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Case Report

Uterus didelphys with obstructed hemivagina with chronic presentation: a case report

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ABSTRACT

This is a case report of a 24-year patient who had uterus didelphys with obstructed hemivagina. Clinical manifestations of this case were of progressive dysmenorrhea, abdominal pain after menarche, cyclic difficulty in urination and constipation, with the existing paravaginal tumor indicated this rare anomaly. The diagnosis was by clinical examination and ultrasonography. The patient underwent successful transvaginal septoplasty and drainage of the hematocolpos and hematometra, which is the appropriate mode of treatment in such cases.

Key words: Clinical picture, Obstructed hemivagina Renal agenesis, Uterus didelphys

INTRODUCTION

Uterus didelphys with obstructed hemivagina is the result of a lateral nonfusion of the mullerian ducts with asymmetric obstruction, and it is almost always associated with renal agenesis.[1] While the classic presentation includes ipsilateral renal agenesis, more recent series have described rare cases of patients with two kidneys.

Patients with hematocolpos typically present early due to dysmenorrhoea, and pelvic pain.[2] Unusual features of this case were: patient presented late with a large paravaginal mass coming out of vagina, especially during menstruation associated with urinary problems during menstruation, clinical findings supported by USG alone, were used for diagnosis, both the kidneys were present, treatment successfully done without laparoscopy.[3-5.]

CASE REPORT

A 24-year-old nulligravida presented to the outpatient clinic of our department with complaints of steadily worsening dysmenorrhea and lower abdominal pain. She attained menarche at 13 years of age and she had irregular menses since then. She used to bleed for 15-16 days with gap of 20 days in between. After 1-2 years of menarche she used to have dysmenorrhea and lower abdominal pain on the left side.

Thereafter she started feeling a swelling in left side of vagina, which increased in size successively with each menstruation and decreased somewhat after menses. The swelling gradually increased so much so that it used to come out of the introitus for the last 1-2 years. Occasionally she had constipation, especially after her

periods along with difficulty in urination when the vaginal swelling increased in size. Her general examination was normal. Height was 151cm and her weight at the time of examination was 43 kg. Her abdominal findings were not significant. Vaginal examination revealed a non-tender bulging mass in the left half of vagina extending downwards from the fornix upto the introitus. The cervix was made out with difficulty, high up and deviated to the right. There was no evidence of bleeding noted at the time of examination.

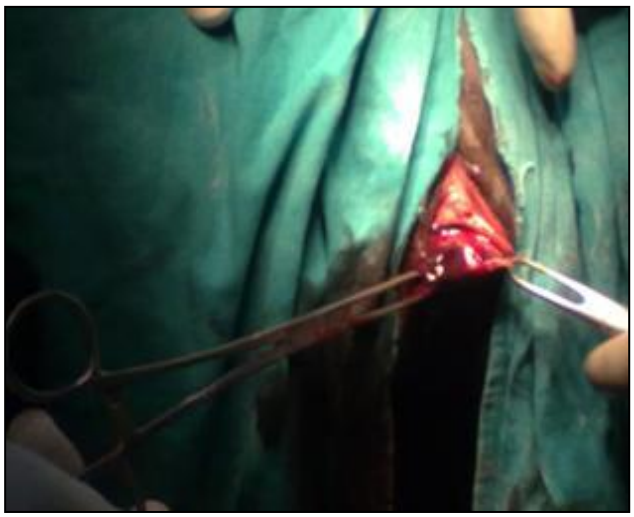

Figure 1: Drainage of left hematocolpos.

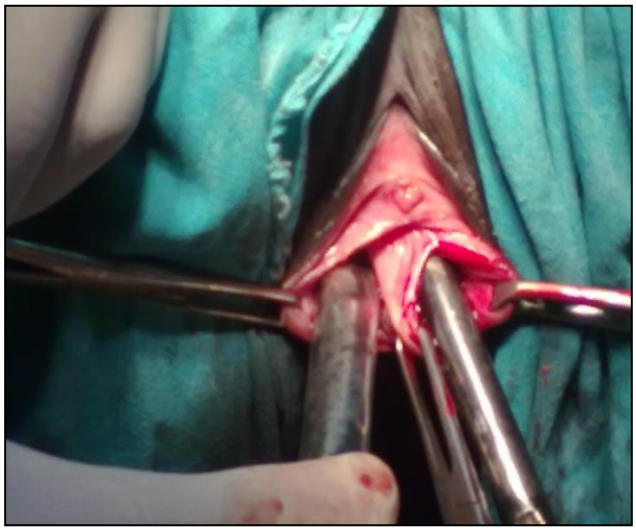

Figure 2: Longitudinal vaginal septum.

