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Three Unique Cases of Uterine Anomaly: Atypical Müllerian Ducts Development

Abstract: Uterine anomalies have different types and are classified in different ways. Here, we report three hysterosalpingographically unique cases of uterine anomalies selected from more than 30,000 cases over duration of 40 years.

Case 1 is a branching out bicornuate uterus, in which the left uterine horn and cervix branch from the cervical canal. The second case is a unicornuate uterus, in which a fallopian tube without uterine cavity directly branches from this corn. The third case is a cross-shaped bicornuate uterus. Considering the embryologic development of uterus, we concluded that these three cases had abnormalities in different stages of their Müllerian ducts' development

Keywords: congenital anomalies, uterine malformations, urogenital abnormalities, hysterosalpingography

Introduction

The congenital anomalies of the uterus provide a fascinating subject for study. The incidence of congenital uterine abnormalities varies between 0.5 percent and 1.0 percent depending on the method used for diagnosis. Uterine malformations are usually detected during routine ultrasound examination and hysterosalpingography. Hysterosalpingography is the most sensitive and accurate method of diagnosis.[1]

In this article, we briefly describe the embryology of female genital system and a simple classification of uterine anomalies. (Table1) (Figure 1)[2-4]

Case Presentation

Case 1: A 26-year-old woman presented with a five-year history of infertility. Her menstrual cycles were irregular and were accompanied by dysmenorrhea. On examination, the vagina was single and a single cervical os was observed. On hysterosalpingography, a single vagina with one cervical os was seen. Contrast medium injected through this os revealed the right horn and its fallopian tube appearing normal; but the left uterine horn, especially the cervix and external os, branched out from the right cervical canal. In laparoscopy, two separate complete horns were observed, that were connected only at the cervix. The fallopian tubes and both ovaries were normal. (Figure 2)

Case 2: A 23-year-old women presented with five years of primary infertility. Her menses were regular with moderate bleeding, but neither intermenstrual bleeding nor dysmenorrhea. On speculum examination, a normal vagina and a normal exocervix were observed. On hysterosalpingography, on the right side, a unicornuate uterus was seen. On the left side, a fallopian tube without uterine cavity branched out of the right unicornuate uterus at the internal os. Laparoscopy confirmed the hysterosalpingographic findings of a right unicornuate uterus with a left hydrosalpinx, not communicating with the uterine cavity but joining the isthmus of the right horn. Both ovaries appeared normal. (Figure 3)

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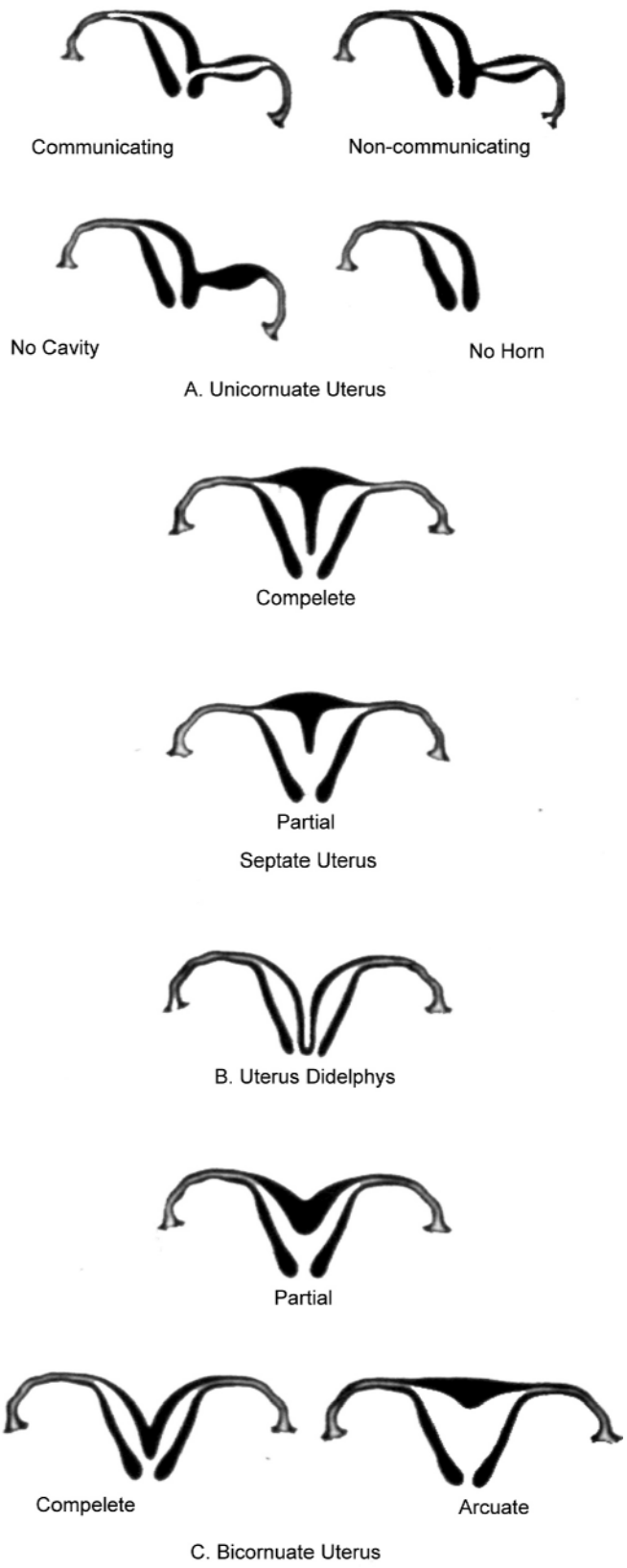

