PREGNANCY IN HERLYN WERNER WUNDERLICH SYNDROME: TWO CASE REPORTS

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ABSTRACT

Herlyn-Werner-Wunderlich (HWW) syndrome is a rare congenital anomaly of the Mullerian duct system presenting with the triad of *uterine didelphys, obstructed hemivagina* and *ipsilateral renal agenesis*. The patient typically presents with dysmenorrhea evolving into continuous pelvic pain, and a palpable pelvic mass. Symptoms primarily occur after menarche. The HWW syndrome is commonly linked with obstetrical complications including infertility, abortion, and preterm delivery. To obviate these disastrous complications, it is essential to have a high level of suspicion for prompt recognition and treatment. We are presenting herein two (2) cases of HWW syndrome with contrasting management approaches resulting to different fertility profiles and obstetrical complication. Prompt diagnosis and appropriate treatment of this syndrome can forestall potential complications like pain and infertility. While the first case suffered infertility and preterm labor, the second case had normal fertility and reached full term.

CASES OF HERLYN WERNER WUNDERLICH SYNDROME

CASE 1

- 28 years old, G3P2(0020)
- Pregnancy Uterine 31 weeks and 4 days age of gestation, in labor
- Diagnosed with Herlyn-Werner-Wunderlich syndrome at age 13
- Previous surgeries include:
  - 1999 Right salpingo-oophorectomy for a ruptured Endometriotic cyst
  - 2000 Adhesiolysis with Right Hemi-hysterectomy
  - 2014 Diagnostic cystourethroscopy, Ultrasound guided drainage of the paravaginal mass, Excision of the longitudinal vaginal septum
- Underwent primary low segment cesarean section at 35 weeks and 5 days age of gestation
- Delivered a live baby girl 37 weeks by pediatric aging, 2490 grams, with APGAR score of 9 remaining 9, appropriate for gestational age

CASE 2

- 28 years old, G4P2 (2-0-1-2)
- Pregnancy Uterine 38 weeks gestation, in labor
- Diagnosed case of Herlyn-Werner-Wunderlich syndrome
- Underwent excision of vaginal septum at a tertiary government hospital 6 years prior to conception
- Delivered via repeat low segment cesarean section with bilateral tubal ligation.
- Delivered a live baby boy, 38 weeks by pediatric aging, 2900 grams, appropriate for gestational age with APGAR score 9,9

DISCUSSION

In patients with HWW syndrome, symptoms result from the obstructing vaginal malformation including cyclical lower abdominal pain, which later evolve into persistent severe pain due to increasing distention from the obstruction. Embryological arrest during the 8th week of embryological development is responsible for the occurrence of a didelphys uterus, which is the most common type of obstructing malformation of the uterus and vagina, together with an obstructed hemivagina. When the partition between the fused Mullerian ducts disappears incompletely, it persists as the longitudinal vaginal septum. Reproductive performance of patients with HWW syndrome may be affected with delayed diagnosis leading to increased risk of abortion and infertility. Literature shows that while abortions occur in 23% and preterm births in 15% of these patients, 87% continue with a successful pregnancy while 62% advance into full-term pregnancies and uncomplicated deliveries. The treatment of choice for an obstructed hemivagina is resection of the vaginal septum to relieve obstruction and prevent the retrograde flow leading to endometriosis. Currently, hemihysterectomy is rarely indicated and no more preferred. The challenge in pregnant patients with uterine malformations is primarily pregnancy maintenance than fertility. Literature is full of data showing its association with adverse obstetric outcomes including increased incidence of spontaneous abortion, malpresentation, placental abruption, intrauterine growth restriction, prematurity, operative delivery, retained placenta and fetal mortality. Our first case suffered long years of infertility, two spontaneous abortions, and preterm labor in the current pregnancy. With prompt diagnosis and appropriate management, normal fertility is expected with these patients having a reasonable chance of getting pregnant. This is well illustrated by our second case who reached a full-term pregnancy with no obstetric complications.