Abdominal ultrasonography revealed a bicornuate uterus with normal sized right uterus. Fluid collection was noted with internal echoes in the left uterine cavity. Left cornu measured 10.6x10.7x4.5 cm while the Right cornu measured 8.6x10.7 x 2.8cm. Endometrium (Right side) was thin and in central position, echotexture was normal. No focal lesion was seen. Cervix of uterus appeared normal. Both ovaries normal. POD was clear. Both kidneys were visualized and were normal. Impression: Hematometra in left cornu of bicornuate uterus.

Hematocolpos was missed on the USG and therefore the confusion in diagnosis. Magnetic resonance imaging was recommended, but could not be done due to monetary constraints. Routine investigations of blood were carried out and found to be normal in a work-up for EUA (examination under anaesthesia).

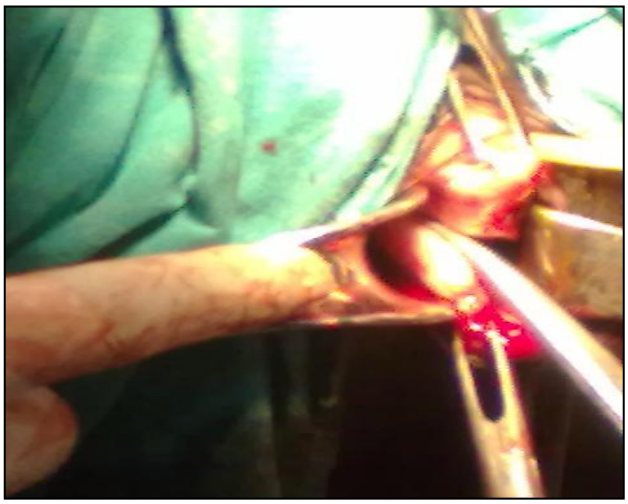

Figure 3: Transvaginal septal excision.

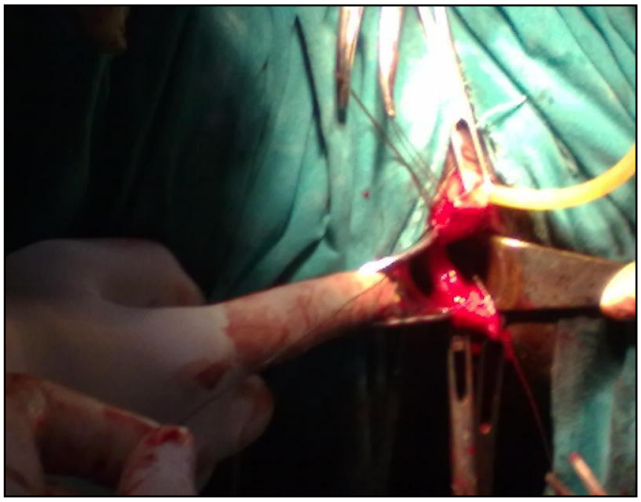

Figure 4: Suturing of edges of the vaginal septum

After appropriate counseling and consent, the patient was then taken to the operating room. Examination under anesthesia was undertaken and the left lateral vaginal mass was palpated. A Foley's catheter was placed in the bladder. A needle was placed into the mass, old collected blood came out and hematocolpos was confirmed. An incision was given on the mass in lower third of vagina and dense chocolate colored fluid escaped from it. Allis clamps were then placed along the cut edges of the incision and the hematocolpos was evacuated.

Two cervices were then palpated, right cervix could be visualized, left cervix (obstructed cervix) was palpated high up, flushed with vagina. The vaginal septum was excised as a oblong piece 6cmx3cm dimension until the two cervices. This revealed that the report of bicornuate uterus in USG was actually a case of uterine didelphys-bicollis-bicolpos.

The patient recovered without complications and postoperative speculum examination revealed healed vaginal mucosa and two cervices visible at the vaginal apex. The obstructed cervix appeared erythematous and flush with the vaginal fornix. An erythematous appearance is normal for the cervix after resection because maturation of squamous epithelium has not occurred.

DISCUSSION

Uterus didelphys with an imperforate hemivagina is an embryonic malformation of the genitourinary system in the female.[6] The lack of fusion of the mullerian ducts or the absence of resorption of the septum, results in varied genital anomalies: vaginal atresia, uterus didelphys, unicornuate or bicornuate uterus.[7] The displaced mullerian duct forms the imperforate hemivagina. Menstrual backflow may occur leading to unilateral hematocolpos, hematometra and endometriosis. The ipsilateral kidney and collecting ducts fail to develop.[8]

