Extra-gonadal Teratomas

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Introduction

- Greek: ***teratos*** ("of the monster") and ***onkoma*** ("swelling")
- **fascinating tumors** owing to the diversity of tissues they contain
- **Primordial germ cell** is probably not the exclusive progenitor of a teratoma, but it is currently classified under GC tumors.
- Tumor sometimes consists of more organized tissue which some consider as a **parasitic twins**, but still classified teratoma by others
- Case reports from Ethiopia….

Contents lists available at ScienceDirect

Journal of Pediatric Surgery Case Reports

journal homepage: www.jpscasereports.com

Case report: A rare presentation of spinal teratoma in neonates: Two cases from Ethiopia

N. Tihitena, A. Mesay*

Addis Ababa University, College of Health Sciences, Ethiopia

International Journal of Reproductive Medicine & Gynecology

Case Report

A Case Report of a Rare Finding of Fetal Anterior Neck Mass - a

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Outline

- Teratomas

(general considerations)

  - Embryology
  - Pathology
  - Presentation
  - Management
  - Challenges in LMIC

1. Sacrococcygeal
2. Thoracic
   - Mediastinal
3. Abdominal
   - Retroperitoneal
4. Head and neck
   - Cervical
   - craniofacial

Teratomas

General considerations

Definition

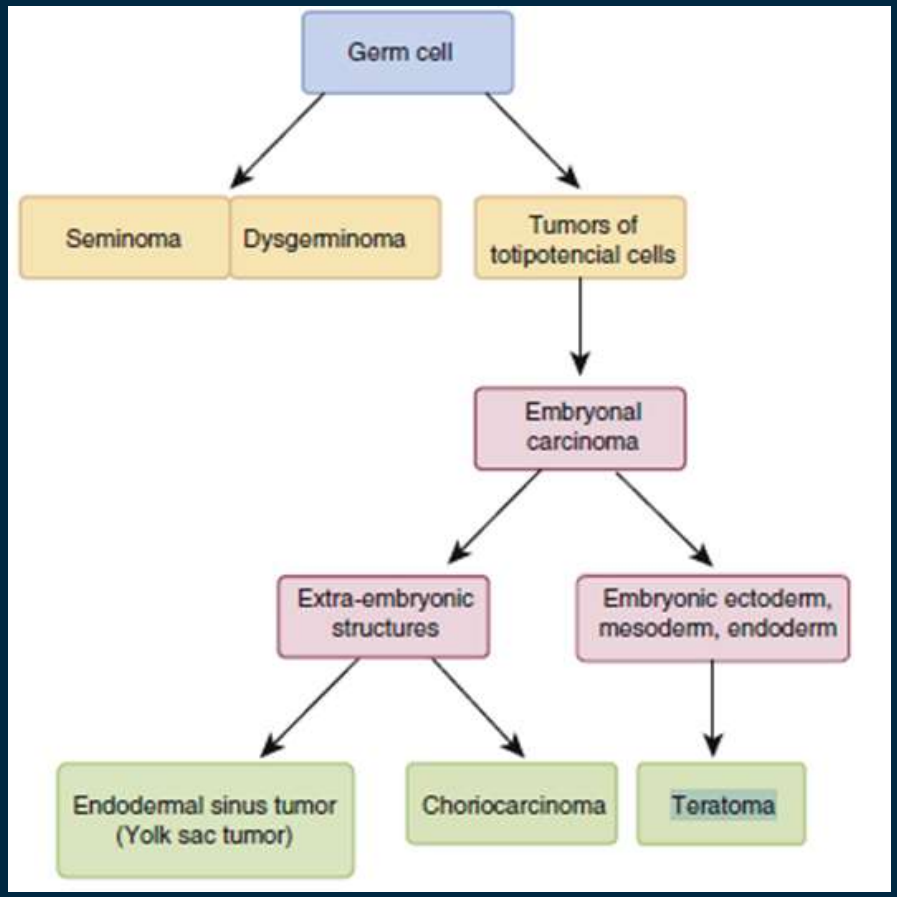
- classified by WHO as a histologic category of germ cell neoplasms
- embryonal neoplasms that contain at least 2 of the 3 germ layers
  - Ectodermal: (frequently present) hair, sebaceous/sweat gland, teeth
  - Mesodermal: (always present) fat, cartilage, bone and muscle.
  - Endodermal: intestine, cyst lined with epithelium, thyroid, neuroepithelial…
- *(*recent classifications also include monodermal types)*

