Uterus didelphys with imperforate hemivagina and ipsilateral renal agenesis complicated by hematocolpos, hematometra and hematosalphinex. The challenge of intact hymen
CASE REPORT

Uterus didelphys with imperforate hemivagina and ipsilateral renal agenesis complicated by hematocolpos, hematometra and hematosalpinex. The challenge of intact hymen


a Department of Obstetrics and Gynecology, Assiut University, Assiut, Egypt
b Department of Radiology, Faculty of medicine, Assiut University, Assiut, Egypt
c Department of Obstetrics and Gynecology Nursing, Faculty of Nursing, Assiut University, Assiut, Egypt

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Abstract Background: Uterine didelphys with obstructed hemivagina is an extremely rare condition. This diagnosis should be considered whenever an adolescent is presented with severe dysmenorrhea and progressive pelvic mass.

Case: A 15-year-old girl presented with uterus didelphys with obstructed left hemivagina and ipsilateral renal agenesis. Excision of the septum was done using monopolar diathermy in the cutting mode under laparoscopic and hysteroscopic guidance, taking into consideration the integrity of the hymen. Complete recovery was achieved within one week.

* Corresponding author. Address: Department of Obstetrics and Gynecology, Faculty of Medicine, Consultant, Women’s Health Centre, Assiut University, P.O. 71116, Assiut, Egypt. Tel.: +20 882 41 4616, +20 88920355; mobile: +20 127432270; fax: +20 882 368377.
E-mail address: drzahranmk@gmail.com (K.M. Zahran).
Conclusion: The prompt and accurate diagnosis of this condition is essential to prevent complications from acute illness and to preserve future fertility. Ultrasonography, CT scan and MRI are the initial steps for its diagnosis; however, laparoscopy is a crucial tool for confirmation of the diagnosis. In order to avoid the associated morbidity of delayed management, surgical intervention should be considered as the first line therapy, taking into consideration the social and traditional backgrounds of the parents and patients.

1. Introduction

Uterine didelphys are rare malformations involving the Mullerian ducts (1,2). During fetal development, the Mullerian ducts represent the primordial components of the female reproductive system. They differentiate into the fallopian tubes, uterus, cervix, and the superior aspect of the vagina (3).

In cases of didelphys with renal agenesis, it is likely that the lesion occurs from embryologic arrest at 8 weeks of gestation that simultaneously affects the adjacent Mullerian and metanephric ducts. In the general population, the true incidence of this anomaly is unknown, but has been reported between 0.1% and 3.8% (4). This wide range could be because of inaccurate diagnosis or the fact that many of these diagnoses are not detected during the women’s lifetime (5). For unknown reasons, anomalies of this type are more common in the Finnish population, with an incidence of 0.5% (6). Some may present at menarche with pelvic pain secondary to hematocolpos, while others present with pain, fever, and abscess formation (7,8). We present here a rare case presented by Uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis complicated by hematocolpos, hematometra, and hematosalpinx.

2. Case

A 15-year-old unmarried, average body built girl was referred to the women’s health hospital, Assiut University, Egypt, because of increasing monthly abdominal pain. She stated that her menarche occurred at age 13 with normal pubertal events, but her menses were irregular. During the few months prior to her referral, she began feeling severe lower abdominal pain particularly during her menstrual period. On examination, her vital signs were stable and her abdominal examination revealed a hardly felt, not tender, pelviabdominal mass extending up to the umbilicus. Her pelvic examination revealed normal looking external genitalia, the hymeneal orifice looked more or less normal, but there was a tense cystic bulge to the left side and above the hymeneal orifice. PV examination was not performed for the sake of integrity of the hymen. PR examination revealed an oblong shaped tense cystic swelling anterior to the rectum; its upper end cannot be reached. Transabdominal ultrasonic examination revealed absent left kidney. The vagina was seen as an oblong shaped cystic mass filled with turbid fluid posterior to the bladder communicated by narrow isthmus to another thick walled cystic mass (the uterus) and another thin walled cystic mass (the fallopian tube) Fig. 1.

The diagnosis of hematocolpos and hematometra was suspected, but, as the girl reported that she is regularly menstruating and there was normal looking hymeneal orifice, the diagnosis of septate uterus and vagina with obstructing hemivagina was highly suspected.

Computed tomography subsequently confirmed the urinary system having a solitary normal right kidney, with the absence of the left kidney. The distended left tube was seen as an oblong shaped structure filled with turbid fluid suggesting hematosalpinx (Fig. 2A). The distended left uterine horn was filled with turbid fluid collection suggesting hematometra and normal right uterine horn (Fig. 2B). The distended left hemivagina was also filled with fluid collection (Fig. 2C,D) suggesting hematocolpos. The diagnosis of Uterus didelphys, longitudinal vaginal septum with imperforate left hemivagina and ipsilateral renal agenesis complicated by hematocolpos, hematometra, and hematosalpinx was highly suspected. Renal function was normal and other patient’s laboratory parameters were within normal limits. A relief operation was subsequently arranged.

