Uterus Didelphys with Unilateral Obstructed Hemivagina and Renal Agenesis on the Same Side : A Case Report

Pongsuk Pinchun MD,
Somkiat Prapasirisulee MD.

Department of Obstetrics and Gynaecology,
Chonburi Hospital, Chonburi,
Thailand

Abstract : *A rare condition, complete or incomplete duplication of the uterus and cervix with unilateral vaginal obstruction, is usually associated with ipsilateral renal agenesis. Clinical presentations are various such as pelvic pain, dysmenorrhea, abnormal vaginal bleeding or vaginal discharge. This patient complained of recurrent foul smelling vaginal discharge for many years, that was initially treated with many antibiotics and failed to recognize the true diagnosis. The menstrual and fertility functions were normal. The case, diagnosed and treated at Chonburi Hospital, is presented for awareness of this syndrome. (Thai J Obstet Gynaecol 1993; 5: 103-106.)*

Key words : uterus didelphys, renal agenesis, vaginal discharge

The specific association of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis was recognized as early as 1922[1]. In a minority of cases, a communication connecting the right and left sides at the level of the vagina was present[2]. The presenting symptom of this patient was recurrent vaginal discharge which did not respond to medical treatment.

Case Report

Mrs BR, a 25-year-old multiparous Thai woman presented to the Gynaecologic Out-patient Division with the complaint of foul smelling purulent vaginal discharge off and on which did not respond to many courses of various antibiotics for 2-3 years. Other symptoms such as dysmenorrhea, abdominal pain, fever or urinary symptoms were not found in this patient. Her first menstruation began at 15 years old, and the menstrual cycle was normal. She had 2 children delivered by normal vaginal delivery. Her past and family histories were negative.

Systemic physical examinations were normal. Pelvic examination re-

vealed normal external genitalia. During a speculum examination, a slightly protruding left vaginal fornix and foul smelling purulent discharge drainage from a small pinpoint area just anterolateral to the cervix was apparent. The uterus and both adnexae were normal.

Wet smear, Gram-stain revealed numerous white blood cells and mixed organisms, neither Trichomonas vaginalis nor fungus was found. On the endovaginal ultrasonogram, small hypoechoic density cystic space measuring 1.8 x 2.2 cm was found at the left lateral vaginal fornix. The uterus and adnexae were normal. No abnormal pelvic mass could be demonstrated.

Intravenous pyelography revealed an absent left urinary system and compensatory hypertrophic change of right kidney. No vesico-vaginal or uretero-vaginal fistulas could be demonstrated (Fig. 1).

Barium enema was performed revealed normal large bowels and no demonstrable recto-vaginal fistula.

The patient underwent hysterectomy because of persistent markedly foul smelling vaginal discharge and her family was complete. At laparotomy, no left kidney and ureter were present, confirming the findings of previous examinations. There were double uteri, the right side was normal in size but the left one was rather small. The fallopian tubes and ovaries were normal. The small left hemivagina was connected to the left small uterus and filled with purulent discharge. Transabdominal hysterectomy was done with no immediate or late complications.

