10 y/o female presented with mass at lower back.
Mass noticed 5 years ago, but enlarged over the last 2 years.
Denied lower back or abdominal pain.
PMH
• Born full term without complications.
PE
• Well developed 10 y/o female, 146 lbs.
• 10 x 12 cm mass over sacrum.
• Abdomen- soft, nontender, nondistended.
• DRE- mass not palpable
• No LE motor or sensory deficit.
Labs
• CBC, Chem, Coags - WNL
CT Scan

- 15 X13X10 cm complex cystic lesion posterior and anterior to sacrum.
- No continuity with spinal canal
Surgical Resection

Posterior Approach

- Ampicillin, Gentamycin, Flagyl given.
- Prone position
- Semilunar incision
- Mass attached to periostium of sacrum and coccyx.
- Coccygectomy
- Yellow fluid from cystic cavity
- Dissection in plane posterior to rectum.
- JP drain over muscle closure.
Surgical Resection
Pathology

- Multiloculated cystic mass 24.5 x 14 x 8 cm
  - External component
  - Internal component
- Culture of yellow thick fluid – negative.
- Epidermoid cyst with chronic inflammation.
Post Op Course

- Pt received abx postop.
- POD #1 - Tolerated PO diet.
- POD #2 - + BM
- POD #3 – Discharge Home with PO abx, JP with SS fluid.
Sacroccocygeal Teratomas
Germ Cell Tumors (GCT)

Teratoma
- Gonadal
  - Ovarian
  - Testicular
- Extragonadal
  - Sacrococcygeal
  - Head and Neck
  - Mediastinal
  - Retroperitoneal

Other histologic types
- Seminoma (Dysgerminoma)
- Yolk sac tumor (Endodermal Sinus)
- Embryonal Carcinoma
- Polyembryoma
- Choriocarcinoma
- Mixed GCT

Rare
Germ Cell Tumors (GCT)

- 2.4 cases per million children
- 1% of cancers diagnosed in children younger than 15 years
- Gonadal and Extragonadal sites
- Originate from the primordial germ cell.
Migration of Germ cells – 4th and 5th Weeks Gestation

- Germ cells migrate from yolk sac to the gonadal ridges and incorporate into gonadal tissue.

- Migration mediated by the c-kit receptor and stem cell factor.

- Failure in migration $\rightarrow$ deposition of germ cells in abnormal locations:
  - Anywhere from the brain to the coccygeal area, usually in the midline.
    - Sacrococcygeal area, mediastinum and retroperitoneum.

- Malignant transformation can occur at any of these sites.

- Germ cells are totipotential $\rightarrow$ tumor types associated with the degree of cell differentiation.
Staging for Malignant Extragonadal Germ Cell Tumors

Stage I: Localized disease, with complete resection at any site (coccygectomy for sacrococcygeal site); negative tumor margins; tumor markers positive or negative

Stage II: Microscopic residual disease; capsular invasion; negative lymph nodes or microscopic lymph node involvement; tumor markers positive or negative

Stage III: Gross residual disease; gross lymph node involvement; cytologic evidence of tumor cells in ascites or pleural fluid; tumor markers positive or negative

Stage IV: Distant metastases involving lungs, liver, brain, bone, distant nodes, or other sites.

*Children's Oncology Group and the National Cancer Institute.*
**Teratomas**

- Most frequently occurring GCT
- Extragonadal → infancy and early childhood
- Gonadal → older children
- More than half of teratomas are observed at birth.
- Diagnosed generally during the first year of life.
- 20% - malignant component
  - endodermal sinus tumor.

### Site of Tumor Occurrence

<table>
<thead>
<tr>
<th>Site</th>
<th>Incidence (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sacrococcygeal region</td>
<td>45-65</td>
</tr>
<tr>
<td>Anterior mediastinum</td>
<td>10-12</td>
</tr>
<tr>
<td>Gonadal (ovary and testis)</td>
<td>10-35</td>
</tr>
<tr>
<td>Retroperitoneum</td>
<td>3-5</td>
</tr>
<tr>
<td>Cervical area</td>
<td>3-6</td>
</tr>
<tr>
<td>Presacral area</td>
<td>3-5</td>
</tr>
<tr>
<td>Central nervous system</td>
<td>2-4</td>
</tr>
<tr>
<td>Other rare sites (&lt;1)</td>
<td></td>
</tr>
</tbody>
</table>
  - (e.g., liver, kidney, vagina, stomach)

Teratomas

- Solid, cystic, or mixed lesions.
- Tissues from each of the three germ layers.
  - Ectoderm
    - Epidermal and dermal structures – hair, sebaceous and sweat glands, teeth.
    - Neuroepithelial and glial tissue.
  - Mesoderm
    - Fat, cartilage, bone, and muscle.
  - Endoderm
    - Intestinal epithelium, Pancreatic, adrenal, and thyroid tissue.
- One germ cell layer may predominate → Monodermal
Most common Teratoma
Most common neoplasm of the fetus and newborn
Incidence of 1 in 20,000 to 40,000 live births
Female predominance from 2:1 to 4:1
17% have malignant histologic or clinical features.
10% of patients have a family history of twinning.
Coexisting anomalies in 10-20%
- Imperforate anus, anorectal stenosis, spina bifida, genitourinary malformations, meningomyelocele, and anencephaly.
- Structural abnormalities of juxtaposed organs from displacement by the teratoma.
Altman’s SCT Classification System

- **Type I (46.7%)**
  - external with minimal presacral extension.

