Uterine Didelphys with Unilateral Cervicovaginal Agenesis: A Case Report

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Unilateral cervicovaginal agenesis in a didelphic uterus is an extremely rare congenital anomaly. Women born with this anomaly present with menarche associated with progressive dysmenorrhea and symptoms of worsening endometriosis. Presented here is a case of a 12 year old nulligravid who presented with progressive cyclic abdominal pain. Physical examination revealed a tender large right adnexal mass and presence of a vaginal canal and cervix tilted to the left. Sonography showed the presence of uterine didelphys, right hematosalpinx and hematocolpos. Preoperative impression was outflow tract obstruction probably due to Herlyn-Werner-Wunderlich syndrome. However, intraoperatively, it was discovered this was a case of uterine didelphys with outflow tract obstruction of the right hemiuterus due to unilateral cervicovaginal agenesis and ipsilateral renal agenesis.

Keywords: cervicovaginal agenesis, mullerian anomalies, uterine didelphys

Introduction

Embryologically, the uterus, bilateral fallopian tubes and the upper two thirds of the vagina are derived from the mullerian ducts. In organogenesis, the mullerian ducts form, fuse in the midline, and septal resorption completes the process to form a single uterus with a functional endometrium and intact outflow tract. Failure of organogenesis leads to unicornsate uterus or mullerian agenesis including cervical agenesis and cervicovaginal agenesis. Failure of the mullerian ducts to fully fuse leads to didelphic or bicornuate uterus. Presence of fusion but failure of septal resorption results in a septate uterus or arcuate uterus. The urinary and genital systems arise from a common ridge of mesoderm; therefore anomalies of the mullerian ducts may be associated with anomalies of the urinary system.

The incidence of mullerian anomalies ranges between 0.1% to 3.8% in the general population. Incidence of congenital obstructive malformations of the female reproductive system is low; the actual incidence is difficult to ascertain since these anomalies are usually undetected at birth, thus unreported and there is lack of uniform diagnostic modalities. Ovarian development is unrelated to mullerian ducts embryologically and thus unaffected in the presence of these anomalies.

Vaginal agenesis in a functional uterus has a reported incidence of 1 in 5000 female livebirths, while the incidence of cervical agenesis is even lower, occurring in 1 out of 80,000-100,000 livebirths. Combined agenesis of the cervix and the vagina in the presence of a functional uterus is an extremely rare mullerian anomaly. Half of patients with cervical anomalies will have concurrent vaginal agenesis and one third will have a uterine anomaly.

Uterine didelphys with unilateral obstruction is frequently associated with ipsilateral renal agenesis. This complex malformation is also known as OHVIRA (obstructed hemivagina ipsilateral renal agenesis). Some reports refer to this condition
as the Herlyn-Werner-Wunderlich (HWW) syndrome. Initially, Herlyn and Werner described the syndrome as the presence of uterus didelphys, unilateral renal aplasia, and ipsilateral blind hemivagina. Meanwhile, Wunderlich described the condition as the presence of simple vagina and unilateral hematocervix due to cervical atresia. The incidence of HWW occurs in 3-4% of women with Mullerian anomalies.5 There is no reported incidence of uterine didelphys with associated cervicovaginal agenesis. Search for literature citing the presence of obstructive uterine didelphys complicated by the presence of cervical or vaginal or cervicovaginal agenesis or atresia resulted to the following 1) two cases of uterine didelphys with unilateral cervicovaginal agenesis6, 2) one case of uterine didelphys with unilateral distal vaginal agenesis7, 3) one case of uterine didelphys with bilateral cervical agenesis and 4) two cases of uterine didelphys and unilateral cervical atresia.5,8 All cases, except the one with bilateral cervical agenesis, had reported ipsilateral renal agenesis.

The Case

This is a case of a 12 year old nulligravid who presented at the emergency room with the chief complaint of abdominal pain. Patient has had cyclic right lower quadrant pain for almost a year, which was described as crampy, non-radiating, and with an intensity of 5 out of 10. Patient experienced the pain during the first day of menstruation and was relieved by intake of paracetamol tablet.

