Clinical Pathological Correlation: Case Two

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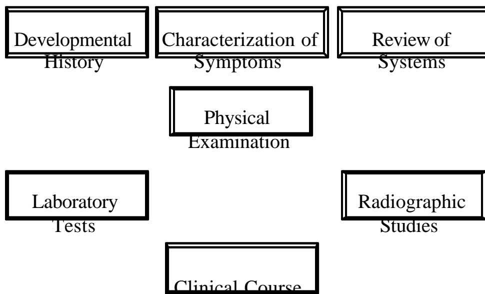
Case Presentation

- Chief Complaint
  - 14 y.o. female
    - Two weeks of diffuse dull lower abdominal pain
    - G_0
    - LMP - ?
      - None for at least one year

Case Two – Road Map



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

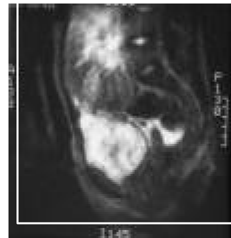
Characterization of Symptoms

- 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*



*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	

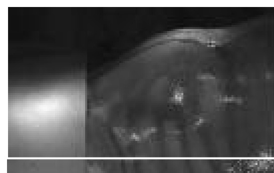
Thoughts?

- 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



Procedure
- LSO, omentectomy, lymph node sampling

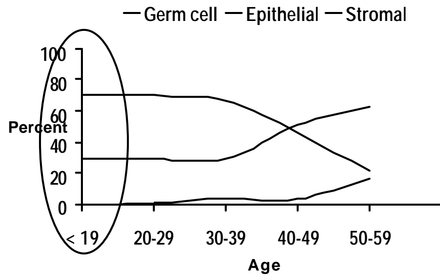
Peri-Menarchal Pelvic Mass

Age Based Differential Diagnosis

Benign
- Teratoma
- functional cyst?

Malignant
- Germ Cell
- Sex Cord - Stromal
- Wilm's Tumor

Distribution of Benign Ovarian Neoplasms



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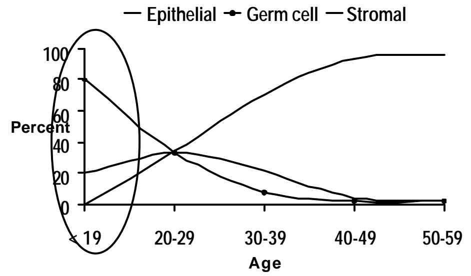
Koonings et al. Obstet Gynecol 74:921 1989

Distribution of Malignant Ovarian Neoplasms



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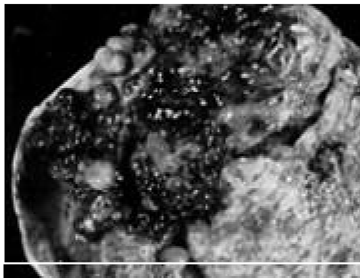
Left Ovarian Tumor



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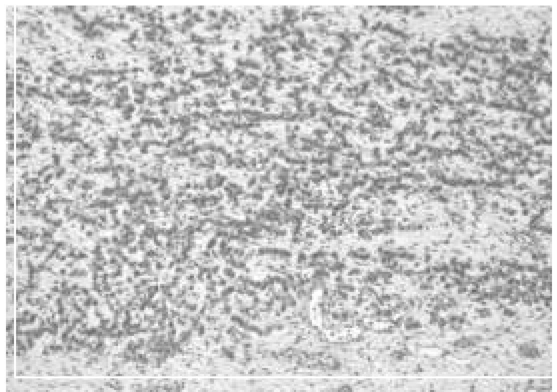

