Syndrome of Herlyn-Werner-Wunderlich: a case - report

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Introduction

Uterine malformations occur in 2 to 4 % of fertile women with normal reproductive outcome and in 13 to 25 % among women with recurrent pregnancy loss.

One of these rare malformations is the Herlyn-Werner-Wunderlich syndrome, which represents a complex female genital malformation with uterus didelphys, unilateral low vaginal obstruction, and ipsilateral renal agenesis.

Case - report

Our patient 11 years old had her menarche 6 months before her first presentation to emergency room for acute abdominal and pelvic pains of two weeks duration. A physical examination indicated a sensitive and painful abdomen. Further evaluation by ultrasonography showed an abdominal mass of 6-8 cm, both ovaries were normal in shape and size. A CT scan was performed in order to evaluate the possible genito-urinary anomaly and demonstrated the presence of the triad of uterine malformation (didelphys uterus), right obstructed hemivagina, and ipsilateral renal agenesis.

The patient underwent a laparoscopy which confirmed the diagnosis of uterus didelphys. Marsupialisation of the blind right hemivagina and hysteroscopy were performed at the same time. Our patient quickly improved after the surgery.

Discussion

HWW is a type of Mullerian duct anomalies (MDA) resulting from disruption of mullerian duct development during the embryogenesis. Three principal mechanisms can cause these malformations:

- non development (agenesia or hypoplasia) of mullerian duct;
- defect of lateral or vertical fusion of mullerian duct;
- failure in resorption of mullerian duct;

MDA are commonly associated with renal anomalies in 30 to 50 percent (agenesis, ectopia, hypoplasia, malrotation, duplication?). However, there is no association with ovarian anomalies.

Magnetic resonance (with oblique coronal T2-weighted images of uterus) and 3-D ultrasonography (with rebuilt coronal views of uterus) are both non invasive and non irradiant exams that can evaluate MDA. These imaging methods provide important diagnosis features such as:

- description of the external uterine contour (a fundal cleft greater than 1 centimeter is the key feature to differentiate bicornuate-didelphys uterus to septate uterus);
- evaluation of internal uterine cavity and zonal anatomy;
- measure of the intercornual angle ( more than 60-100 degrees in bicornuate-didelphys uterus and less in septate uterus);
- measure of intercornual distance (4 cm or more in didelphys-bicornuate uterus, and less than 4 cm in septate uterus);

Clinically, this usually occurs to postpubertal adolescent or adult women when hematometrocolpos produces a more pronounced mass effect and pain on the side of the obstructed hemivagina.

Conclusions

The diagnosis of HWW can be difficult to elaborate due to the absence of specific findings through physical examination and to non specific symptoms. It is desirable to achieve an early diagnosis and treatment of HWW syndrome, in order to relieve acute symptoms, preserve fertility and prevent both acute complications as acute
pyo-hematocolpos, pyosalpinx, and pelviperitonitis and long term complications as endometriosis, pelvic adhesions, obstetric problem (recurrent pregnancy loss, preterm delivery, malpresentation?) and infertility.