Common uterine anatomic anomalies

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Anatomy Elective
May 2007
quick basics/embryology review

• the main structures of the female reproductive tract: uterus, fallopian tubes, cervix, vagina, ovaries

Embryology:
• 2 paired müllerian ducts ultimately develop into the uterus, fallopian tubes, cervix, and the upper two thirds of the vagina.
• germ cells develop into ovaries
• urogenital sinus forms the lower one third of the vagina

• Female external genitalia:
• Clitoris develops from the genital tubercle (by slight elongation)
• Labia minora develop from the genital folds (by remaining separate)
• Labia majora develop from the genital swellings (by enlarging greatly)
• Vestibule develops from the lower most part of the urogenital sinus.
Normal uterine structure contains a single uterine cavity and flat or convex outer fundal contour.
anatomic anomalies of the uterus

- majority caused in embryologic development by mullerian duct anomalies
- categorized most commonly into 7 classes according to the American Fertility Society (AFS) Classification Scheme (1988) as follows:
Class I (hypoplasia/agenesis):

- uterine/cervical agenesis or hypoplasia
- most common form is the Mayer–Rokitansky–Kuster–Hauser syndrome, which is combined agenesis of the uterus, cervix, and upper portion of the vagina
- Patients have no reproductive potential
Class II (unicornuate uterus):

- complete, or almost complete, arrest of development of 1 müllerian duct
- arrest is incomplete in 90% of patients, and a rudimentary horn with or without functioning endometrium is present
- if the rudimentary horn is obstructed, it may present as an enlarging pelvic mass
- if the contralateral healthy horn is almost fully developed, a full-term pregnancy is believed to be possible (see didelphys uterus)
Class III (didelphys uterus):

- complete nonfusion of both müllerian ducts
- individual horns are fully developed and almost normal in size
- two cervices are present
- a longitudinal or transverse vaginal septum may be present—has the highest association with transverse vaginal septa, although septa may also be seen with other anomalies
- patients and doctors may consider metroplasty (plastic surgery of uterus)
- since each horn is almost a fully developed uterus, patients have been known to carry pregnancies to full term
Class IV (bicorhnuate uterus):

- partial nonfusion of the müllerian ducts
- the central myometrium may extend to the level of the internal cervical os (bicorhnuate unicollis) OR external cervical os (bicorhnuate bicollis).
- bicorhnuate bicollis vs didelphys uterus:
  - bicorhnuate bicollis has some degree of fusion between the two horns
  - classic didelphys uterus has the two horns and cervices separated completely
- horns of the bicorhnuate uteri are not fully developed; are typically smaller than horns of didelphys uteri
- some patients are surgical candidates for metroplasty
Class V: septate uterus

- failure of resorption of the septum between the two uterine horns
- septum can be partial or complete, in which case it extends to the internal cervical os
- septum may be composed of myometrium or fibrous tissue.
- uterine fundus is typically convex but may be flat or slightly concave

Women with septate uterus have the highest incidence of reproductive complications. Differentiation between a septate and a bicornuate uterus is important: septate uteri are treated using transvaginal hysteroscopic resection of the septum; bicornuate uterus is treated, if possible and/or indicated, by an abdominal approach to perform metroplasty.
Class VI (arcuate uterus):

- a single uterine cavity with a convex or flat uterine fundus
- the endometrial cavity is the uterine fundus, which has a small fundal cleft or impression ($\geq 1.5$ cm)
- outer contour of the uterus is convex or flat
- often considered a normal variant since it is not significantly associated with the increased risks of pregnancy loss and the other complications found in other subtypes.

[Images of normal and arcuate uterus]
Class VII (diethylstilbestrol–related anomaly):

- Several million women were treated with diethylstilbestrol (DES; an estrogen analog prescribed to prevent miscarriage) from 1945–1971.
- The drug was withdrawn when its teratogenic effects on the reproductive tracts of both males and females were discovered.
- The uterine anomaly is seen in the female offspring of as many as 15% of women exposed to DES during pregnancy.
- A variety of abnormal findings, including uterine hypoplasia and a T-shaped uterine cavity.
- Patients also may have abnormal transverse ridges, hoods, stenoses of the cervix, and adenosis of the vagina with increased risk of vaginal clear cell carcinoma. Imaging findings are pathognomonic for this anomaly.
Clinical Details: Suggestion of müllerian duct anomaly may arise in different clinical situations:

In the newborn/infant, initial presentation may be an obstructed system discovered as a palpable abdominal, pelvic, or vaginal mass (mucocolpos)

In adolescent girls, presentation may be delayed menarche and/or an obstructed system presenting as an intra-abdominal mass (hematocolpos). Cyclical pain often present

Women of childbearing age may present with varied problems, including infertility, repeated spontaneous abortions, or premature delivery. Part of an infertility workup includes routine imaging to look for anatomic anomalies.

The anomaly may be discovered incidentally during imaging eval for another condition or during surgery such as elective sterilization.
Examination and imaging of anomaly

- Imaging criteria to distinguish forms of uterine anomalies are based on the configuration of the endometrial cavity and (primarily) on the configuration of the uterine fundus.
- **Ultrasound**: first imaging is pelvic ultrasound (US) with transabdominal and transvaginal imaging (if anatomically possible).
- Newer 3-dimensional (3D) sonographic techniques offer relatively higher sensitivity and specificity than 2D images.
- **Hysterosalpingography** (HSG), performed under fluoroscopy, evaluates the uterine cavity and tubal patency. Anomalies may be suggested but positive findings often are nonspecific for precise diagnosis.
- **MRI**: gold standard for imaging uterine anomalies. Most types of uterine anomalies can be diagnosed using pelvic MRI. Provides high-resolution images of the uterine body, fundus, and internal structure. Can help evaluate the urinary tract for concomitant anomalies.
Effect of uterine anomalies on reproduction capabilities

- Study of 1089 women with no history of reproductive problems found 983 had normal uterine cavity, 72 an arcuate, 29 a subseptate, and 5 a bicornuate uterus

- Women with congenital uterine anomalies were more likely to have adverse pregnancy outcomes than women with a normal uterus

“Reproductive Outcomes in Women With Congenital Uterine Anomalies Detected by Three-Dimensional Ultrasound Screening”. Brigitte Woelfer, MD, Rehan Salim, MD, Saikat Banerjee, MD, Janine Elson, MD, Lesley Regan, MD, PhD and Davor Jurkovic, MD, PhD Obstetrics & Gynecology 2001;98:1099–1103
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