Two months prior to admission, due to increasing intensity of the pain, the patient consulted the outpatient department and a transrectal ultrasound was requested. The transrectal ultrasound revealed the presence of uterine didelphys, dilated right fallopian tube suggestive of hematosalpinx, hematocolpos, normal ovaries and incidental finding of absent right kidney (Figures 2 & 3). The impression was Herlyn-Werner-Wunderlich Syndrome and she was advised surgery. However due to financial constraints, the patient refused to be admitted.
One day prior to admission (day 1 of her menses), the patient experienced severe hypogastric pain, crampy in character, non-radiating and 8/10 in pain intensity. There was no associated fever, vomiting or dysuria noted. The patient was subsequently admitted for further management.

Her past medical history was non-contributory. There was no family history of menstrual disorder or congenital malformations. The patient has one female sibling who had her menarche at 11 years old and has no gynecologic complaints. She had her menarche when she was 11 years old; she had regular menstrual cycle occurring every 28 days with duration of 2 days. She uses 2 fully soaked pads per day and she experienced dysmenorrhea usually on the first day of her menses with no other associated symptoms. The patient denies any sexual contact.

On physical examination, the patient had normal secondary sexual characteristics (Tanner staging 3 and 4 for breast and pubis). She was not pale and had pink palpebral conjunctiva. She had a soft flat abdomen with direct tenderness of the
hypogastric area and no rebound tenderness. Pelvic examination revealed normal appearing external genitalia and presence of what seemed to be a hymen. As the patient was a virgin, a bimanual rectal examination was done revealing the presence of a right lateral tender mass 3 cm from the anal verge measuring 5 cm in widest diameter that was thought to be the hematocolpos or trachelometra. Based on sonography and physical findings the initial working impression was Right hematosalpinx secondary to Herlyn-Werner-Wunderlich syndrome.

Different causes of outflow tract obstruction that included obstructive transverse vaginal septum, presence of cervical or vaginal or cervicovaginal agenesis or atresia were discussed with the patient and her guardian. Treatment options such as excision of transverse vaginal septum, uterovaginal anastomosis and hysterectomy and their complications were discussed as well. A decision for hysterectomy was undertaken by the patient and her guardian in case of the presence of cervical, vaginal or cervicovaginal atresia or agenesis. Informed consent was also obtained for possible discussion of this case in a scientific forum. Permission was given by the patient and her guardian for necessary appropriate pictures to be taken during the preoperative and intraoperative period that will aid in the discussion of the case.

At the operating room, internal examination under anesthesia revealed the presence of one vaginal canal directed more to the left with one cervix palpated. No bulging mass was noted. There was presence of a 12 cm movable cystic mass palpated at the right adnexal area. Trachelometra was doubted due to the absence of a firm bulging mass in the vagina and so a decision to proceed with laparotomy was undertaken.

Upon laparotomy, there was minimal hemoperitoneum noted. The left hemiuterus was small with attached left adnexa. The left adnexa was grossly normal with no noted adhesions. There were dense adhesions noted between the right fallopian tube and the right ovary and omentum. The right fallopian was dilated to 11.5cm x 6.5cm x 4.0cm containing approximately 100cc of chocolate like fluid on cut section. The right hemiuterus was slightly enlarged with no palpable cervix. No right vaginal tract was palpated abdominal or vaginally. The right hemiuterus measured 10.5cm x 5.5cm x 5.0cm containing approximately 50cc of hematometra. There was no visible cervical portion noted in the uterus (Figures 4 & 5). Right hemihysterectomy with right salpingectomy was performed.

The patient had an uneventful postoperative recovery and was allowed to go home after 4 days of confinement. Histopathology showed basal
endometrium; endometriosis, chronic salpingitis, hydrosalpinx and foci of old and new hemorrhage of the right fallopian tube.

Final diagnosis was Right hematosalpinx secondary to uterine didelphys with right cervicovaginal agenesis; ipsilateral renal agenesis.

Discussion

This is a case of a 12 year old nulligravid who presented with a chief complaint of cyclic abdominal pain temporarily relieved by intake of analgesic. She presented with normal development of secondary sexual characteristics. Bimanual rectal examination revealed a right adnexal mass. Sonography showed the presence of uterine didelphys, right hematosalpinx and hematocolpos. Preoperative impression was outflow tract obstruction probably due to Herlyn-Werner-Wunderlich syndrome. However, intraoperatively it was discovered this was a case of uterine didelphys with outflow tract obstruction of the right hemiuterus due to cervicovaginal agenesis and ipsilateral renal agenesis.

