Uterus Didelphys

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Morbidity and Morbidity
Case Presentation

- 13 year old female presented with dull, intermittent abdominal pain x 2 days in the lower abdomen.
- The pain initially relieved with Motrin.
- Similar pains in 12/2009.
History continued..

- **PMH:** left renal agenesis, pneumonia

- **PSH:** Exploratory laparotomy, excision of cyst wall, drainage of intraperitoneal blood and appendectomy- 12/2009 (Brooklyn Hospital)

- All immunizations were up to date

- **SH:** non-contributory
Physical Exam

- **VS:** T 98.9  BP 119/78  HR 83  \( O_2 \) sat 100%
- **General:** well nourished, AAO x 3, no acute distress
- **HEENT:** NCAT, no conjunctival pallor, PEERL
- **CV:** RRR, \( S_1 S_2 \) normal
- **Pulm:** CTA bilaterally; no rales, rhonchi, crackle
- **GI:** BS present, soft nondistended, **TENDER IN THE LLQ**, no rebound tenderness
- **Rectal:** good sphincter tone, no gross blood
Labs

CBC: 9.2/13/41/325

BMP: 139/5 101/26 12/0.8 <80

LFTs: 9.7/7.1/29/11/180/0.1
Differential Diagnosis

- Ruptured ovarian cyst
- Hematosalpinx
- Ureterovaginal malformation
- Uterus didelphys
Radiologic Studies

- **Abdominal ultrasound**: absent left kidney
- **Pelvic US**: mass in LLQ
- **MRI (5/4)**: uterus didelphys with left hematocolpos and distended left sided endometrium and distended endocervix
- **MRI (5/6)**: right uterine horn and cervix extending into single vagina; *divergent rudimentary left uterine horn with fusion abnormality of left mullerian duct*
Hospital Course

- Taken for EUA, vaginoscopy and possible vaginal aspiration on HD #2
- Patient was and taken for an exploratory laparotomy on HD #3
- Intra-operative findings: didelphys uterus
  - Right: normal uterine system with a right ovary and a cervix opening into the vagina
  - Left: hematosalpinx with a distended uterus filled with dark blood and normal ovary.
Postoperative Course

- Pathology:
  - weak proliferation of endometrium;
  - endocervix with hematosalpingeal with paratubal adenomatoid tumor
  - Foci of endometriosis

- Diet was advanced on POD #1

- Discharged on POD #2
Normal Uterus
Paradenomatoid tumor
Calretinin immunostain
Outline

- Uterovaginal development
- Mullerian malformations
- Congenital fusion abnormalities
- Herlyn-Werner-Wunderlich Syndrome
Figure 106-23 Development of uterus and vagina. During the 10th week, the paramesonephric ducts fuse at their caudal ends to establish a common channel and come into contact with a thickened portion of the posterior urogenital sinus called the sinovaginal bulb. This is followed by development of the vaginal plate, which elongates between the 3rd and 5th months and becomes canalized to form the inferior vaginal lumen. (Modified from Sadler TW: Langman's Medical Embryology. Baltimore, Williams & Wilkins, 1985.)
Müllerian malformations

- True duplication of the uterus is rare
- Failed resorption of the common medial wall of the paired mullerian ducts
- More than 50 cases of uterus didelphys with a unilateral imperforate vagina
- Renal and axial skeletal abnormalities
Mullerian Ductal Defects

a. Agenesis and hypoplasia
b. Vertical fusion defects
c. Lateral fusion defects

Incidence: 1 of 200-600 fertile women
Congenital fusion abnormalities

- Bicornuate uterus - 37%
- Arcuate or incomplete septum - 28%
- Complete septum - 9%
- Didelphic uterus - 11%
- Unicornuate uterus - 4%
Figure 129-12  Fusion anomalies of the müllerian system. Upper left to lower right, bicornuate uterus (partial), bicornuate uterus, uterine duplication, complete uterine and vaginal duplication, duplication of the uterus and cervix with a unilateral imperforate vagina.  (From Yerkes EB, Rink RC: What urologists should know about pediatric gynecologic abnormalities. Contemp Urol 2002;14:12.)
Herlyn-Werner-Wunderlich Syndrome

- Didelphic uterus, obstructed hemivagina and ipsilateral renal and ureteral agenesis
- Failure of both lateral fusion and vertical fusion

Theory:
- Damage to the caudal portion of the wolffian duct
Anatomical defect is a failure of lateral fusion of mullerian ducts combined with failure of vertical fusion between mullerian duct and urogenital sinus (hemivaginal septum ipsilateral to the side of renal agenesis. (Journal of Ped Surgery 2006. 41, 987-992)
Associated anomalies

- Ipsilateral renal agenesis is commonly seen
  - Inferred that an early teratogenic process active in 4th week of gestation resulted in arrested growth of one mesonephric duct → Agenesis of the ureteric bud

- IVC duplication, intestinal malformation, ovarian malposition
On history and physical...

- **Sx:** asymptomatic until menarche
  - cyclic or chronic pelvic pain
  - Does not result in primary amenorrhea

- **PE:**
  - Unilateral abdomino-pelvic mass that terminates in a bluish bulge in lateral vaginal mass
Diagnosis

- **Imaging:**
  - **MRI:** two uterine horns usually widely separated, with preservation of the endometrial and myometrial widths
  - **US:** renal anomalies on the side ipsilateral to the obstructed system
Complications

• If left untreated:
  – Endometriosis
  – Pelvic adhesions
  – Pyosalpinx or pyocolpos

• Spontaneous abortion – 40%

• Fertility is comparatively good
Management

- **Diagnostic Laparoscopy**

- **Treatment:**
  - Wide incision of vertical vaginal septum to release the entrapped menstrual blood
  - Resection of the vaginal septum
  - Marsupialization of vaginal margins
  - Hemihysterectomy and salpingo-oophorectomy
Management and outcome of patients with combined vaginal septum, bifid uterus, and ipsilateral renal agenesis (Herlyn-Werner-Wunderlich syndrome)

- 80 patients with uterine and vaginal abnormalities
  - 12 patients with HWWS
  - Preferred surgical approach: full excision and marsupialization of the vaginal septum
  - Hemihysterectomy with or without salpingooophorectomy is rarely indicated


Wein: Campbell-Walsh Urology, 9th ed.; Chapter 129 - Surgical Management of Intersexuality, Cloacal Malformation, and other Abnormalities of the Genitalia in Girls > ... > Obstructive Genital Anomalies

Adam: Grainger & Allison's Diagnostic Radiology, 5th ed.

Thank you

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Dr. Velcek.

Small part choking hazard
The ovaries may be pulled off and become a choking hazard. Keep away from children.