Figure 1: Classification of congenital uterine malformations

Table 1: Müllerian Duct Anomalies

I. Segmental Müllerian duct agenesis
II. Unicornuate Uterus
  A. With contralateral rudimentary horn
    1. With endometrial cavity
      a. Communicating
      b. Non-communicating
    2. Without endometrial cavity
  B. Without contralateral rudimentary horn
III. Didelphic uterus
IV. Bicornuate uterus
  A. Complete
  B. Partial
  C. Arcuate
V. Septate uterus
  A. Complete
  B. Incomplete

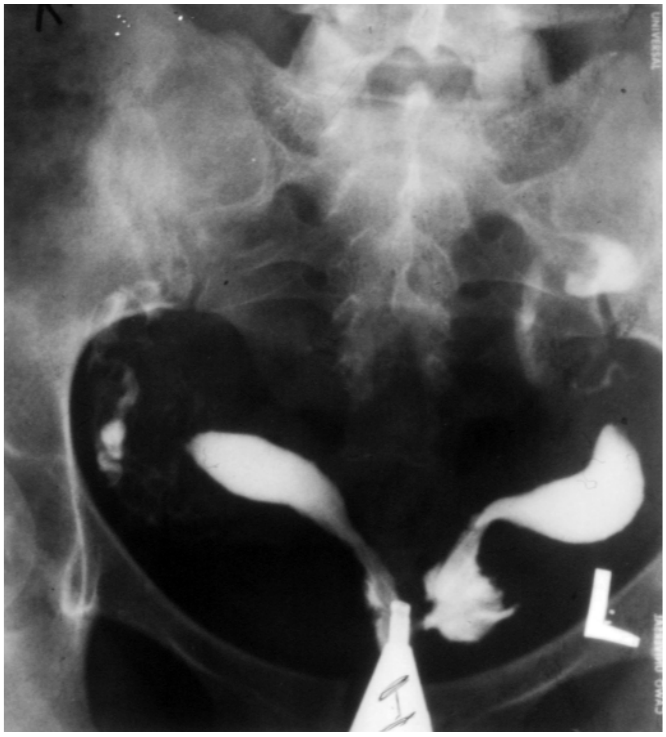

Figure 2: Hysterosalpingogram of case 1. The left uterine horn and cavity branched from right cervical canal.

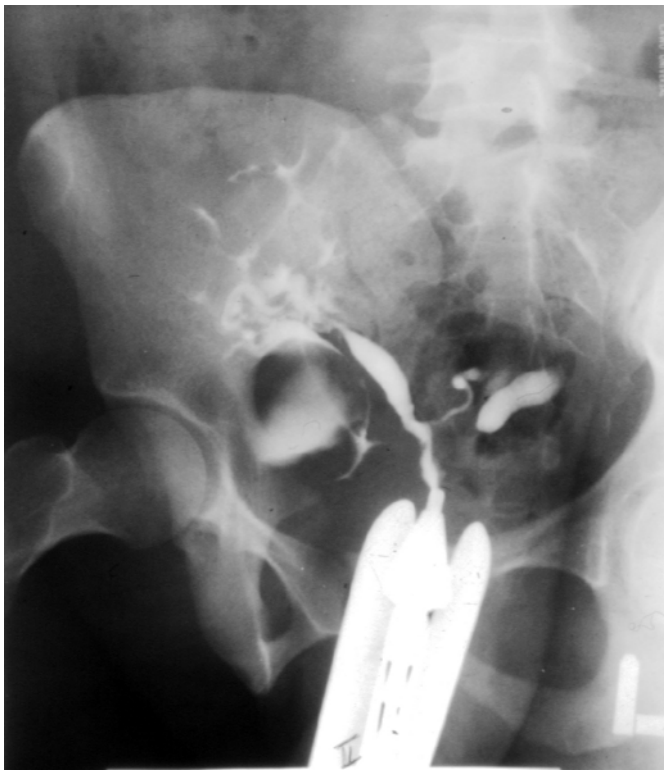

Figure 3: Hysterosalpingogram of case 2. A unique unicornuate uterus that a fallopian tube without left uterine horn and cavity branches from right cervical canal.

