TUMORI TESTICOLARI ETA’ PEDIATRICA

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Università di Catania
TUMORI TESTICOLARI PEDIATRICI

Pre-pubertal

Benign (74%)

Germ cell tumors
- Teratoma
- Epidermoid cyst
- Yolk sac tumor

Malignant low metastatic tendency

Testis sparing surgery, if possible

Post-pubertal
Until recently, the majority of prepuberal testicular tumors have been considered to be malignant.

Tumor registries (AFIP; AAP; PTTR), urological textbooks and literature surveys all assert that yolk sac tumors are the most common primary testicular tumors in boys 12 years and younger.

It now appears that benign testicular tumors have been under-reported. Metcalfe PD et al. J Urol 2003; Pohl HG et al. J Urol 2004; Agarwal PK, Palmer JS. J Urol 2006
Patient age at diagnosis was *prenatal* to approximately **11 years**
(mean 43 and median 18 months).
PREPUBERTAL TESTIS TUMORS: ACTUAL PREVALENCE RATE OF HISTOLOGICAL TYPES

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From the Departments of Urology, Children’s National Medical Center, Washington, D. C. (HGP, HGR), Division of Pathology (DSH), Children’s Hospital of Philadelphia, Philadelphia, Pennsylvania (ARS), Hospital for Sick Children, Toronto, Ontario, Canada (PDM, DJE), and Children’s Hospital, Boston, Massachusetts (BGC, ABR)

Single Institution study

PREPUBERTAL TESTIS TUMORS

Incidence of pathological types of prepubertal testis tumors

<table>
<thead>
<tr>
<th>Pathological Type</th>
<th>Hospital for Sick Children</th>
<th>Children’s Hospital of Philadelphia</th>
<th>Children’s National Medical Center</th>
<th>Children’s Hospital</th>
<th>% Total Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yolk sac</td>
<td>4</td>
<td>8</td>
<td>1</td>
<td>2</td>
<td>15</td>
</tr>
<tr>
<td>Teratoma:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>62</td>
</tr>
<tr>
<td>Mature</td>
<td>19</td>
<td>11</td>
<td>7</td>
<td>6</td>
<td>48</td>
</tr>
<tr>
<td>Immature</td>
<td>3</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>14</td>
</tr>
<tr>
<td>Subtotal</td>
<td>22</td>
<td>12</td>
<td>7</td>
<td>6</td>
<td>23</td>
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<tr>
<td>Epidermoid Cyst</td>
<td>5</td>
<td>6</td>
<td>2</td>
<td>1</td>
<td>14</td>
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<tr>
<td>Gonadal stromal:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>3</td>
</tr>
<tr>
<td>Granulosa</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Leydig</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Sertoli</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Mixed</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Subtotal</td>
<td>2</td>
<td>6</td>
<td>1</td>
<td>4</td>
<td>13</td>
</tr>
<tr>
<td>Other:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>11</td>
</tr>
<tr>
<td>Gonadoblastoma</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Cystic dysplasia</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>1</td>
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<tr>
<td>Lymphoma</td>
<td>3</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Inflammatory</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1</td>
</tr>
<tr>
<td>Subtotal</td>
<td>1</td>
<td>6</td>
<td>2</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Overall</td>
<td>34</td>
<td>38</td>
<td>13</td>
<td>13</td>
<td>98</td>
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</table>

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PREPUBERAL GERM CELL TUMORS

TERATOMA
- Mature
- Immature
  - BENIGN

YOLK SAC TUMOR
- MALIGNANT
  - PROGNOSIS BETTER THAN POST-PUBERTAL

Virtually absent or only rarely detected
Prepubertal teratomas account for 30% of testicular germ cell tumors in children.

**Ultrasound**: solid mass with cystic areas; calcifications

**AFP serum levels**: normal or slightly increased (*less than 100 ng/ml*)

They usually show a pure histology (without yolk sac component) with no evidence of intratubular germ cell neoplasia (88% in adults).

These teratomas are uniformly benign in prepubertal testes irrespective of histology (mature or immature).

Teratomas have a malignant behaviour in postpubertal children and adults.
Prepubertal teratoma arises from a benign germ cell

Post-pubertal teratoma arises from IGCNU
PREPUBERTAL TESTIS TERATOMAS

Testis sparing enucleation

No evidence of recurrence

Benign behaviour
Epidermoid cyst

- AFP normal
- No associated IGCT
- Benign course after sparing surgery
Stromal-sex cord tumors

Juvenile Granulosa Cell Tumor (1.2%) most common in infancy

Multicystic hypoechoic circumscribed mass

AFP normal

BENIGN: no recurrence or metastases
At least **93%** of yolk sac tumors stain positively for this marker and are associated with increased serum levels.

AFP normalizes after surgery, unless metastatic disease.

85% presents at stage I (vs. 35% in adults).

**PROGNOSIS BETTER THAN POST-PUBERTAL**
Testicular tumors incidence is about 0.5-2 per 100,000 in children.

Testicular tumors are 10 times more frequent in postpubertal males than in boys 12 years and younger (5.4 per 100,000 in adults).

