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Case Report

An interesting case of Herlyn–Werner–Wunderlich syndrome

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Abstract

Herlyn–Werner–Wunderlich syndrome (HWWS) is a rare congenital anomaly characterized by uterus didelphys with blind hemivagina and ipsilateral renal agenesis. Usually, such patients present with dysmenorrhea shortly after menarche, increasing pelvic pain and a palpable mass due to the obstructed hemivagina. Interestingly in the present case, the patient had her menarche seven years ago, but dysmenorrhea started only 1-year back. She never sought medical help previously as she was mostly asymptomatic all through the years. It was only after she conceived and got investigated for antenatal concerns that she was found to have HWWS. A tortuous history and an unusual clinical presentation made this case an interesting one.

Keywords: *Herlyn–Werner–Wunderlich syndrome, Mullerian duct anomaly, Pandora's box*

Introduction

Herlyn–Werner–Wunderlich syndrome (HWWS) is a rare variant of mullerian duct anomalies which was first described in 1922 in a young woman with regular menstruation and gradually increasing pelvic pain and a pelvic mass after menarche [1]. HWWS comprises of didelphic uterus with obstructed hemivagina and ipsilateral renal agenesis [2,3]. The common presenting symptoms are dysmenorrhea shortly after menarche and increasing pelvic pain. An abdominal mass secondary to hematometrocolpos from retained menstrual blood is another common finding [4]. In the present case, the patient surprisingly had her menarche 7 years back but experienced dysmenorrhea recently. She never sought medical help previously as she was mostly asymptomatic all through the years. It was only after she conceived and got investigated for antenatal concerns that the surprise sprung up.

Case report

A 19-year-old female who had been married for 8 months presented with a history of amenorrhea for the past 4 months and a progressively growing lump abdomen for the same duration. When she was 1 month overdue her periods, a urine pregnancy test (UPT) was performed at a primary health center which was positive and was thus antenatally registered there. She was apparently well until 4 days back when she had bleeding per vagina, and her abdomen was found distended more than her period of amenorrhea, therefore, was referred to our hospital as a case of primigravida with 4 months' amenorrhea with suspected twin gestation and threatened abortion. There was no ultrasonography (USG) available with the patient. On examination, she had mild pallor, and her vitals were normal. Her cardiovascular system examination was normal. Abdominal examination revealed a relaxed uterus corresponding to 20 weeks' gestation and fetal heart sound was not localized on stethoscope. On per speculum examination, minimal bleeding was seen. An immediate USG was done which showed 2 separate uterine horns with left horn showing echogenic foci of 1.5 am × 1 cm size suggestive of retained products of conception. There was also a right-sided 10 cm × 10 cm × 8 cm cystic lesion which was suspected to be endometriotic. Another striking finding was the absence of the right kidney. With suspicion of HWWS with incomplete abortion, the patient was put through a thorough investigative workup. Meanwhile, the patient passed a fleshy mass per vagina following which a repeat USG showed empty left horn suggesting complete abortion. The blood investigations reported hemoglobin 9 gm%, serum β-Hcg = 0.1 mIU/mL, CA125 = 50 U/mL, and alpha-fetoprotein and lactate dehydrogenase within normal range. An intravenous pyelogram showed the absence of right kidney which was seconded by a magnetic resonance imaging that also reported a didelphic uterus and a right ovarian cyst [Figures 1 and 2]. A meticulous menstrual history elucidated that patient attained menarche at 12 years with regular cycles and dysmenorrhea had begun just 1 year back. The patient was posted for examination under anesthesia (EUA), where on per speculum examination, a single external os and cervix were visualized. There was no obvious bulging septum. A uterine sound was carefully negotiated

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through the visible os which freely went toward the left side but was not negotiable in the right direction. An exploratory laparotomy was subsequently performed in view of Mullerian anomaly with suspected endometriotic cyst due to right-sided outflow obstruction. On laparotomy, there was a right-sided mucinous cystadenoma with didelphic uterus, normal fallopian tubes, and normal left ovary [Figures 3 and 4]. Intraoperatively, the right cyst adenectomy was performed preserving the right fallopian tube and aspiration from the right hemiuterus yielded minimal fluid. There was no evident hematometra, and on palpation, the right renal bed was found empty while the left kidney was present. A thorough counseling of the patient and her attendants was done regarding the anomaly and her future obstetric prospects. The patient underwent vaginal septum resection with Strassman's metroplasty and was discharged on the 12th postoperative day.

Discussion

The incidence of HWWS varies between 0.1% and 3.8% and is attributed to embryologic arrest affecting both mullerian and metanephric ducts at about 8 weeks' gestation [2,5,6]. The common presenting symptoms include dysmenorrhea, increasing pelvic pain and abdominal mass secondary to hematometrocolpos due to obstructed vagina. Infection of the retained clotted blood result in pelvic inflammatory disease and tubo-ovarian abscess which could also lead to urinary retention in some cases secondary to the enlarging lump [7]. From obstetric aspect, HWWS can present with infertility, recurrent miscarriages, preterm labor, and malpresentations [8]. Our case was interesting because a rare entity like HWWS was initially referred from a rural health center as a case of twin pregnancy with threatened abortion. This referral diagnosis was based on history of 4 months' amenorrhea, a positive UPT and overdistended abdomen. Furthermore, this was due to the lack of USG at rural health center. When this case was investigated at our institution, it was surprisingly found to be a case of HWWS with complete abortion and a coexistent right ovarian cyst. Since the tumor markers were found to be within normal range except CA125 which was slightly raised it prompted us to consider the possibility of an endometriotic cyst. Predicament arose when EUA revealed no septal bulge suggesting minimal collection or obstruction, while on the other hand, the distended

