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Case Report

Uterine Didelphys with Hematometra: Case Report

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ABSTRACT

Uterine didelphys represents a uterine malformation where the uterus is present as a paired organ when the embryogenetic fusion of the mullerian ducts fails to occur. As a result there is a double uterus with two separate cervices, and often a double vagina as well. The condition is less common than these other uterine malformations: arcuate uterus, septate uterus, and bicornuate uterus. The complete form is characterized by two hemiuteri, two endocervical canals with cervices fused at the lower uterine segment. Each hemiuteri is associated with one fallopian tube. Ovarian malposition may also be present. The vagina may be single or double, with duplication a frequent component.

Key words: Didelphys uterus, Dysmenorrhoea, Hematometra, Mullerian ducts, Renal agenesis, Ultrasonography.

INTRODUCTION

The uterus is formed during embryogenesis by the fusion of the two mullerian ducts. This process usually fuses the two mullerian ducts into a single uterine body but fails to take place in these affected women who maintain their double mullerian systems. A didelphic uterus will have a double cervix and is usually associated with a double vagina. The cause of the fusion failure is not known. Associated defects may affect the vagina, the renal system and, less commonly, the skeleton. Renal agenesis most commonly occurs in association with uterine didelphys than with any other type of mullerian anomaly. [1] Obstructed unilateral vagina in patients with uterus didelphys is frequently associated with ipsilateral renal and ureter agenesis; this is known as Wunderlich-Herlyn-Werner syndrome (HWW), a rare but well-established anomaly. [2,3] Piccinini et al reported case of Herlyn-Werner-Wunderlich syndrome a rare congenital disorder in which uterus didelphys is associated with obstructed hemivagina and renal agenesis. [4] Renal agenesis is thought to be due to developmental arrest in one wolffian duct that in turn affects induction of nephrogenesis and positioning of the ipsilateral mullerian ducts. The malpositioned mullerian duct is incapable of successful fusion, and two hemiuteri develop. [1]

CASE REPORT

20 years female presented with abdominal pain and dysmenorrhoea. History revealed nothing significant. On presentation, the patient was hemodynamically normal and a febrile. She was advised 3D/4D color doppler ultrasonography of lower abdomen. On ultrasonography there were two uteri and two cervices. Right uterus was normal in

size and echo texture and cervix was normal. Left uterus and cervix were distended with fluid / blood with cervical stenosis. Left ovary was normal in size and echo texture and showed normal small follicles. Right ovary was normal in size and echo texture and showed normal small follicles. No fluid was seen in Pouch of Douglas.

DISCUSSION

Mullerian duct abnormalities cover a wide range of developmental anomalies, resulting from non-development, defective fusion, or defects in regression of the septum during fetal development. Didelphys uterus arises when midline fusion of the mullerian ducts is arrested, either completely or incompletely. Approximately 11% of uterine malformations are didelphys uterus. [5] Approximately 75% of patients with didelphys uterus have a complete or partial vaginal septum, [6] which is most commonly longitudinal in HWW and is thought to reflect a disorder of lateral fusion between the inferior portions of the two mullerian ducts. [7] Studies of uterine organogenesis have implicated the Hox and a Wnt gene as regulators of uterine morphology didelphys uterus is reported to occur in association with other anomalies. [8] The association of uterus didelphys with an obstructed hemivagina is explained by a defect in embryogenesis at the eighth week of gestation affecting the mesonephric and mullerian ducts, in which there is non-fusion of mullerian ducts or failed resorption of the uterine/vaginal septum. Patients with a uterine didelphys are usually asymptomatic, unless an obstruction is present. In such cases, hematometrocolpos, hematometra, and hematosalpinx may develop. Women with the condition may be asymptomatic and unaware of having a double uterus. However, a study by Heinonen showed that certain conditions are more common. In his study of 26 women with a double uterus gynecological complaints included dysmenorrhea and dyspareunia. All patients displayed a double vagina. [9]

CONCLUSION

Uterus didephys reported here emphasize the following fact that early recognition is important in order to avoid complications such as retrograde tubal reflux and consequent endometriosis, as well as to preserve fertility. [13,14]

Associated anomalies

Some coexisting anomalies include the following: bladder exstrophy with or without vaginal hypoplasia; congenital vesicovaginal fistula with hypoplastic kidney; and cervical agenesis. Other defects which can be found include renal dysplasia, duplication of the kidneys and ureters, [10,11] ectopic ureter, as well as high-riding aortic bifurcation, IVC duplication, intestinal malrotation [12] and ovarian malposition.

In the present case report, uterus didelphys was present with hematometra. There were no other associated anomalies.

Diagnosis

Nonobstructive uterus didelphys is usually asymptomatic until menarche. The diagnosis is often rendered during the initial pelvic examination, when two cervices are identified. In hemivaginal obstruction, the clinical presentations are variable and depend on the degree of obstruction and whether the obstruction has an opening. The most common presenting symptoms are onset of dysmenorrhea within the first years following menarche and progressive pelvic pain. A unilateral pelvic mass is detected on examination with the right affected nearly twice as frequently as the left.

Techniques to investigate the uterine structure are transvaginal ultrasonography, hysterography, hysterosalpingography, MRI, and hysteroscopy. More recently 3-D ultrasonography has been advocated as an excellent non-invasive method to evaluate uterine malformations.

Prognosis

Prognosis is good, with the major concern being preservation of fertility. Women with uterus didelphys have a high

likelihood of becoming pregnant [14,15,16,9] with approximately 80% of patients able to conceive [17,18] but with elevated rates of premature delivery (22%) and abortion (74%); cesarean section is necessary in over 80% of patients. [18,19]

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