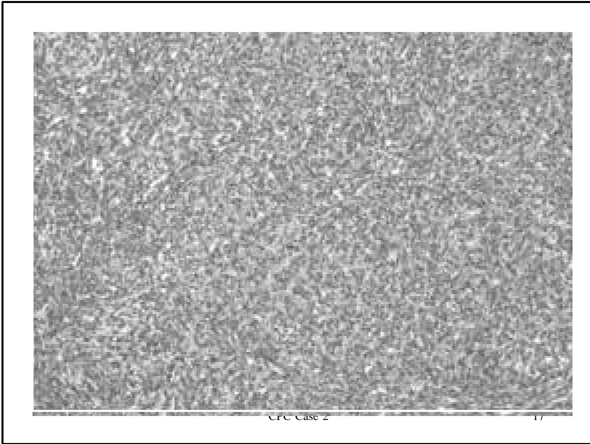
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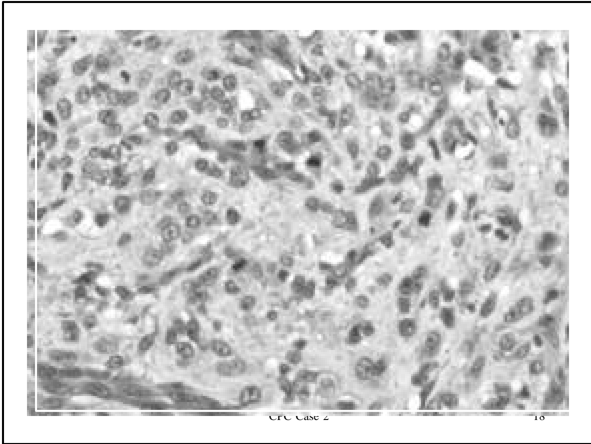
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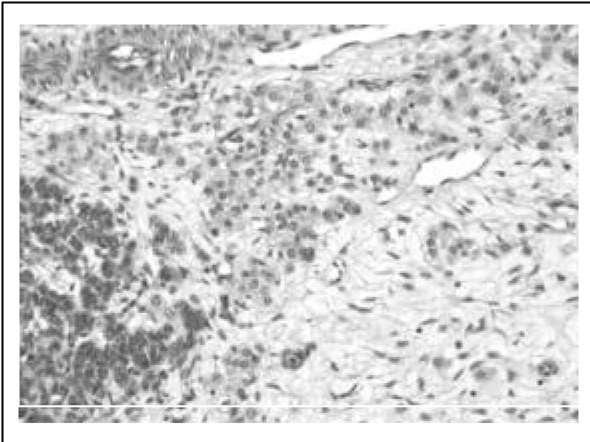
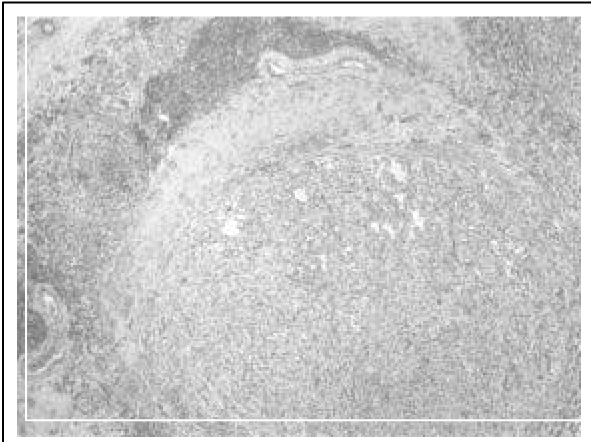
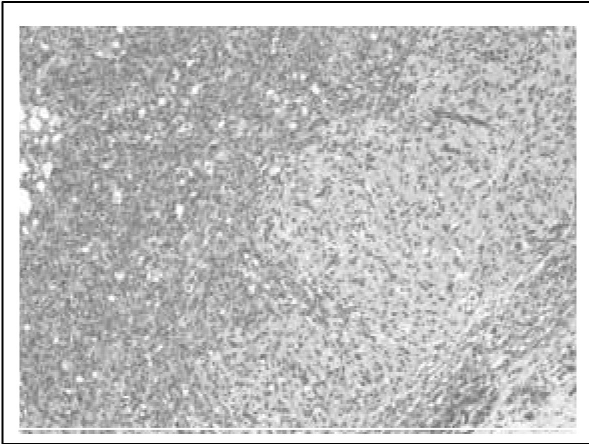
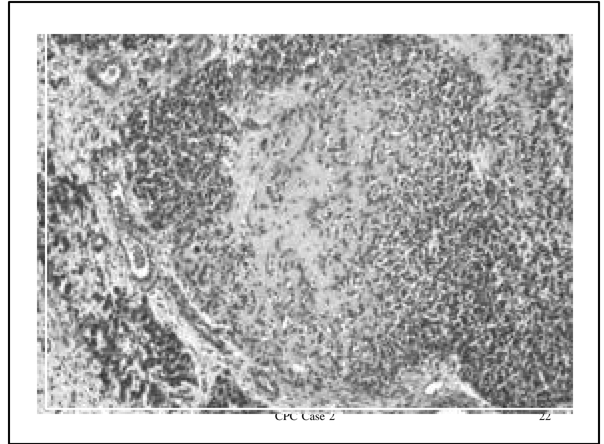








Pathologic Diagnosis

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

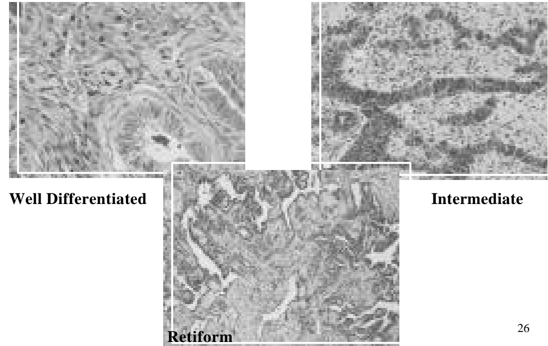
Sex Cord - Stromal Tumors

- 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



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

Follow-Up: High Grade SLT with heterologous elements

- chemotherapy
  - 6 cycles of BEP
    - bleomycin, etopiside & 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*