Embryologic theories

- Totipotent primordial germ cell theory
  - arise from totipotent primordial germ cells
  - normally migrate to the gonadal ridges during wk4-5
  - miss their target destination and give teratoma anywhere in the midline
- Primitive node theory
  - arising from remnants of the primitive streak or primitive node.
  - This would explain the more common occurrence in sacrococcygeal
- Incomplete twinning theory

Classification

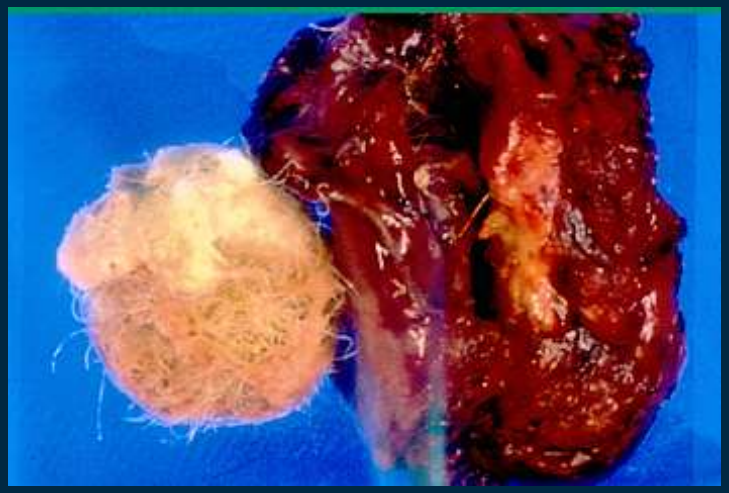
- mixed GCT types are often found within a single neoplasm, thereby making the detailed histological examination so important



Pathology

- Mature: differentiated tissues
  - little or no tendency to malignant degeneration
  - An apparently mature teratoma may recur several after years as a malignant YST
- Immature: incompletely differentiated, fetal tissue elements
  - May have microscopic foci of yolk sac tumor that may be missed.
  - favorable prognosis in newborn since immature elements may be appropriate for age
- Malignant: presence of malignant tissue elements
  - 20 % of tumors contain malignant components
  - Whether malignant from onset or replaced benign component is difficult to ascertain.

Pathology



- Mature teratoma: cystic mass filled with cheesy, sebum-like material and hair, forming a "hair-ball."

Presentation

- Tumor location correlates with the age
  - In children two-thirds of GCTs are located in extra-gonadal sites
- 75–80 % in females
- > 50 % evident at birth
- Associated congenital anomalies depends on the tumor site and size

Presentation

- Large tumors
  - Hydrops, placentomegally, polyhydraminos
  - Dystocia, tumor rupture
  - hyperkalemia from tumor necrosis
- Compressive symptoms (vary based on location)
- Symptoms related to malignancy
- Growing teratoma
  - enlargement of benign elements of teratoma
  - Initial resection and avoidance of chemotherapy in lack of definite malignancy

Labs

- Alpha-fetoprotein (AFP)
  - principle tumor marker, to assess residual or recurrent disease.
  - High level may indicate malignancy but is found in benign cases as well.
- β-hCG
  - indicates the presence of a component of choriocarcinoma
  - β-hCG can lead to precocious puberty.
- CA125

Treatment & Outcome

- Poor prognosis
  - prenatally detected teratomas (mortality rate 3x those diagnosed postnatally)
  - tumor location (head and neck involvement)
  - presence of hydrops (hyperdynamic state)
- adjuvant chemotherapy: Cisplatin-based chemo, survival> 80–90%
  - for higher grade immature, metastatic, incomplete resection, LN involvement
  - Immature elements: recommend the ongoing surveillance, no adjuvant chemo
- Biopsy followed by chemo before resection: Older patients with large malignant tumors