Congenital obstructive reproductive tract anomalies usually become clinically relevant in adolescence. They may present with primary amenorrhea and cyclic pelvic pain or progressive dysmenorrhea. The presence of menarche does not exclude obstructive mullerian anomalies because the obstruction may be unilateral and involves a hemiuterus of a uterine didelphys or rudimentary horn of a unicornuate uterus. The index patient presented with progressive dysmenorrhea due to an obstructed right hemiuterus of a uterine didelphys.

Presently, four systems have been proposed for the classification of female genital tract anomalies. The 1988 American Fertility Society (AFS) classification, the 2011 the embryological-clinical classification system of genitourinary malformations (Acien), the vagina, cervix, uterus, adnexae and associated malformations system based on the tumor nodes metastases (TNM) principle in oncology and the European Society of Human Reproduction and Embryology (ESHRE) and the European Society for Gynaecological Endoscopy (ESGE) classification of female genital tract anomalies. The AFS classification is the most popular and widely used system. In this system the patient falls under Class III category (Figure 1). However the AFS classification has serious limitations when it comes to further subdivisions for certain categories of anomalies such as in this case. In the ESHRE/ESGE classification, the main classes of anomalies have been divided into subcategories expressing anatomical deviations with clinical significance. The cervical and vaginal anomalies are further classified into subclasses that make it easier to classify mullerian anomalies with multiple uterine and cervicovaginal malformations. In this classification, the index patient is classified as Class U3b/C3 (Figure 6). Limitation in this classification is the absence of subcategory for unilateral vaginal agenesis or atresia.

Cervicovaginal agenesis is somehow difficult to diagnose. Thorough clinical history and physical examination with aid of imaging modalities are the keys to diagnosis. Physical examination during menstruation reveals the presence of an enlarged uterine corpus distal to the introitus indicating collection of menstrual blood in the uterus, cervix and/or vaginal canal. Adnexal mass and/or tenderness may be present as a consequence of pelvic endometriosis. Presence of a functioning endometrium and outflow obstruction caused by the absence of the cervix and vagina prevent menstrual debris from being discharged. Menstrual blood will collect inside the uterus and reflux along the fallopian tubes into the peritoneal cavity leading to pelvic endometriosis. Since most patients presenting with the clinical profile of obstructive mullerian anomalies are adolescents with no previous sexual contact, examination is limited to a bimanual rectal exam making it difficult to differentiate different causes of outflow tract obstruction. Differential diagnoses include imperforate hymen, transverse vaginal septum, cervical agenesis or atresia and vaginal agenesis. Imaging studies such as magnetic resonance imaging (MRI) and ultrasound are the most reliable modalities in diagnosing these cases. MRI is the gold standard in the diagnosis of mullerian anomalies. However, it is quite costly and not readily available and accessible. In experienced
Figure 6. Female genital tract malformation classification by the European Society of Human Reproduction and Embryology.

hands and with appropriate clinical correlation, sonography has a high accuracy in diagnosing different types of mullerian anomalies. Three-dimensional ultrasound has a higher accuracy rate over conventional two-dimensional scanning. Two-dimensional sonography was used in this case. Despite all the advanced diagnostic modalities, the true nature of outflow tract obstruction is often difficult to diagnose as in the case of our index patient.

Conservative management of cervicovaginal agenesis in a single uterus is important to preserve reproductive function of the patient. However, complications of conservative surgery for patients with cervicovaginal agenesis result to poor functional outcome and high failure rates. Complications include recurrent obstruction of the uterovaginal neocanal, pelvic infection and persistent infertility. Therefore in many cases, hysterectomy is still considered the first line
treatment in patients with cervicovaginal agenesis. Fortunately for this patient, the presence of a normal left hemiuterus and normal left adnexa has preserved her reproductive function despite right hemihysterectomy. Postoperative monitoring for possible progression of endometriosis is also essential. A follow-up pelvic ultrasound 6 months after the patient's surgery revealed no progression of the endometriosis.

Summary

In patients with obstructive mullerian anomalies, the goals of management include establishing an intact outflow tract or removal of the obstruction to relieve the symptom of pain and to preserve sexual and reproductive function if possible. Delay in establishing the diagnosis of unilateral obstructive anomalies of the reproductive tract increases the risk of pelvic endometriosis.

This is a case of a 12 year old gravida 0 with outflow tract obstruction of the right hemiuterus in a uterine didelphys complicated by right hematosalpinx. The cause of the obstruction was agenesis of the right cervix and vaginal canal. Early evaluation and appropriate surgical intervention has decreased the risk of future reproductive complications in this patient.

References