An eventful journey from menarche to successful motherhood

ABSTRACT

Herlyn–Werner–Wunderlich syndrome is an uncommon variant of mullerian duct anomaly and the approach to its diagnosis requires a high index of suspicion and vigilant work up. Presented here is a case of a 26-year-old woman who had the aforementioned anomaly and was pursued for nearly 10 years to provide her with a fruitful obstetric outcome after having undergone Strassman’s metroplasty at a young age of 15 years.

KEY WORDS: Herlyn–Werner–Wunderlich syndrome, mullerian duct anomalies, Strassman’s metroplasty

INTRODUCTION

Mullerian duct anomalies (MDAs) are rare and to witness a successful pregnancy outcome after surgical correction of the same is a delightful experience for the treating clinician. MDA results from nondevelopment (agenesis or hypoplasia), defective vertical or lateral fusion, or resorption failure of the mullerian ducts.[1] In the general population, the prevalence of these anomalies ranges from 0.001% to 10% while in women with bad obstetric history it ranges from 8% to 10%.[2,3] Didelphic uterus is seen in about 11% of MDA cases and are associated with unilateral anomalies, i.e., obstructed hemivagina and ipsilateral renal agenesis in 15–30% of cases.[4] This association of didelphic uterus with obstructed hemivagina and ipsilateral renal agenesis has been referred to in the literature as the Herlyn–Werner–Wunderlich syndrome (HWWS).[5,6] The term obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome is used to denote the triad of OHVIRA and uterine anomaly other than didelphys.[7] Both the syndromes are rare and are discussed as same entities in many case reports and their incidence is estimated to be between 0.1% and 3.5% of all mullerian anomalies.[8] The cause is attributed to embryologic arrest affecting both the mullerian and metanephric ducts concurrently at about 8 weeks gestation.[9] The most common presenting symptoms are dysmenorrhea within first few years of menarche and increasing pelvic pain. A palpable mass can be encountered due to the associated hematocolpos or hematometra, which result from retained menstrual blood in the obstructed vagina. Due to their extreme, the diagnosis is often missed or delayed. Narrated here is an interesting case of a 26-year-old woman with HWWS who had an eventful journey from her menarche to successful motherhood.

CASE REPORT

A 26-year-old primigravida was admitted from our antenatal clinic at 19 weeks gestation with threatened abortion. She had a history of having undergone abdominal metroplasty at the age of 15 years. She was a familiar face as she had been a frequent visitor at our gynecological outpatient clinic with complaint of severe dysmenorrhea since her menarchial age of 13 years. Her early symptoms included irregular menses,
progressive pelvic pain, constipation, and rectal pain during defecation. She was put on medical therapy, but her symptoms remained intractable. Her clinical workup showed an ultrasonography report suggestive of bilateral cystic ovarian mass with absent left kidney. Diagnostic laparoscopy was performed which revealed two hemiuteri out of which the left hemiuterus was markedly distended. Since the laparoscopic findings were suggestive of gross mullerian anomaly and the magnitude of symptoms was deteriorating the patient’s quality of life, laparotomy was carried out subsequently after a week. The findings of diagnostic laparoscopy were confirmed on laparatomy, showing a didelphic uterus with obstructed hemivagina and absent left kidney. As the hemiuteri were hypoplastic, Strassman’s metroplasty was performed to drain the left noncanalized hemiuterus and to create a single uterine cavity with a bigger caliber, aiming to provide better prospects for her future obstetric career. Postoperatively, the patient was greatly relieved of her symptoms and in due course of time, she got married in 2007. Following this, she had tedious 7 years of primary infertility and was under regular follow-up. Both the partners were investigated for infertility. Her husband’s Semen analysis was found normal. The hormone profile (luteinizing hormone, follicle stimulating hormone, and estradiol), serum prolactin, and thyroid profile of the patient were normal. Hysterosalpingography was performed which revealed a deformed uterine cavity resembling an arcuate uterus with bilateral spillage of dye. Finally, she conceived spontaneously and was admitted at 19 weeks with threatened abortion. On examination, her pulse rate was 80/min, and her blood pressure was 120/70 mmHg. Her cardiovascular system examination was normal. On per abdominal examination, her uterus corresponded to 20 weeks, and a gentle speculum examination revealed an old clot with no active bleeding and her cervical os was closed. An ultrasonography was done immediately to ascertain fetal well-being and cervical length which was found normal. She was planned for institutional observation and strict surveillance throughout her pregnancy till safe confinement. Her surveillance program included maternal and fetal monitoring with regular ultrasonography scans to estimate cervical length, rule out intrauterine growth retardation, and Doppler studies to detect placental invasion. All her antenatal investigations were reported normal, and we had planned her delivery at 37 weeks gestation by elective cesarean section. In anticipation of preterm labor, she was administered corticosteroids. She was carrying her pregnancy well until at 36 weeks 6 days gestation, she developed a premature rupture of membranes and was taken for emergency cesarean. A 2.2 kg male baby was delivered. Her intraoperative findings revealed a unified arcuate-shaped uterus having a deep midline scar [Figures 1 and 2]. The posterior surface of the unified uterus revealed the remnant rectovesical ligament which was transected during previous procedure of Strassman’s metroplasty [Figure 3].
The placenta was delivered with ease, and there were no intraoperative complications. Cesarean was successfully conducted followed by an uneventful postpartum period, and our patient went home on 7th day postoperatively with a healthy baby.

**DISCUSSION**

MDA is an infrequent finding in routine clinical practice and often their accurate diagnosis and complete assessment are missed. Most of the patients with MDA present during their reproductive years with infertility, recurrent pregnancy loss, prematurity, and other obstetric complications, whereas in pubertal age group, the symptoms include amenorrhea, cryptomenorrhea, or dysmenorrhea. A symptom such as dysmenorrhea is often treated conservatively, and the question arises as to what extent we can go while evaluating a case of primary dysmenorrhea. This was the salient feature of our case where a young teenage girl presenting with agonizing dysmenorrhea was evaluated to the extent of diagnostic laparoscopy followed by laparotomy and surgical correction. Our patient had a rare variant of MDA known as the HWWS with a didelphic uterus, obstructed left hemivagina, and left renal agenesis. This was a striking feature contrary to the evidence provided in the literature of predominance of right-sided renal involvement.[9]

While investigating such rare cases, the imaging modalities are of variable utility as the hematometra and the hematocolpos frequently distorts the pelvic anatomy leading to inconclusive results. Laparoscopy and laparotomy prove to be appropriate in defining the abnormality and treating it as seen in our case.[9,10] It was noteworthy to see the compliance of our patient for nearly a decade and how we pursued her to accomplish the goal of successful motherhood after corrective Strassman’s metroplasty.

In an era where laparoscopic surgeries are more rampant, to witness a fruitful obstetric outcome in a patient who underwent abdominal Strassman’s metroplasty provides relevance to this lesser practiced technique. Moreover, it also finds its place in low-resource set up like ours, where facilities for laparoscopic surgeries are unavailable. Our case was a great learning experience and a boost for us because our endeavors were not only merely providing symptomatic relief to our patient but also giving her a promising obstetric career ahead.

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**Conflicts of interest**

There are no conflicts of interest.

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