- **Type II (34.7%)**
  - external with significant intrapelvic portion.

- **Type III (8.8%)**
  - primarily pelvic and abdominal, but apparent externally.

- **Type IV (9.8%)**
  - presacral, no external manifestation.
Malignancy correlates with:

- **Anatomic type**
  - 8% in type I versus 38% in type IV

- **Age at diagnosis**
  - 8% malignant dx at birth
  - 40-80% malignant dx older than 6 mo

- **Male gender**
  - Solid tumor > cystic

- **No correlation with size of the tumor.**

Malignant tissue:

- Yolk sac tumor or embryonal carcinoma.
Prenatal Diagnosis of SCT

- Diagnosed with Ultrasound - 22 and 34 weeks' gestation.
- Significantly less favorable prognosis than does diagnosis at birth.
- Mortality rate for SCT diagnosed in neonates is 5% vs that for fetal SCT is close to 50%.
- Hydrops or polyhydramnios and placentomegaly associated with fatal outcome.
- US findings
  - Mass arising from the sacral area of the fetus
  - Mass composed of solid and cystic areas, foci of calcification.
- Most prenatally diagnosed SCTs are extremely vascular and can be seen on color-flow Doppler.
Physical examination
- 90% of SCTs are noticed at delivery.
- Protruding mass extending from the coccygeal region.
- DRE to assess intrapelvic component

Intrapelvic tumors
- Delayed postnatal presentation
  - Infants and children between 4 months and 4 years of age.
- Clinical presentation
  - Symptoms from tumor compressing the bladder or rectum, constipation, anal stenosis.
  - Sacral defects and anorectal malformations
Postnatal Diagnosis of SCT

- Imaging studies
  - PXR
    - Sacral defects or tumor calcifications.
  - CT scan with IV and rectal contrast defines the
    - Intrapelvic extent of the tumor
    - Lymph node or distant metastases (liver)
    - Urinary tract displacement or obstruction
  - MRI
    - When spinal involvement is suspected or if the diagnosis is in doubt.
Differential Diagnosis of SCT

- Lumbosacral myelomeningocele
  - Similar findings on US.
    - Assessment of the integrity of the fetal spine - MRI
    - Assessment of lower extremity function.

- Other DDx
  - Neuroblastoma
  - Hemangioma
  - Leiomyoma
  - Lipoma
SCT- Operative Management

- The treatment of choice is complete surgical resection on an elective basis early in the newborn period.
  - Emergent surgery for tumor rupture or hemorrhage.
- Operative approach
  - Tumors with significant external component (type I and II)
    - Posterior sacral approach
  - Tumors with extensive intrapelvic component (type III and IV)
    - Abdominal approach
SCT - Surgical Resection - Posterior approach
SCT- Operative Management

- Operative goals
  - Complete tumor resection
  - Resection of the coccyx to prevent tumor recurrence
    - Recurrence rate as high as 37% if coccyx is not removed.
  - Reconstruction of the muscles of anorectal continence
  - Restoration of a normal perineal and gluteal appearance.
- Mortality - hemorrhagic shock.
- Morbidity
  - intraoperative hemorrhage
  - fecal incontinence
    - 30% to 40% of premature infants, large SCTs, levator muscles are severely attenuated.
Indicated when malignant component is identified.
- **Platinum-based chemotx** (cisplatin) most successful
- Other agents- etoposide, bleomycin.
- Survival rates -88% with local disease and 75% with distant metastases.

For unresectable primary malignant tumor, chemotherapy decreases size of tumor facilitating subsequent resection.

Localized malignant recurrence
- Complete resection with adjuvant chemotherapy.
- Radiation tx rarely given

Metastatic foci in the lungs and liver.
The long-term outcome in newborns with SCT is generally excellent.

**Age at diagnosis** - dominant prognostic factor.
- Diagnosis made before 2 months of age or excision is performed before 4 months of age, the malignancy rate is 5% to 10%.

**Cystic tumor** (mature) → better prognosis.

**Solid tumors** (immature) → hemorrhage, malignancy.

The Pediatric Oncology Group–Children's Cancer Group study (Marina et al, 1999)
- 80% event-free survival and 100% survival with complete resection alone in immature teratomas, even when microscopic foci of yolk sac tumor were present.

Children's Cancer Group study (Rescorla et al, 1998)
- 11% tumor recurrence rate with mature teratoma and a 4% recurrence rate with immature teratoma.
- Although 43% to 50% of these recurrences were malignant, the chemotherapy sensitivity of all tumors (mature and immature) is very high.
Close follow-up at 3-month intervals for 3 to 4 years is recommended.

- Physical exam
  - DRE- may detect a presacral recurrence

- Imaging if suspicious for local recurrence
  - Abdominal US
  - Abdominal CT scan or MRI

- Chest and Abdomen CT scan
  - R/o metastasis
Germ Cell Tumor
Tissues of 3 germ cell layers or monodermal
Treatment – Complete resection
Excellent prognosis
References

References