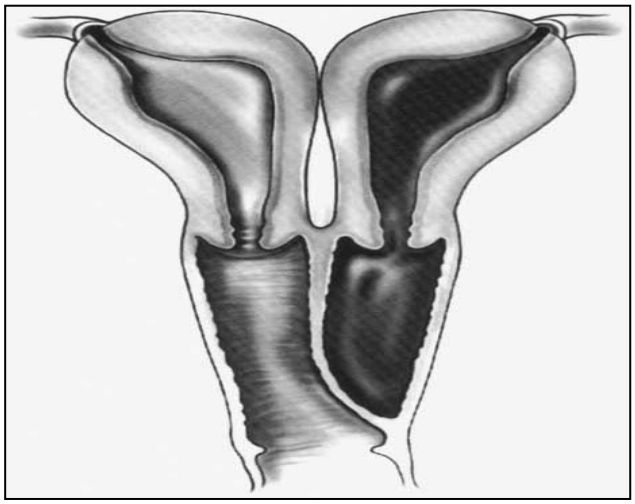

Figure 5: Complete obstructive longitudinal vaginal septum.

Uterovaginal duplication with obstructed hemivagina accompanied by ipsilateral renal agenesis is a rare entity that is referred to as Herlyn-Werner-Wunderlich syndrome, Patients with rare obstructed Mullerian anomalies such as Herlyn-Werner-Wunderlich syndrome present with nonspecific signs and symptoms.

The acronym OHVIRA (obstructed hemivagina and ipsilateral renal anomaly) was suggested in 2007 by Smith and Laufer to provide a common nomenclature for the syndrome. There has been a gradual increase in case reports of OHVIRA syndrome in the medical literature in recent years, likely reflecting a greater awareness of the condition among physicians.

Patients with this syndrome menstruate normally, so this syndrome as a diagnosis can easily be overlooked, as is obvious from our case presenting at 24 years' age. The diagnosis typically is made on the basis of history and examination that lead to increased pretest suspicion, followed by imaging. The imaging evaluation usually begins with ultrasonography, which is a noninvasive easily performed method for evaluation of the female genital tract. Magnetic resonance imaging is currently regarded as the the gold standard for delineating the anatomy.

Cases of obstructive uterine didelphys are particularly difficult to diagnose on USG owing to anatomical distortion secondary to mass effect from hematocolpos. In cases of obstructive uterine didelphys, it is most important to identify the pelvic mass as a hematocolpos. Real-time high resolution ultrasonography usually can differentiate among different blood filled portions of the occluded genital tract.[9] USG may determine that a uterine anomaly exists but often cannot differentiate between anomalies reliably as happened in this case where the report was of bicornuate uterus. Given the association between uterine and renal abnormalities, a brief evaluation of the renal fossae should be performed.

One clearly needs to be as definitive as possible in making the correct diagnosis before any surgical intervention is initiated. Accurate preoperative evaluation of the female genital tract using noninvasive technique with the awareness of the association of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis ensures appropriate conservative surgical intervention and decreases long-term morbidity.

Excision of the vaginal septum is recommended by most investigators. Traditional excision of the septum requires hymen disruption and wide exposure of the vagina using vaginal valves and often there are technical difficulties because of the small size of the vagina and thus of the surgical field.[10]

In present case, the septum protruded significantly into the vagina, as in the presentation of a complete transverse vaginal septum. Furthermore, the septum was quite thick in our case, making the excision challenging. Complete removal of the septum is the ultimate goal because this will help to reduce dyspareunia and intrapartum complications.

Good knowledge of normal embryology and sexual differentiation is important for timely diagnosis and appropriate management. This type of anomaly is diagnosed most frequently in adolescence but in present case it was diagnosed 10 years after menarche, because

these anomalies are uncommon, many practicing obstetrician-gynecologists may have little or no experience in diagnosing and treating these conditions. Furthermore, pelvic examination in the adolescent may be possible only under anesthesia, especially in early adolescence

Congenital developmental anomalies of Müller's canals are associated with menstrual difficulties; they are significant etiological factor of infertility and increase the risk of obstetrical complications. Long-term follow-up of reproductive characteristics of 38 patients with this type of developmental anomaly of Müller's canals within Haddad's study8 reported 20 pregnancies in 9 patients were reported. In four patients, pregnancy developed in ipsilateral hemiuterus on the side of atretic vagina following the resection of vaginal septum. These results suggest that transvaginal excision of vaginal septum is the appropriate therapeutical procedure. It should be emphasized that early diagnosis and surgical treatment are the best prevention of complications that may significantly diminish already reduced reproductive capacity, and sometimes even cause severe psychological problems.

The inclusion of a laparoscopy in the surgical treatment plan is still controversial. The natural history of endometriosis in the setting of obstructive lesions is not well-understood, and while two 11 authors documented spontaneous resolution of implants, none reported interval laparoscopy to confirm persistence. Therefore, until further evidence becomes available regarding the persistence of endometriosis with obstructive anomalies, routine laparoscopy with the goal of treatment of endometriosis may not be necessary.

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