5 x 5.5 x 4 cm with ground glass appearance was also seen.



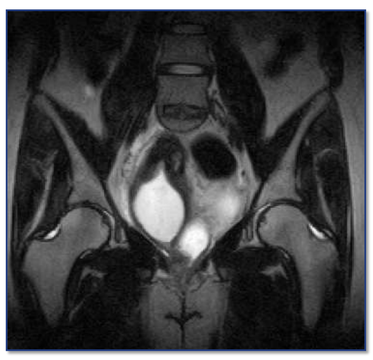


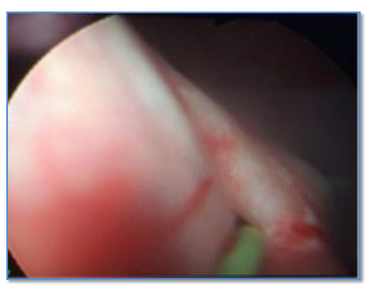
USD of KUB noted an empty right renal fossa while the left kidney was present with normal measurements and characteristics





Pelvic MRI confirmed that there are two widely spaced uterine corpora with one fallopian tube and cervix each. The two cervices were separated by a septum measuring 0.8 cm. No fistula or communicating channel was seen. The right cervix continues to a blind-ended vaginal dilatation which was filled with 72cc fluid. Both ovaries were normal. Impression was Uterine didelphys, Septated vagina with hematocolpos from obstructed right hemivagina and right renal agenesis suggestive of Herlyn-Werner-Wunderlich syndrome.

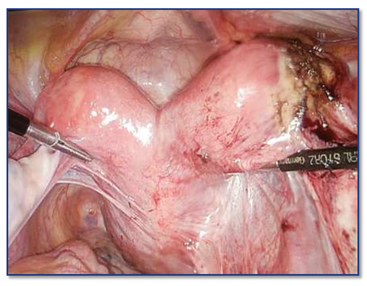




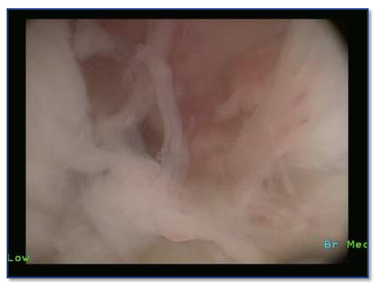
Cystourethroscopy confirmed the absence of right ureteral orifice. The left ureteral orifice was appreciated with efflux of urine. The rest of the bladder mucosa appeared normal.

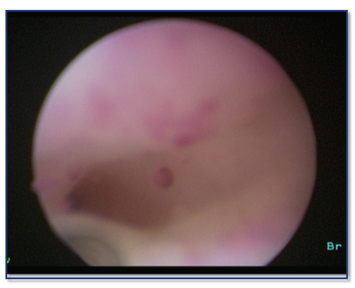
On vaginoscopy, a slit-like opening was identified at the left paravaginal wall probably the cervix of the left uterus. Vaginal rugae were normal-looking and intact. Along the right paravaginal wall is the previously noted bulging mass. Overlying rugae over this area appear flattened. No draining sinus was noted from this bulge. Drainage of the cystic vaginal mass was done. It was filled with brownish purulent foul-smelling fluid amounting to 200 ml.

Laparoscopy was done showing a uterine didelphys configuration. The right uterine horn was slightly larger then the left. Both ovaries and fallopian tubes were grossly normal. A bulging mass was seen at the cul-de-sac more on the right. Multiple puckered lesions and neovasularization were seen at the posterior cul-de-sac and posterior uterine horns. Lysis was done on the portion of the omentum which was adherent to the posterior surface of the right uterine body.

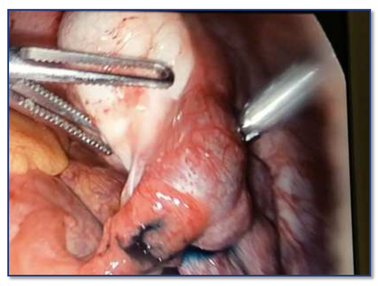


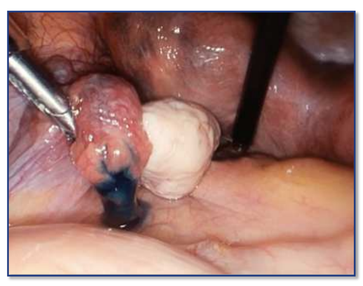
On hysteroscopy, the left uterus revealed thin endometrial lining with the left ostium seen. On the right uterus, slight endometrial thickening was visualized and right ostium was also appreciated.



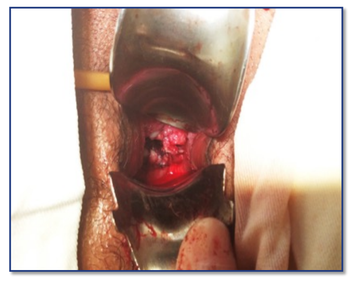


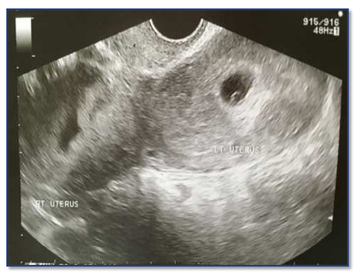
Egress of methylene blue was noted on both fallopian tubes when chromotubation was done.





