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CASE REPORT

Double Uterus with Unilateral Obstructed Hemivagina and Ipsilateral Renal Agenesis

Vorapong Phupong MD,
Damrong Tresukosol MD,
Surasak Taneepanichskul MD,
Charintip Somprasit MD,
Srinaree Krewruedee MD,
Pramuan Virutamasen MD.

Department of Obstetrics & Gynecology, Faculty of Medicine, Chulalongkorn University, Rama IV Road, Pathumwan, Bangkok 10330, Thailand

ABSTRACT

A double uterus with a unilateral obstructed hemivagina is a rare condition, usually associated with ipsilateral renal agenesis. We report a case presenting with abdominal pain and pelvic mass. Hemihysterectomy was performed leaving the contralateral uterus intact. The postoperative course was uneventful and the patient did well at the six-week follow-up. An accurate diagnosis, appropriate management and the prevention of future fertility problems were discussed.

Key words : Double uterus, hemivagina, renal agenesis

The unique clinical syndrome consisting of a double uterus, obstruction of the vagina, and ipsilateral renal aplasia is a rare condition.[1-6] The clinical presentation varies greatly depending on whether the unilateral hemivagina is completely obstructed and on the location of its opening.[2] The most common clinical presentation is dysmenorrhea in association with the finding of a pelvic mass which resulted from a hemiobstructed vagina. Other presentations are pelvic pain, hypermenorrhea, menometrorhagia, intermittent vaginal spotting, malodorous vaginal discharge and urinary symptoms.[2-6] Since the clinical presentation varies, arriving at a diagnosis is difficult.[1] Early and accurate diagnosis and appropriate management will prevent future fertility problems.

When a post menarcheal young girl is found to have a pelvic mass, an ovarian germ cell tumor should first be listed in the differential diagnosis. Despite its rare condition, mullerian duct anomalies should be enlisted as well. One of the easiest clues is to prove that there is ipsilateral renal agenesis which is commonly associated with urinary and genital tract development.

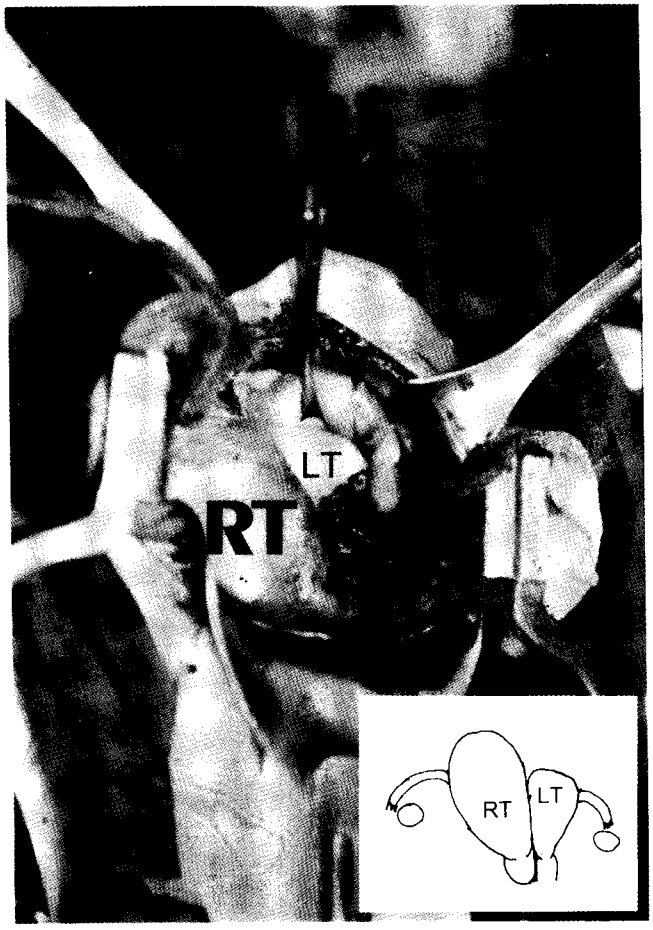
In this study, we report on a puberty patient who presented with lower abdominal pain, a pelvic mass and normal menstruation. Preoperative diagnosis was an ovarian tumor. We discuss a pitfall of diagnosis and management in the case.

Case Report

A 13-year-old girl was admitted to King Chulalongkorn Memorial Hospital because of right lower quadrant abdominal pain one month prior to admission. Her first menstruation period had been 24 months previously and had become regular since then. Her last menstrual period was April 17, 1999. She has never had dysmenorrhea. Her past medical and family history were unremarkable.

Physical examination revealed a cystic mass, sized 8x10 cm, extending from the pubic symphysis to the right paraumbilicus and mild tenderness on pressure. Pelvic examination disclosed an intact hymen and normal external genitalia. On rectal examination, there was mild tenderness near the small sized uterus and a cystic pelvic mass located on the right anterior of the uterus. Preoperative intravenous pyelography (IVP) and transabdominal ultrasonography demonstrated the absence of the right kidney, a dumbell like mass and mixed echogenicity. Preoperative diagnosis was right ovarian tumor.