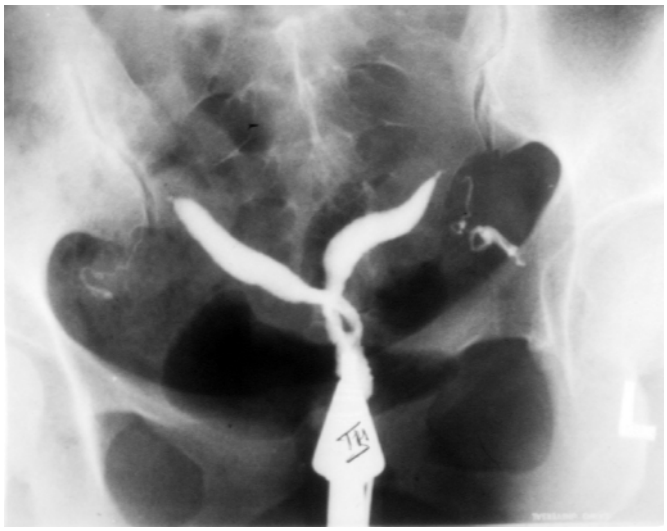

Figure 4: Hysterosalpingogram of case 3. Bicornuate uterus crossing at the region of isthmus.

Case 3: A 27-year-old woman, married one year before, presented with her second 8-week miscarriage. Her first 7.5-week abortion had occurred 5 months earlier. Hysterosalpingography showed one cervical canal and two separate cavities (uterus bicornis unicollis) and two crossing, non-parallel isthmi. Three months after the second abortion, she underwent hysteroscopic surgery. On laparoscopy and hysteroscopy done by another surgeon, a bicornuate septated isthmus (but not crossing) was confirmed. Only the septum between the horns was removed hysteroscopically, sparing the septum of the isthmi. Postoperative hysterosalpingography showed the septum of the isthmi and the repaired uterine cavity. (Figure 4) On intravenous pyelography (IVP), the right kidney was absent, but the urinary system was otherwise normal.

One year after the operation, the patient became pregnant spontaneously.

Discussion

For a better understanding of uterine anomalies, we first explain the process through which the uterus is developed: formation, fusion and absorption; according to AFS classification.[6]

Stage1: The uterus begins to form ten weeks after fertilization. The structures which give rise to uterus are a pair of tubes called the Müllerian ducts. At their cranial ends, these two tubes will form the fallopian tubes. Their caudal ends lie side by side close together. They will form the uterus by fusing together to become one structure. Fusion occurs between the two middle walls which face one another. They form one central wall (median septum) at the middle of one newly formed, larger tube.

Stage 2: The fusion of two Müllerian ducts at the beginning of the tenth week produces a cylindrical structure of equal diameter throughout. However, the fully developed uterus is a widening of the top of the central wall. As this wall grows, the whole tube becomes wider at the top, forming the part of the uterus known as the fundus. This process occurs in the embryo ten to thirteen weeks old.

Stage 3: The uterus is completed during the thirteenth to twentieth weeks of development. This completion is dependent on degeneration of the top of the median septum. This septum separates the two horns (cornua) of the uterus which are portions of the uterus that lie at the top furthest from the center. Its dissolution begins at the bottom of this septum and progresses toward the fundus of the uterus. When it is complete, a single, continuous uterine cavity results.[6]

Our reported cases are different with regard to these embryonic stages of development of the uterus. In unicornuate uterus, we almost have a rudimentary horn with a fallopian tube communicated or not to the principle corn; but in our case 1 (Figure 3), the left uterine horn and cervix branched from the right cervical canal. This is not a didelphic uterus because in more than 75 percent of didelphic cases, there are two complete separate uterine cavities and two cervical orae with a vaginal septum. However, this

case had a bicornuate uterus, with two united cervices in between and only one cervical os.

In case 2 (Figure 4), the left fallopian tube without uterine cavity branches out of the right uterine isthmus. This case does not fit in the unicornuate uterine classification.

Unicornuate uteri are divided into two classes (Table 1): class A, with a contralateral rudimentary horn and Class B without a rudimentary horn. Hydrosalpinx ensues with a risk of infection.

As it shows, neither of the cases 1 and 2 can be explained by the embryonic fusion stage of Müller's theory.

In case 3 (Figure 4), a crossed bicornuate uterus, the two Müllerian ducts are not side by side but cross each other .In hysteroscopy, this septum was observed but since it was inconvenient or unnecessary to be removed, just the cavity was repaired by hysteroscopy . On post-operative hysterosalpingography, this septum is obviously visible. (Figure 3) Absent right kidney on IVP is another unusual finding in this patient.

These three cases represent atypical Müllerian ducts development and hesitate this theory: In case 1, the fallopian tube directly originated from uterus and in case 2, one corn originated from cervix while in case 3, the corns cross each other in the region of isthmus.

Since 1967, Muller's[2] theory about embryonic formation of female genital system has remained almost unchanged. Nevertheless, it can not explain all malformations of the uterus. In 1957, Crosby et al [3] proposed that fusion of the two Müllerian ducts starts at their caudal ends in Müller's tubercle, and proceeds cranially up to the fundus. In 1929, Puddicomb [4] assumed that resorption may start at any level of the fused Müllerian ducts.

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