Challenge in low income countries

- **Lack of prenatal diagnosis** leading to Obstetric complications
- **Delayed postnatal treatment** due to
  - lack of recognition of the symptoms
  - lack financial resources to travel to an appropriate hospital
  - overwhelming volume of patients in referral hospitals
  - fear of surgical treatment and seek traditional healers instead
  - Sometimes these children are considered monsters and left to die at home
- **Availability / affordability of facilities**
  - imaging (ultrasound, CT) and pathology
  - TPN and mechanical ventilation
  - Chemotherapy
- **Surgery by inadequately trained personnel**, leading to malignant recurrence
- **Lack of anesthesiologist** make surgery in neonate challenging
- **difficulties in follow-up** when patients come from remote areas

1. Sacrococcygeal Teratomas

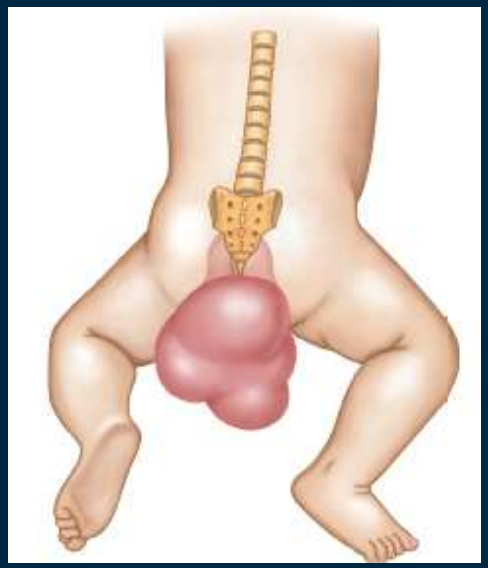
Epidemiology

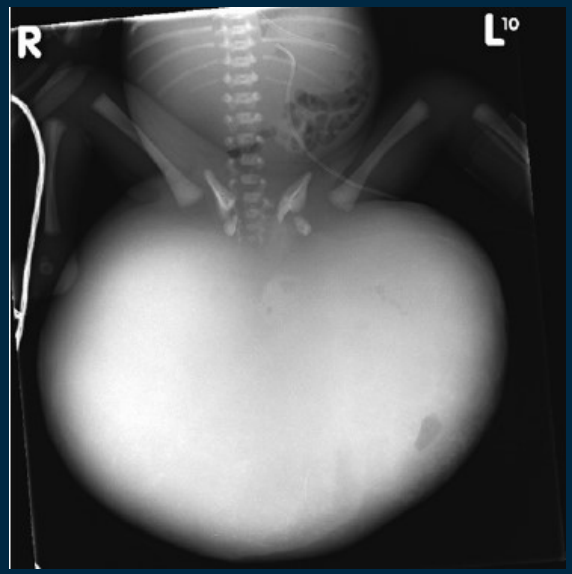
- 1:20,000 to 1:40,000 live births
- F:M = 3:1 (1.5:1 in Currarino triad)
- Most common tumor in newborns
- most common extragonadal location irrespective of age
- 35–60% of teratomas (gonadal included)

Associated anomalies

- Most are sporadic with a low recurrence risk
  - 10-20% have ass anomalies. sacrum, vertebrae and GI or urogenital tracts
- familial teratoma with an autosomal dominant mode
  - partial sacral agenesis with intact first sacral vertebrae.
  - Currarino triad (presacral teratoma, ARM, sacral defect)
  - urogenital malformations

