RESEARCH ARTICLE

PSEUDO UNICORNUATE UTERUS: CLINICAL CASE AND LITERATURE REVIEW

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Abstract

The pseudo unicornuate uterus is a rare uterine malformation resulting from incomplete unilateral Müllerian aplasia and is estimated to occur in about 10-14% of all uterine anomalies. It is the consequence of a developmental arrest of one of Muller's ducts, which results in a normal hemi-uterus and a rudimentary horn with or without a cavity.

We present a case that illustrates this pathology: This is Mrs. S. H, 30 years old, without any notable history, G2P2: G1: First pregnancy, followed at the health center, with normal evolution, with delivery by cesarian section at term for breech presentation in a primiparous woman. The patient did not report any anomalies about the first childbirth. G2: Second pregnancy, followed up until 39 weeks of amenorrhea at the health center, admitted in early labor, obstetrical ultrasound revealed a single fetal pregnancy with breech presentation, the indication for extraction by the high route was indicated for breech presentation in a scarred uterus. On exploration we noted the presence of a right hemi-uterus in which the pregnancy had developed with a homolateral horn and adnexa, and a small rudimentary remnant on the left continuing with a tube.

Introduction:-

Pseudo unicornuate uterus is a rare uterine malformation, resulting from incomplete unilateral Müllerian aplasia and estimated to occur in about 10 to 14% of all uterine anomalies. It is the consequence of a developmental arrest of one of the Müllerian ducts, which results in a normal hemi-uterus and a rudimentary horn with or without a cavity.

Observation:-

Mrs. S. H., 30 years old, with no notable history, G2P2:
G1: First pregnancy, followed at the health center, with normal evolution, with delivery by cesarian section at term for breech presentation in a primiparous woman. The patient did not report any anomalies about the first childbirth.
G2: Second pregnancy, followed up at 39 weeks of amenorrhea, admitted in early labor, obstetrical ultrasound revealed a breech pregnancy with a single fetus, the indication for emergency high extraction was indicated for breech presentation in a scarred uterus.

On exploration we noted the presence of a right hemi-uterus in which the pregnancy had developed, with a homolateral horn and adnexa, giving the a first impression of a unicornuate uterus.
After exploring the contralateral:
On the left side, there was a small uterine remnant corresponding to a rudimentary horn with no cavity and not communicating with the other part, which continued with a tube and an ovary of normal appearance. Absence of pelvic ectopic kidney.

The whole illustrated by the following images:

**Figure 1:** First look: presence of a single right annex: unicorne uterus?

**Figure 2 and 3:** Visualisation of the left ovary and fallopian tube linked to a rudimentary horn.
Classification
The malformation in our patient's case is classified as U4C0V0 according to the ESHRE, corresponding to a pseudo-unicornuate uterus with a rudimentary horn, probably non-functional; an MRI was requested to confirm or deny the presence of a cavity.

| Uterine anomaly | Main class | Sub-class | Cervical/vaginal anomaly | Co-existent class |
|----------------|------------|-----------|--------------------------|------------------|
| U0 Normal uterus |            |           | C0 Normal cervix           |                  |
| U1 Dysmorphic uterus | a. T-shaped | b. Infantilis | C1 Septate cervix |                  |
| | c. Others |           | C2 Double 'normal' cervix |                  |
| U2 Septate uterus | a. Partial | b. Complete | C3 Unilateral cervical aplasia |                  |
| U3 Bicorporeal uterus | a. Partial | b. Complete | C4 Cervical aplasia |                  |
| | c. Bicorporeal septate |           |                  |                  |
| U4 Hemi-uterus | a. With rudimentary cavity (communicating or not horn) | b. Without rudimentary cavity (horn without cavity/no horn) | V0 Normal vagina |                  |
| U5 Aplastic | a. With rudimentary cavity (bi- or unilateral horn) | b. Without rudimentary cavity (bi- or unilateral uterine remnants/aplasia) | V1 Longitudinal non-obstructing vaginal septum |                  |
| U6 Unclassified malformations | | | V2 Longitudinal obstructing vaginal septum |                  |
| | | | V3 Transverse vaginal septum and/or imperforate hymen |                  |
| | | | V4 Vaginal aplasia |                  |

Discussion:
The incidence of pseudo-unicornuate uteri, although difficult to specify, is estimated to be 1 per 1000 women. In 10% of cases the rudimentary horn communicates with the unicornuate uterus, while non-communicating rudimentary horns with a cavity represent 36%. The prognosis of pregnancies in the unicornuate uterus is good with a rate of almost 80% of pregnancies and 60% carried to term with 17% of prematurity, with a tendency to breech presentation (34%), and a caesarean section rate of 45%, with evidence to support this, the case of our patient.

The circumstances of discovery are diverse, as reported in the largest series of pseudo-unicornus uteri with 42 cases collected from 1962 to 1995: 65% of cases in the presence of dysmenorrhea, 20% for pregnancy in the rudimentary horn, 15% on the occasion of an infertility assessment. The discovery of this anomaly can also be fortuitous.

Indeed, if there is a clinical symptomatology, this is secondary to the existence of a retained cavity, sometimes lined with a functional endometrium, exposing it to numerous gynaeco-obstetrical risks and requiring laparoscopic resection:

Pregnancy in a rudimentary uterine horn is rare, the incidence is estimated at 1/100,000 to 1/140,000, and is thought to result from intraperitoneal migration of spermatozoa or oocyte. The major complication of these pregnancies is the rupture of the rudimentary horn (90%), most often in the second trimester of the pregnancy, leading to a picture of hemoperitoneum or even a state of maternal shock.
Tubal pregnancy homolateral to the rudimentary horn is also described, leading to a classic picture of extra uterine pregnancy.

The problems of hypofertility have not been fully elucidated, but are present: difficulties in fertilization of the oocyte contralateral to the hemi-uterus have been mentioned, as well as problems linked to associated endometriosis. Endometriosis, the most frequent differential diagnosis due to the symptoms presented (menstrual pain, hypofertility, etc.), is associated in approximately 21 to 33% of cases.

Abnormalities of the urinary tract are frequently associated with this uterine malformation (38%) and are dominated by renal agenesis homolateral to the side of the blind horn, detected by MRI.

**Conclusion:**
This malformation is often asymptomatic. However, it may be revealed by pain in relation to hematometry, or by possible complications: pregnancy in the pseudo horn, dystocic presentations on the normal hemi-uterus as in the case of our patient, but also endometriosis or sterility. In some cases treatment is necessary and will be laparoscopic.

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