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 lactorrhea
- 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*
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
- Procedure: LSO, omentectomy, lymph node sampling

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

- Germ cell
- Epithelial
- Stromal


Distribution of Malignant Ovarian Neoplasms

- Epithelial
- Germ cell
- Stromal


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

Virilization may occur ~30%

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

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
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