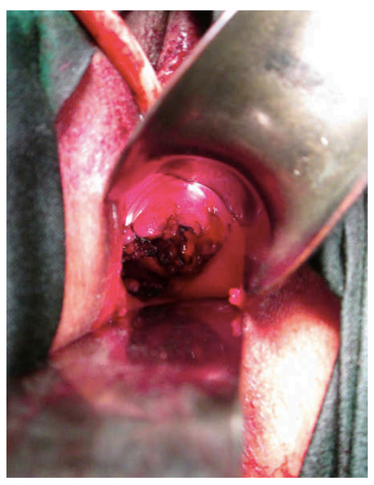

FIGURE 1. Excised vaginal septum with sutured margin (Case 1)

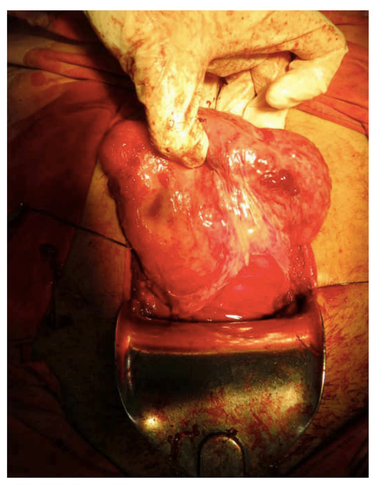

FIGURE 2. Uterus showing incision site at the lower uterine segment (Case 1)

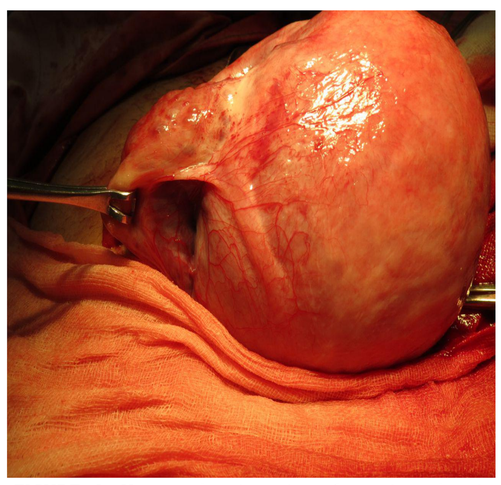

FIGURE 3. Uterus showing site of previous hemi-hysterectomy (Case 1)

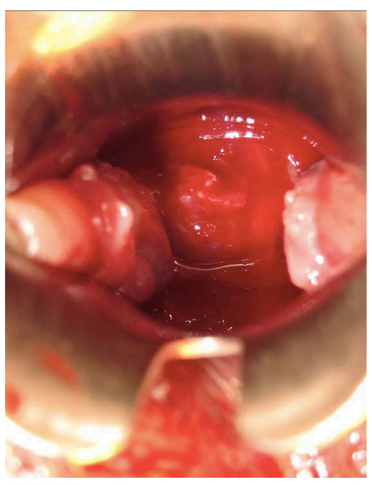

FIGURE 4. Speculum exam showing 2 cervices (Case 1)

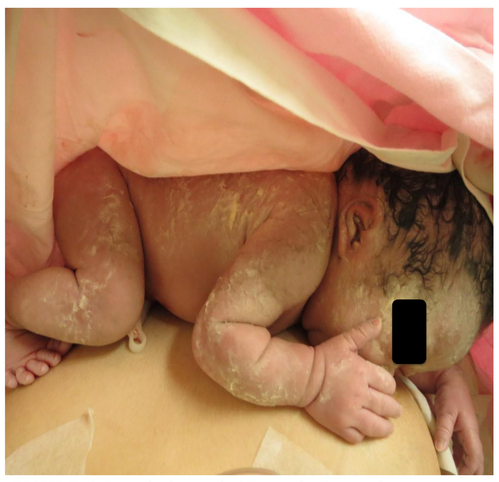

FIGURE 5. Live baby girl. 37 weeks by pediatric aging, 2490 grams, with APGAR score of 9 remaining 9, appropriate for gestational age (Case 1)

CONCLUSION

We have illustrated two (2) cases of pregnant patients with Herlyn-Werner-Wunderlich syndrome. A high index of suspicion in a patient with early-onset dysmenorrhea presenting with an abdominopelvic or vaginal mass would lead to a timely diagnosis and prompt management. Relief of obstruction with excision of the imperforate vaginal septum is the ideal surgical approach to prevent fertility complications. Pregnancy maintenance remains a problem with numerous adverse maternal and fetal outcomes reported in the literature. Proper counseling of pregnant patients with uterine anomalies and antenatal surveillance for intrauterine growth restriction, malpresentation, and preterm labor are essential. With prompt recognition and surgical relief of obstruction, patients with HWW syndrome may have good reproductive outcomes.

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