In children, most testicular tumors (74%) are benign, especially before puberty.

98% of malignant germ cell tumors in prepubertal testes is a pure yolk sac tumor.

Seminoma, embryonal carcinoma and choriocarcinoma, the more commonly encountered malignant germ cell tumors in adults, are not reported in prepubertal testis.
### Table 1. Presenting signs and symptoms

<table>
<thead>
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<th>Condition</th>
<th>No. Pts (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mass</td>
<td>42 (85)</td>
</tr>
<tr>
<td>Trauma, bruising</td>
<td>4 (8)</td>
</tr>
<tr>
<td>Hydrocele, hernia</td>
<td>3 (6)</td>
</tr>
<tr>
<td>Cryptorchidism</td>
<td>1 (2)</td>
</tr>
<tr>
<td>Virilization</td>
<td>1 (2)</td>
</tr>
<tr>
<td>Incidental finding</td>
<td>1 (2)</td>
</tr>
</tbody>
</table>
ULTRASONOGRAPHY

INTRATESTICULAR
SENSITIVITY UP 100%

PARATESTICULAR

TERATOMA

Well circumscribed masses with heterogeneous solid and cystic areas, and occasionally calcification.

EPIDERMOID CYST

Well circumscribed masses with typical lamellated internal echoes.

LEYDIG CELL TUMOR

Well circumscribed hypoechoic mass.
Ultrasonography Underestimates the Volume of Normal Parenchyma in Benign Testicular Masses

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Fig. 3. Ultrasound shows testicular teratoma. A, complex mass with cystic and solid elements. Arrowheads indicate thin rim of normal parenchyma. B, postoperative image demonstrates normal sonographic appearance.
Testis Tumors, Cryptorchidism, Varicocele

CLINICAL BEHAVIOR AND A CONTEMPORARY MANAGEMENT ALGORITHM FOR PREPUBERTAL TESTIS TUMORS: A SUMMARY OF THE PREPUBERTAL TESTIS TUMOR REGISTRY

JONATHAN H. ROSS, LISA RYBICKI AND ROBERT KAY

From the Section of Pediatric Urology, Cleveland Clinic Urological Institute and Cleveland Clinic Department of Biostatistics and Epidemiology, Cleveland, Ohio

<table>
<thead>
<tr>
<th>TABLE 3. AFP levels in infants with yolk sac tumor or teratoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. pts.</td>
</tr>
<tr>
<td>---------</td>
</tr>
<tr>
<td>No. pts.</td>
</tr>
<tr>
<td>AFP (ng./ml.):</td>
</tr>
<tr>
<td>Median</td>
</tr>
<tr>
<td>Range</td>
</tr>
</tbody>
</table>
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Our clinical approach conforms to the recommendation of Ross et al that only infants between 6 and 12 months with serum AFP greater than 100 ng/ml and patients older than 1 year with AFP greater than the normal range in adults should be excluded from a testis sparing approach. In addi-
Fig. 5. Algorithm for initial surgical management of prepubertal patients with testis tumor.

Intraoperative diagnosis of prepubertal testis tumor is highly reliable in distinguishing benign from malignant lesions.
Testicular mass in children (0 weeks to 12-13 years)

Testicular ultrasound

Doubt

Suggestive of benign tumor

AFP serum levels

AFP high levels

Frozen section

Malignant or suspicious

Benign

TERATOMA (mature or immature)

age: 11-13 years

Prepuberal

Frozen section adjacent normal parenchyma

Puberal

Radical orchietomy

AFP normal or age-related slightly increased

OTHER (sex cord-stromal tumor; epidermoid cysts)

Technically salvageable parenchyma

No

Si

Testis sparing surgery
CASE 1

6-month-old infant

- Testicular mass
- Ultrasound: suggestive of teratoma
- AFP: 79 ng./ml

excisional biopsy

Frozen section

Teratoma (mature or immature)

Testis sparing surgery if technically salvageable prechnyma
CASE 2

12-month-old infant

• Testicular mass
• Ultrasound: suggestive of teratoma
• AFP: 90 ng./ml

→ excisional biopsy

→ Frozen section

Teratoma
(mature or immature)

→ Testis sparing surgery
if technically salvageable prencyma
CASE 3

9-month-old infant

- Testicular mass
- Ultrasound: suggestive of benign tumor
- AFP: 1700 ng./ml

Yolk sac tumor highly suspected

ORCHIECTOMY
CASE 4

12-years-old child

- Testicular mass
- Ultrasound: suggestive of teratoma
- AFP: normal

![Diagram of surgical decision tree]

- Excisional biopsy
  - Frozen section
    - Tumor mass
      - Teratoma? (Mature or Immature)
        - Prepubertal
          - Testis sparing surgery
            (if technically salvageable parenchyma)
        - Pubertal
          - Adjacent normal parenchyma?
            - Orchiectomy
              Follow-up for malignant tumor