Fetal Tumors

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Learning Objectives

- Describe the histologic types and biologic behavior of fetal tumors
- Recognize pertinent imaging features, which help to differentiate fetal tumors from the more common congenital malformations
- Understand the prognostic implications of each of the common tumor types

Disclosure

- Medical Editor Amirsys Content, Elsevier

Congenital Tumors

- Extracranial teratomas
- Neuroblastoma
- Soft tissue tumors
- Brain tumors
- Renal tumors
- Liver tumors
- Leukemia

Sacrococcygeal Teratoma

- Most common of the germ cell tumors
  - 1/35,000 – 1/40,000 live births
  - Female predominance 3:1
  - 10% have other abnormalities
    - Lower genitourinary or gastrointestinal
    - Pelvic girdle/lower extremity distortion with gait abnormalities
    - Malignant potential

- Type 1 – external
- Type 2 – external + internal extension
- Type 3 – external + internal into abdominal cavity
- Type 4 – completely internal
Type 1 – external
Type 2 – external + internal extension
Type 3 – external + internal into abdominal cavity
Type 4 – completely internal

Sacrococcygeal Teratoma
- Mortality rate newborn 5%
- Mortality rate fetus 50%
  - More common in solid tumors presenting < 30 wks
  - Can evaluate specific parameters
    - Tumor volume to fetal weight ratio (TFR)
    - Tumor volume to head volume ratio
    - Tumor morphology score
    - Tumor growth rate
    - Hydrops and placentomegaly

Sacrococcygeal Teratoma

- Mortality rate newborn 5%
- Mortality rate fetus 50%
  - More common in solid tumors presenting < 30 wks
- Causes
  - Hydrops (failure)
  - Arteriovenous shunting
  - Metabolic demands of large tumor
  - Tumor rupture or internal hemorrhage
  - Prematurity
- Mother can develop mirror syndrome
Size increased from 7 cm to 14 cm
Capsular breech
Severe polyhydramnios, fetal tachycardia, decreased fetal movement

Teratomas
- Head and neck
  - Thyrocervical, palate, nasopharynx
- Chest
  - Pericardium, mediastinum
- Abdomen
  - Retroperitoneum
EXIT Procedure
Ex Utero Intrapartum Treatment

- Partial delivery of fetus via caesarian section
- Placenta, umbilical cord remain intact
- Utero-placental gas exchange maintained
DDx Mediastinal Mass
- Normal thymus
- Don’t confuse with lung mass
  - Sequestration
  - Congenital pulmonary airway malformation

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Neuroblastoma
- Most common congenital malignancy
- 30% of fetal tumors
- Derives from primordial neural crest cells
- 90% occur in fetal adrenal
- Most diagnosed > 32 wks

Solid
- Echogenic
- May see Doppler flow
- Occasional stippled calcifications

Partially cystic
- Completely cystic
  - Probably involuting
  - Excellent prognosis

Favorable stage (I, II, IV-S)
- 67% Stage I
- Confined to adrenal
- 4% Stage II
- Beyond adrenal but not across midline
- 2% Stage III
- Extension across midline
- 5% Stage IV
- Distant metastases
- 22% Stage IV-S
- Mets to skin, liver, < 10% bone marrow

Neuroblastoma

- Blueberry muffin appearance

- 22% Stage IV-S
  - Mets to skin, liver, < 10% bone marrow

Excellent prognosis
- Most have favorable stage and biologic markers
- No MYC-N gene amplification

International Neuroblastoma Risk Group Staging System
- Low risk group (localized and 4s disease)
  - 70% of neonatal neuroblastoma
  - 95-100% 5-year survival
  - Conservative treatment approach
- Intermediate risk group (non-localized +/- mets)
  - 25% of neonatal tumors
  - 85-95% 5-year survival
- High risk group (above with MYC-N amplification)
  - 5% of neonatal neuroblastoma
  - 30-40% 5-year survival

Normal fetal adrenal medulla contains neuroblastic nodules
- Indistinguishable from neuroblastoma
- 100% of 2nd trimester fetal adrenals
- 0.5-2.5% of newborn adrenals

Neuroblastoma

- Metastases
  - Local invasion
  - Liver mets in 25% (IV, IV-S)
  - Discrete masses or diffusely infiltrating
  - Placenta
    - Fetal catecholamines enter maternal circulation
      - Preeclampsia
      - Headaches
    - 70% mortality rate

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Suprarenal Mass DDx
- Normal adrenal fakeout
- Extralobar sequestration
- Renal duplication
- Gastric duplication cyst
- Adrenal hemorrhage

Normal Adrenal Gland
- Potential Pitfall
  - Adrenal 20x larger than adult compared to body mass
  - Prominent echogenic medulla
  - Ice cream sandwich

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Soft Tissue Tumors

- Wide array of diverse tumors
- 12-22% of congenital tumors
- 2/3 benign but may be locally aggressive

Soft Tissue Tumors

- Fibrous connective tissue tumors
  - Fibromatosis
  - Myofibromatosis
  - Digital fibromatosis
  - Infantile fibrosarcoma

Soft Tissue Tumors

- Rhabdosarcoma
  - Extremities
  - GU tract
  - Face, neck
- PNECT
- Rhabdoid tumors
- Hemangiopericytoma

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Intracranial Tumors

- 10% of congenital tumors
- Most are supratentorial
- Histology
  - Teratoma (≈ 50%)
  - Astrocytoma
  - Craniopharyngioma
  - PNECT
  - Choroid plexus papilloma
  - Lipoma

Choroid Plexus Papilloma

- 5-9% of congenital brain tumors
- Lateral ventricle
- Lobular
- Hyperechoic
- Rapid onset hydrocephalus
- 73% survival rate
Lipoma

- 10% of congenital brain tumors
- Echogenic midline mass
- Agenesis of CC
- May extend into lateral ventricles
- Often asymptomatic
- Seldom resected

Mesoblastic Neprhoma

- 5% of tumors
- Benign mesenchymal tumor
- Homogeneous
- Well-defined
- Polyhydramnios ~70%
- Hypercalcemia, HTN
Liver Tumors
- 5% of congenital tumors
- Liver derived from both mesenchymal and endodermal tissue
  - Infantile hemangioma
  - Mesenchymal hamartoma
  - Hepatoblastoma
  - Metastases
    - Neuroblastoma
    - Leukemia

Benign Hepatic Vascular Mass
- Confusing terminology
  - Infantile hemangioma
  - Infantile hemangioendothelioma
  - Congenital endothelial hemangioma
  - Hepatic arteriovenous malformation
  - Vascular benign lesion of the liver
- Newer terminology
  - Rapid involuting congenital hemangioma (RICH)
  - Noninvoluting congenital hemangioma (NICH)

- Mixed echogenicity vascular masses
- May see significant vascular shunting
  - Hydrops
  - Kasabach-Merritt sequence
    - Hemolytic anemia, thrombocytopenia, consumptive coagulopathy
- May coexist with hemangiomas (10-15%); visceral or cutaneous
- Often grow in first 6 months of life followed by regression and involution (RICH)
Mesenchymal Hamartoma
- Loose mesenchyme with bile ducts
- Large fluid-filled cysts
- Avascular
- Surgery curative

Hepatoblastoma
- Most common liver malignancy
- Well-defined (pseudocapsule)
- “Spoke-wheel”
- Poor prognosis

Leukemia
- Hepatosplenomegaly
- Hydrops
- 15x increased risk with Down syndrome
- Cordocentesis

Leukemia
- WBC > 300,000 with 93% blasts
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