After proper counseling and obtaining written informed consent from the patient and her parents, the patient was prepared for the operation. The day before the operation, she was given enemas until her fluids returned clean and antibiotic prophylaxis (ampicillin, sulbactam 1.5 g) was given to this patient 1 h before the operation. Under general anesthesia, the patient was placed in a lithotomy Position. After proper sterilization, Laparoscopic exploration revealed 2 widely separated uterine horns. The right side was of reasonable size with normal looking right tube and ovary, the left uterine horn and left tube were markedly distended with the left ovary attached to it, (Fig. 3).
Through the hymeneal orifice, hysteroscope was introduced into the right side of the vagina up to the cervix confirming the presence of normal looking cervix. Aspiration of the left cystic vaginal mass revealed dark altered blood of hematocolpos. Excision of the septum was done using monopolar diathermy in the cutting mode, free flow of the old collected blood was allowed. Perfect hemostasis of the edges was done and repair of the small hymeneal tear was done by 2 interrupted 2/0 cat gut sutures.

Laparoscopic reevaluation revealed a dramatic reduction of the hematometra, hematosalpinx, and hematocolpos. Postoperatively, the patient remained afebrile and her recovery was uneventful. One week later, pelvic examination revealed a normal-looking vulva and hymeneal orifice. Ultrasonic evaluation revealed complete collapse of the hematocolpos, hematometra, and hematosalpinx.

Official medical report was given to the parents. The patient and her parents were very satisfied with the results of the operation.

3. Discussion

Uterus didelphys with obstructed hemivagina is a rare condition. This diagnosis should be considered whenever we deal with adolescents with severe dysmenorrhea and progressive pelvic mass. Prompt diagnosis and excision of the obstructed vaginal septum can relieve these symptoms completely and prevent further sequelae (9,10).

Diagnostic methods include hysterosalpingography, transvaginal ultrasound, computed tomography, magnetic resonance imaging, laparoscopy, and hysteroscopy. Hysteroscopy can detect intrauterine adhesions and communication between the duplicated endometrial cavities. Magnetic resonance imaging has 96–100% accuracy in classifying uterine anomalies, while...
transvaginal ultrasound has 85–92%, and hysterosalpingography has 6–55% (11). Unfortunately, many of these diagnostic tools cannot be utilized in that case for the sake of keeping the hymen intact.

Zurawin et al., 2004, reported that laparoscopy should now be considered the gold standard for the complete evaluation of congenital anomalies of the female reproductive tract (12). This is supported by a cohort study of 22 patients that compared MRI and laparoscopy as diagnostic tools for Mullerian anomalies. MRI picked up only 53% of the Mullerian anomalies that were found at the time of laparoscopy (13).

The treatment for these patients is resection of as much of the obstructing vaginal septum as possible. A large distended hematocolpos is easy to visualize and palpate and aids in resection. Whereas the traditional approach utilizes retractors, scalpel, and suture, there have been several reports of the vaginoscopic use of a resectoscope (14,15). Resection may be carried out with preservation of the hymen.

Social and traditional backgrounds should be taken into consideration during counseling the parents and patients. The family was counseled regarding the need for evaluation of the vagina. The possible need for laparoscopy and vaginal septum resection was also explained.

The main difficulty of the operation is how to keep the integrity of the hymen. In our society, hymeneal integrity is a sign of virginity which should be kept intact up to marriage. It is not acceptable at all to have the hymen injured or traumatized before marriage, which was the difficult part in counseling the patient and her parents. The family was reluctant to give consent at that time, only after we assured them that we will do our best to keep the hymen intact or repair it meticulously in case of injury, they accepted the operation.

4. Conclusion

Uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis is considered to be an extremely rare condition. The prompt and accurate diagnosis is essential to prevent complications from acute illness and to preserve future fertility. This diagnosis should be considered whenever an adolescent is presented with severe dysmenorrhea and progressive pelvic mass. Ultrasonography, CT scan, and MRI are the initial steps for its diagnosis; however, laparoscopy is a crucial tool for confirmation of the diagnosis. In order to avoid the associated morbidity of delayed management, surgical intervention should be considered as first line therapy, taking into consideration the social and traditional backgrounds of the parents and patients.

Conflicts of interest

The authors have no conflicts of interest concerning the work reported in this paper.

References