Clinical Pathological Correlation: Case Two

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Case Presentation

- Chief Complaint
  - 14 y.o. female
  - Two weeks of diffuse dull lower abdominal pain
  - G0
  - LMP - ?
    - None for at least one year

Developmental History (from mother)

- Product of an uneventful term pregnancy
- Telarche & adrenarche
  - Uncertain
- Menarche
  - Age 12
  - Irregular, infrequent, light for ~one year
Review of Systems

- Urinary frequency without dysuria
- Early satiety
- No fever, anorexia, weight loss, or diarrhea
- Denies sexual activity

Physical Examination

- 60”, 110 pounds
- Generalized facial and truncal acne
- Normal breast development; no lactorhea
- Hirsuitism - male escutcheon
- Clitoromegaly
- Visible abdominal protuberance
  - Indistinct, firm, immobile pelvic-abdominal mass

Characterization of Symptoms

- Pain
  - Left greater than right, non-radiating
  - Dull & colicky
    - Worse with movement
    - Progressively more constant over the last two weeks

Radiographic Studies

Ultrasound

- Very large, predominantly solid, pelvic mass
- Thick septations & solid areas
- 26 x 26 x 14cm
- Normal kidneys

CT Scan*

*Representative image
**Laboratory Results**

- β-hcg negative
- α-fetoprotein 1.0 ng/ml (normal <10)
- CEA 0.9 ng/ml (normal <3)
- CA-125 586 IU/ml (normal <35)
- Testosterone 2.19 ng/ml (nl 0.1-0.9)
- Karyotype unknown

**Thoughts?**

- What could this possibly be?
- Differential diagnosis
- What is my next diagnostic step?
- Clinical Approach
- How do I counsel this patient and her parents?
- Support & education

**Operative Findings**

EUA
- mass above umbilicus

Intraoperative
- large left ovarian mass
  - appeared hemorrhagic, but intact
- normal uterus and right ovary/tube

**Peri-Menarchal Pelvic Mass**

**Age Based Differential Diagnosis**

**Benign**
- Teratoma
- functional cyst?

**Malignant**
- Germ Cell
- Sex Cord - Stromal
- Wilm's Tumor
Distribution of Benign Ovarian Neoplasms

Distribution of Malignant Ovarian Neoplasms

Left Ovarian Tumor
Pathologic Diagnosis
- Sertoli-Leydig cell tumor
  - high grade
    - roughly equal amounts of:
      - intermediate S-L differentiation
      - sarcomatoid differentiation
    - >15 mitoses/10hpf
  - heterologous elements
    - osteoid
    - rhabdomyoblastic differentiation

Sex Cord - Stromal Tumors
- Granulosa Cell
  - Adult
  - Juvenile
- Thecoma-Fibroma
  - thecoma
  - fibroma
  - fibrosarcoma
- Sertoli-Leydig
Sertoli-Leydig Cell Tumors

- Sertoli Cell
- Sertoli-Leydig
  - well differentiated
  - intermediate
  - poorly differentiated
  - heterologous
- Retiform Sertoli
  - younger ages, more aggressive

- All ages
  - 75% <30yo
  - 10% >50yo
- Virilization may occur ~30%

Sertoli-Leydig Tumors

- Well Differentiated
- Intermediate
- Retiform

Sertoli-Leydig Cell Tumors with heterologous elements

- occur in 20% of S-L tumors
  - usually intermediate to poorly diff tumors
- intestinal differentiation - 20%
- carcinoid tumor - 16%
- heterologous elements - 5%
  - cartilage, bone, skeletal muscle

Sertoli-Leydig Cell Tumors

- Stage I - Most Common
- Survival Rates (5 year)
  - Well differentiated - 100%
  - Intermediate - 89%
  - Poorly diff - 41%
- Heterologous Elements
  - range 20-81% 5 yr
  - cartilage & muscle appear to be worse prognostically
  - Recurrence usually within 6-12 months

CPC Case 2
Follow-Up: High Grade SLT with heterologous elements

- chemotherapy
  - 6 cycles of BEP
    - bleomycin, etoposide & platinum
- NED
  - Currently 3 years out

The Hormonally Active Pelvic Mass

Sex cord-stromal tumors
- granulosa tumors
  - adult or juvenile types
  - up to 80% isosexual precocity
- Sertoli-Leydig tumors
  - 20-50% virilizing
  - retiform variant most common SLCT < age 20

Germ Cell Tumors
- precocious puberty can occur
  - usually associated with hCG production
    - Endodermal sinus tumor
    - Embryonal carcinoma
    - Dysgerminoma (5%)

CPC Case Two — Summary

- Pelvic mass in a child
- differential diagnosis
- overview of age based tumor prevalence
- Histologic features of Sertoli-Leydig Tumor
  - classification
  - heterologous elements
- Tumor marker differentiation
  - don't forget the extra blood for markers