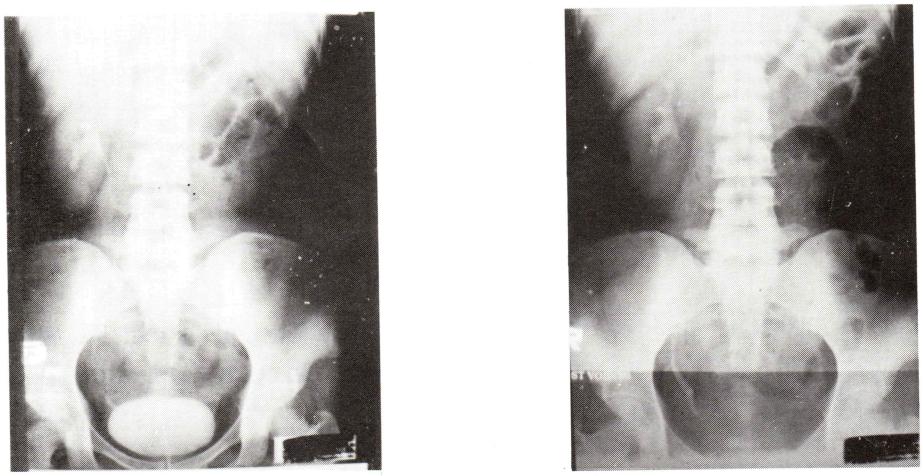

Fig. 1 IVP revealed renal agenesis of left kidney and ureter.

Discussion

At the sixth week of development, both male and female embryos have two pairs of genital ducts[3]. The first pair are mesonephric ducts which run along the lateral side of the mesonephros to the cloaca. The second pair are paramesonephric ducts which run first lateral to the mesonephric duct, but then cross it ventrally to grow caudomedial in direction. In the female embryo, the paramesonephric duct comes to full development and develops into the main genital duct of the female. Its cephalad part develops into the Fallopian tube. Its caudal part fuses with one on the opposite side by the end of the 7th week and finally differentiates into the uterus and upper vagina[3,4]. The growing paramesonephric duct is completely dependent on the mesonephric duct[4]. The ureteric bud is formed by an outgrowth of the mesonephric duct near the opening into the cloaca and penetrates the metanephric blastema which finally develops into metanephros or permanent kidney[3,5]. The failure to form the ureteric bud or make contact of the bud with metanephric blastema may result in agenesis of the kidney on that side[6].

So it is possible that abnormality in the development of the caudal portion of one mesonephric duct may result in failure of ipsilateral kidney development and also involvement of the ipsilateral paramesonephric duct, like in this patient. The incidence of unilateral renal agenesis with paramesonephric duct anomalies was reported as 1/2300 autopsies[7].

The abnormal laterally displaced paramesonephric duct cannot come into contact with the urogenital sinus in the center to form a normal vagina and only a blind sac, i.e. an imperforated or obstructed hemivagina is formed. In this case, we found uterus bicornis bicollis with unilateral hemivagina and the little communication between the two sides of vagina at the level of the vagina. The communication is rather small so it can cause obstruction of the discharge in the blind pouch vagina and become secondarily infected. The communication may be acquired in nature[5].

The presenting symptoms depend on the site of communication such as : progressive dysmenorrhea, foul smelling vaginal discharge, abdominal pain and urinary symptoms. In this case there is only the symptom of persistent vaginal discharge. This symptom and sign may suggest an abnormally developed paramesonephric duct with a communication[5]. No dysmenorrhea can be explained by nonfunctioning endometrium of the affected side.

The anomalies that had unilateral hemivagina were more difficult to be diagnosed. This case had a small cystic mass at the vaginal wall requiring differentiation from more common lesions of the lateral vaginal wall, Gartner's duct cyst. Purulent discharge from the small opening at the vagina or fornix require differentiation from recto-vaginal fistula or

other infection with fistula to vagina.

In cases of unilateral hemivagina, the ultimate goal of treatment is adequate excision of vaginal septum to create a common vagina. The septum should be totally removed or made as wide as possible in one procedure. But in cases of superimposed infection, simple excision of the bulging vaginal septum for drainage the pus must be done initially, and should be followed by complete excision of the septum later on. This procedure is important to facilitate the examination and treatment later on because the patient may be pregnant in the defective side of the uterus. If abortion or term pregnancy does occur, it is difficult to do curretage or vaginal delivery through a small opening or an inadequately excised vaginal septum.

In patients with a single kidney, there is a higher risk than normal. During pregnancy there is a higher incidence of urinary tract infection, this will increase the maternal mortality and the fetal wastage. The remaining kidney is nearly always affected with some chronic renal disease[8].

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