At laparotomy, there was a double uterus; the left side was normal in size, the right enlarged to about 6x10x5 cm^3 (Fig. 1). Both uteri had only one adnexa each. There was a moderate degree of red spots of endometriosis-like appearance on the right fallopian tube and ovarian surface. Perituboovarian adhesion was also observed. Aspiration from the right uterus was made. A dark brown-coloured fluid, which looked like old menstrual blood, was thought to be hematometra. A vertical incision of the affected uterus was performed and a blind pouch vagina disclosed. A right hemihysterectomy was perfomed leaving the vaginal pouch opened. The postoperative course was uneventful and the patient left the hospital on the 5th postoperative day. She had recovered well at the six-week follow-up.



Legends for illustrations

Fig. 1. Intraoperative finding showing double uterus with right hematometrocolpos (RT); normal left uterus (LT).

Discussion

A double uterus with an obstructed hemivagina and ipsilateral renal agenesis is a relatively rare clinical episode.[1-6] Renal agenesis on the side of the obstructed vagina associated with a double uterus and cervix is suggestive of an embryologic arrest occurring during the 8th week of gestation that simultaneously affected the Mullerian and metanephric duct.[1] The exact cause of this developmental defect is unknown.[1] If one of the Wolffian ducts is absent, the kidney and ureter on the same side will fail to develop. The Mullerian ducts may also fail to fuse in the midline, either completely or incompletely. If the failure to fuse is complete, a uterus didelphys is formed. The

Mullerian duct on the side where the Wolffian duct is absent displaces itself laterally and cannot come into contact with the urogenital sinus in the center. Thus, the contralateral Mullerian duct gives way to the vagina, while the already displaced component on the other side forms a blind sac, i.e, an imperforate or obstructed vagina. The distal part of the vagina, which derives from the urogenital sinus, being distal from the hymenal ring, is not affected.[4] The more common defect was on the right side[3,7,8] as in our case.

The common presentation is pelvic pain and worsening dysmenorrhea since menarche is associated with the finding of a pelvic mass.[2,4-6,9,10] Most of the cases have a normal and regular menstruation period.[2,4] Occasionally, patients may present with symptoms from urinary tract infection.[7] Acute urinary retention as the initial symptom has also been reported.[11] Rarely, necrosis and rupture of the hematosalpinx resulting in a typical peritonitis has been reported.[9] Our case, in contrast with previous reports,[2,4-6,9,10] presented with a symptom of right lower quadrant abdominal pain and a finding of right cystic pelvic mass. She has had normal menstruation and no dysmenorrhea since the menarche.

The diagnostic tool valuable to confirm the diagnosis of this syndrome is ultrasonography.[2] It is both noninvasive and useful in the evaluation of renal agenesis.[2] There have been reports about magnetic resonance imaging (MRI) also being useful for diagnosing this syndrome.[12,13] Another investigative method is IVP which not only confirms the absence of a normal kidney on the affected side but also detects abnormalities of both the contralateral kidney and the ureter.[5] In our case, both transabdominal ultrasonography and IVP were not helpful but leaded to presumed ovarian neoplasm. This may be due to the lack of experience in this rare syndrome.

The management of choice recommended by most authors is excision of the vaginal septum.[2,4-6,8-10,14] However, hemihysterectomy is recommended in patients with a high, thick-walled obstruction, massive ovarian involvement, endometriosis and adenomyosis.[9] Pyocolpos due to closure of the septal incision and dysmenorrhea due to cervical stenosis have been reported.[9] Hence, careful follow-up of conservatively managed cases is essential. We managed our case contary to previous recommendations[2,4-8,13,14] involving hemihysterectomy and with the left blind pouch of the vagina opened. The reasons for this management were the thick-walled obstruction, as well as the educational and economic background of our case in that she might lose follow-up when managed conservatively. The advantages of our management are: decreasing long term complications after conservative management such as pyocolpos due to closure of the septal incision, dysmenorrhea due to cervical stenosis and recurrent hematometrocolpos. There were reports about endometrial carcinoma occurring in cases of a double uterus.[15,16] Correct diagnosis in these situations may be difficult due to anatomic difficulties or failure to recognize a second uterine cavity. If we leave both uteri, we must be concerned about the risk of endometrial carcinoma in the future. About future fertility, it is usually consistent with that of cases with a double uterus unless delay in diagnosis and resection of the obstructing septum has resulted in destroyed tubal function or the development of endometriosis.[1] The pregnancy rate, ranging from 40% to 87% has been reported.[5,6]

In conclusion, when ipsilateral renal agenesis is found in an adolescent girl presenting with abdominal pain and a pelvic mass, the syndrome of double uterus, obstructed hemivagina and ipsilateral renal agenesis should be considered. This should allow early diagnosis, appropriate management and decreased morbidity.

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