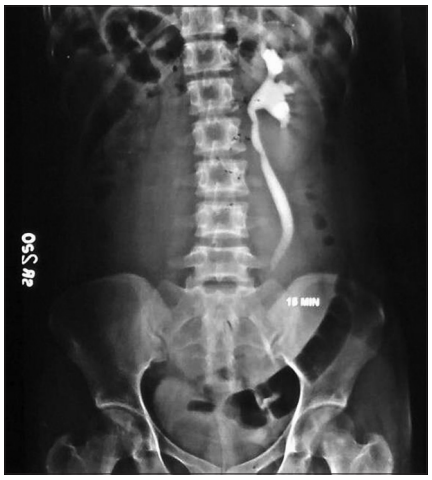

Figure 1: Intravenous pyelogram showing absent right kidney

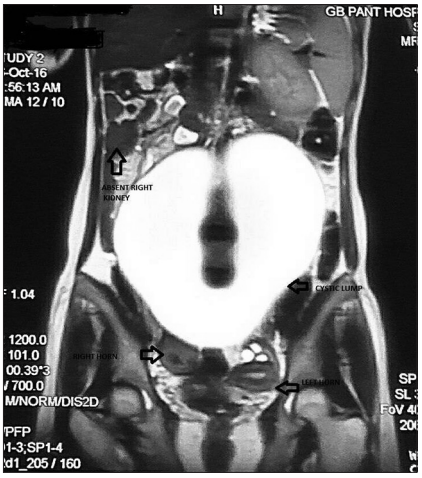

Figure 2: Magnetic resonance imaging showing didelphic uterus with a huge cystic abdominal lump and absent right kidney

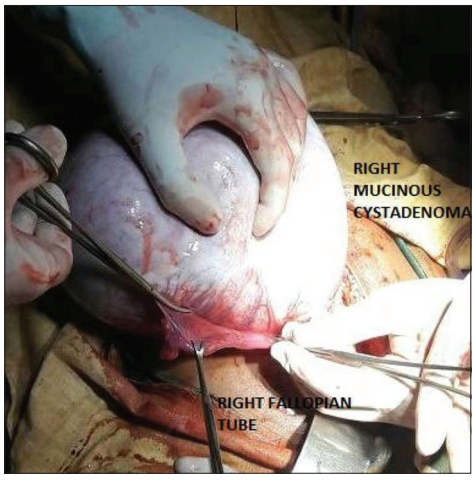

Figure 3: On laparotomy huge right-sided mucinous cystadenoma seen with normal right fallopian tube

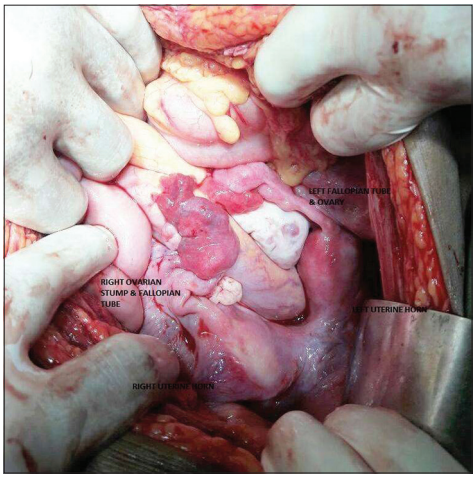

Figure 4: Postcystadenectomy the didelphic uterus is uncovered. The two uterine horns are seen with normal fallopian tubes, normal left ovary and the right ovarian stump. No evidence of endometriosis seen elsewhere

abdomen kept us thinking about hematometrocolpos. This was because hematometrocolpos is associated with HWWS as result of obstructed hemivagina. Nonetheless, as the lump had grown in a quick span of 4 months and initially mimicked pregnancy, hematometrocolpos was our last differential diagnosis.

The management of HWWS requires thorough and careful anatomic considerations. Surgical reconstruction of the internal genitalia with restoration of normal menstruation and maintenance of a patent genital tract is challenging. Full excision and marsupialization of the vaginal septum are the preferred surgical treatment for uterine didelphys with obstructed unilateral vagina [9]. This can be challenging as hematometrocolpos causes distortion of the adjacent anatomic structures [9]. In our case, as the two hemi-uteri were hypoplastic Strassman's metroplasty along with resection of vaginal septum was performed. Strassman's metroplasty facilitated drainage of the right noncanalized hemiuterus and creation of a single uterine cavity with a bigger caliber. This was done aiming to provide better obstetric prospects in future as she had already suffered miscarriage of her first pregnancy.

The peculiar aspect of our case was that the patient had developed dysmenorrhea recently despite attaining menarche 7 years ago. This was contrary to the common presentation of dysmenorrhea and pelvic pain within the first few years of menarche. Another fascinating aspect was the growth of an ovarian cyst which coincided with the time of conception therefore initially it gave a picture of ongoing pregnancy, but later on investigation, it was a complete surprise to encounter HWWS.

Declaration of patient consent

The authors certify that the patient has obtained appropriate patient consent form. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initial will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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