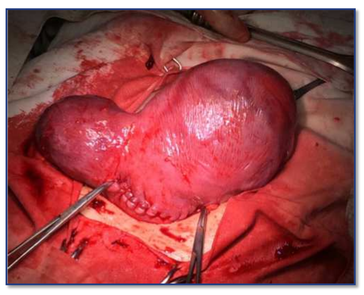
A 4-cm elliptical incision was made along the vaginal mass which allowed further evacuation of fluid content. Copius washing performed on the vaginal walls. Edges of the incision were clamped with Allis forceps and sutured with circumferential running interlocking sutures.

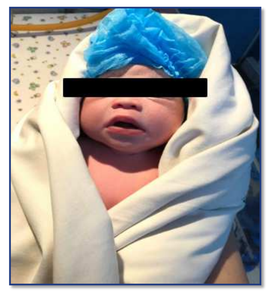




Regular menses resumed a month after the procedure with resolution of symptoms. On the fourth month, there was note of amenorrhea. Pregnancy test done at home was positive. Transvaginal ultrasound confirmed a pregnancy on the left uterus.

Antenatal care was maximized. She delivered via cesarean section to a healthy baby boy without complications and is currently living with her family without recurrence of symptoms.





DISCUSSION

Mullerian duct anomalies are congenital anomalies of the female genital tract which result from non-development or non-fusion of the mullerian ducts or failed resorption of the uterine septum during the sixth to ninth weeks of fetal life causing a wide-ranging series of reproductive ducts malformations.

Herlyn-Werner-Wunderlich syndrome usually presents with progressive and recurrent pelvic pain or enlarging abdominal mass after menarche. It is not initially diagnosed, because of the regular menstrual flow from the unobstructed vagina. Some time after menarche, the retention of menstrual blood in the obstructed hemivagina leads to the formation of a hematocolpos, which is usually clinically detected as a pelvic mass. However this patient presented with symptoms 19 years after menarche which is already late for majority of the cases.

Ultrasonography is a very helpful tool in the diagnosis of Müllerian duct anomalies, in fact the detection of haematocolpos, which appears as a fluid collection with low-level echoes, can make the diagnosis of genito-urinary tract anomaly easier. MRI is a suitable technique for the non-invasive evaluation of female pelvic anatomy because it provides more detailed information regarding uterine morphology, the continuity with each vaginal (obstructed and non-obstructed) lumen, and fluid content nature. However, laparoscopy should now be considered the gold standard for the evaluation of female reproductive tract anomaly. The decision to perform a laparoscopy in this case was based on the following: the interval between menarche and diagnosis, the severity of the symptoms, the presence of a hematometra or pyometra. For this patient, there is a long interval between menarche and diagnosis (19 years) which puts her at risk for long term complications: she has inflammatory responses exaggerated during menses and MRI findings reveal hematocolpos.

To alleviate symptoms and retain fertility in these patients, the most effective treatment is surgery. Resection of the vaginal septum is the treatment of choice of obstructed hemivagina. If treatment is delayed, complications may develop, such as endometriosis caused by retrograde menstruation, infections and pelvic adhesions, which in turn might cause obstruction of the genital organs.[7,9] This patient had cyclical dysmenorrhea and intraoperatively, adhesions were noted between the omentum and the right uterine horn. There were also multiple puckered lesions seen at the posterior cul-de-sac indicative of endometriosis. The underlying pathophysiological mechanism is potentially the increased risk of retrograde menstruation because of the obstructed vaginal outflow, the risk of endometrial tissue reflux through the fallopian tube, which can result in retrograde endometriosis, is higher.

The prognosis of HWWS is good with early diagnosis and early treatment. The outcome of pregnancy in these patients reveals that 87% go on to have a successful pregnancy with 62% having full-term pregnancies and uncomplicated deliveries. Women with uterine didelphys have a reasonable chance of getting pregnant, but the abortion rate is high (74%) and premature delivery is common (22%). A caesarean section is required in 82%. Shortly after the obstruction was relieved, this patient naturally conceived on the left uterus and had a stable antenatal, intrapartal and postpartal condition.

SUMMARY

We are presented with a reproductive-aged woman who manifested with signs and symptoms of an inflammatory process during her menses on her third decade of life. She has been wanting to get pregnant for the past two years but attempts have been unsuccessful. After a year of presenting with fever, foul-smelling menstrual blood, dysmenorrhea and chills, medical consultation revealed that she has a congenital urogenital anomaly namely uterine didelphys, obstructed right hemivagina and an ipsilateral renal agenesis. These are the characteristics of Herlyn-Werner-Wunderlich syndrome. Manifestations were delayed because of the unobstructed menstrual flow from the left uterus (unobstructed side). Laparoscopy, cystourethroscopy, vaginoscopy, hysteroscopy and excision of the obstructing hemivagina were done. These procedures confirmed and treated her mild endometriosis from the retrograde menstruation of the obstructed uterus, evacuated the hematometra and hematocolpos as well as relieved the displacement of the contralateral uterus and cervix. A month after the treatment, there was notable relief of symptoms and regular menses resumed. She naturally conceived on the left uterus after four months. Pregnancy was carried to term without complications. Delivery was uneventful. She is now happily living with her family and thankful for the improvement in her quality-of-life.

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