Sex cord stromal tumours of the testis

Edinburgh 2013

Dan Berney
Testicular pathology in practice

• Increased U/S examination
• Non-palpable lumps
• Germ cell marker negative
• ? Clinical decisions...
• Many are sex cord/stromal tumours.
Increase in incidental lesions in the testis.

- Victim Of Modern Imaging Technology
- VOMIT
- Brainless Application of Radiological Findings
- BARF
Derivation of Sex cord stromal cells
Coelomic mesothelium = Sex cords = Sertoli cells

Mesenchyme = Stroma = Leydig cells
Rule for Testicular Tumours

Anything that happens in the ovary can happen in the testis
(except for spermatocytic seminoma)
• Leydig cell tumours
• Other sex cord/stromal tumours
• Criteria for malignancy
• Treatment strategies
• Associated syndromes
• Common pitfalls in diagnosis
• Immunohistochemistry
Sex cord stromal Tumours

Leydig
Sertoli
Granulosa
Sertoli/Leydig
Fibroma and thecoma
Sex cord stromal NOS
Leydig cell tumours

- Typical appearance
- Oncocytic
- Reinke Crystalloids
- Lack of IGCNU
- 30% have endocrinopathy
- Children 20%
  - German Zoologist 1821-1908
German anatomist, 1862-1919

40%?
Variation in LCTs

• Spindled
• Adipose metaplasia
• Ossification
Leydig cell hyperplasia and variants

- High hCG
- Klinefelter’s syndrome
  - Bilateral nodularity
  - Not a reason for Orchidectomy
  - Cord and tunica involvement
- Congenital Adrenal Hyperplasia
  - Testicular Adrenal Rest Tumours (TART)
Congenital adrenal hyperplasia

- Many autosomal recessive mutations inhibiting formation of cortisol from cholesterol
- Excess androgens common
Testicular tumours of the adrenogenital syndrome (TTAGS)

Testicular adrenal rest tumours (TART)

Adrenal cortical cell rests in cord and hilum, especially rete testis.

Hyperplasia

Multiple nodules
Sertoli Cell Tumours

• Sometimes oestrogens
• Variable! Tubules, retiform, solid, clear cells
• Charcot-Böttcher filaments
• Any tubular component is suspicious
• Enrico Sertoli 1842-1910
Sertoli cell adenoma/hyperplasia

- Cryptorchidism
- ‘Pick’s’ adenoma
- Androgen insensitivity
Sertoli cell tumours

- Sertoli cell tumour NOS
- Sclerosing sertoli cell tumour
- Large cell calcifying sertoli cell tumour
Leydig cell Malignancy Criteria

- Size
- Invasiveness
- Mitoses
- Necrosis
- RPLND??
Does Retroperitoneal Lymph Node Dissection Have a Curative Role for Patients with Sex Cord–Stromal Testicular Tumors?

Ashraf A. Mosharafa, M.D.¹
Richard S. Foster, M.D.¹
Richard Bihrlle, M.D.¹
Michael O. Koch, M.D.¹
Thomas M. Ulbright, M.D.²
Lawrence H. Einhorn, M.D.³
John P. Donohue, M.D.¹

¹ Department of Urology, Indiana University School of Medicine, Indianapolis, Indiana.
² Department of Pathology, Indiana University School of Medicine, Indianapolis, Indiana.
³ Department of Medicine, Indiana University School of Medicine, Indianapolis, Indiana.

<table>
<thead>
<tr>
<th>Pathologic feature</th>
<th>No. of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tumor size &gt; 5 cm</td>
<td>2</td>
</tr>
<tr>
<td>Necrosis</td>
<td>7</td>
</tr>
<tr>
<td>Moderate or marked nuclear atypia</td>
<td>6</td>
</tr>
<tr>
<td>Angiolympathic invasion</td>
<td>2</td>
</tr>
<tr>
<td>Infiltrating margin/paratesticular invasion</td>
<td>4</td>
</tr>
<tr>
<td>Greater than 5 mitotic figures per 10 HPF³</td>
<td>10</td>
</tr>
</tbody>
</table>

HPF: high power field.
³ With a × 40 objective.
Large cell calcifying SCTs

- Peutz-Jeghers
- Carney syndrome: myxomas, lentigines, blue naevi, Cushing's 2ary to PANH, pituitary adenomas.
- Younger age
LCSCTs

• Intra tubular growth
• Syndrome associated show less calcification
• Malignancy in older
Sertoli Leydig

• Hardly reported
• Mimics by invasion of tumour into Leydig cells
• ? Heterologous elements?
Invasion of hyperplastic rete with mucinous metaplasia
Granulosa cell tumours

• Usual type
  – Rare in pure form: morphology.
  – Usually benign

• Juvenile
  – Commonest testis tumour in infants
  – XX/XY mosaicism
  – Invariably benign

• Fibroma/thecomas very rare on own.
Mixed sex cord stromal tumours

• More common.
• Benign under 10
• Malignancy reported in older group
Immunochemistry

• Germ cell markers negative
• Inhibin variable
• Calretinin variable
• Actin spindle elements
• CD99 variable
• S100
Mixed sex cord/stromal/germ cell tumours

- Gonadoblastomas
- Other mixed tumours
Gonadoblastoma

- An ‘in situ’ malignancy
- Risk of any germ cell tumour is high
Gonadoblastomas

- Mixture of seminoma and sertoli cells
- Dysgenetic gonads
- Intersex conditions: 20% phenotypic males
- Bilateral in one third.
Presentation

• Usually young males with feminizing symptoms.
• Often persistent mullerian duct structures
• Karyotype can be normal but always a Y chromosome.
Tips for diagnosis

• Think of hyperplasia.
• Syndromes.
• Always look for IGCNU.
• Check nuclear morphology.
• Immunochemistry.