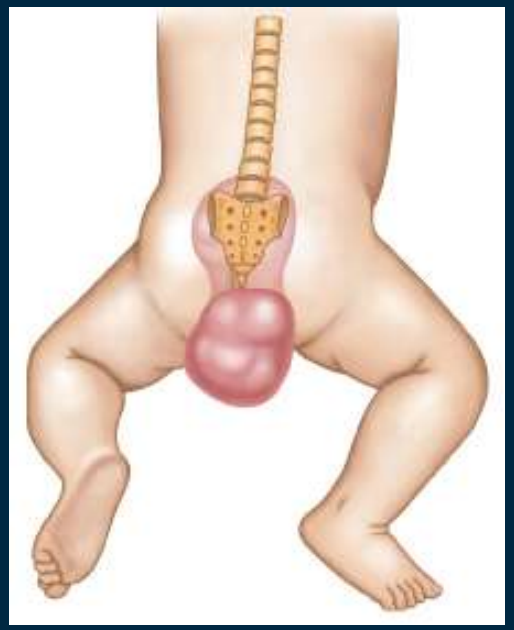
Classification





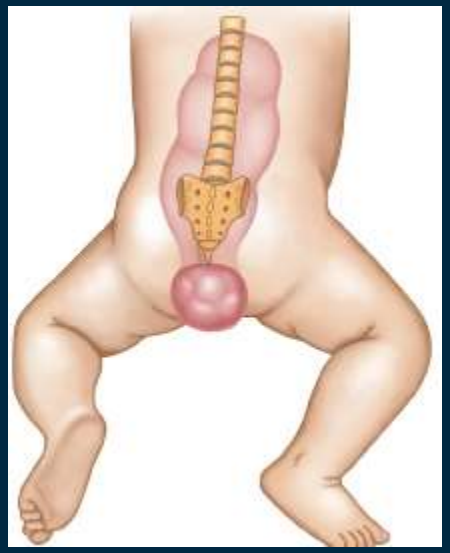
- Type I (46.7 %) - predominantly external with minimal presacral extension.

Classification



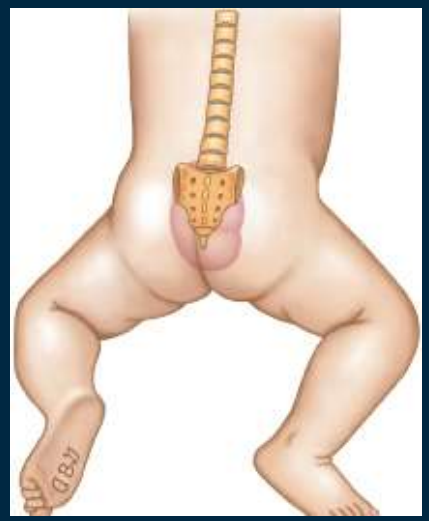
- Type II (34.7 %) arises externally and has a significant intrapelvic component.

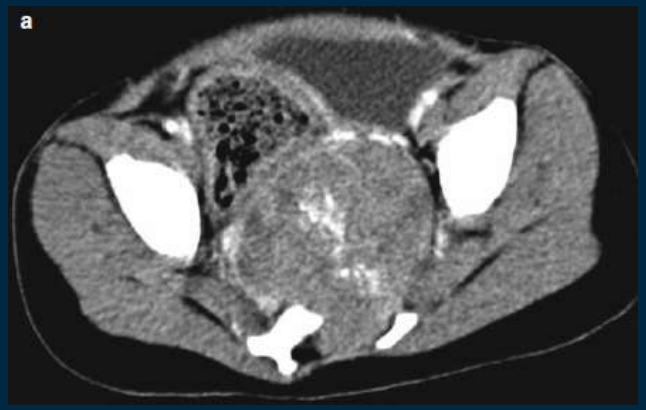
Classification



- Type III (8.8 %) is primarily pelvic and abdominal but is apparent externally

Classification





- Type IV (9.8 %) is presacral and has no external manifestation. Tend to present at a later age in a malignant form

Pathology

- Can grow at an unpredictable rate to tremendous dimensions
- Tend to be benign
- Malignancy
  - Age: less than 10% at birth but rises to 40–75% after age 1 year
  - Sex: more frequent in males
  - Tumor type: more in solid tumors than complex and cystic
  - anatomic type: 8 % in type I and 38 % in type IV

Imaging

- Lumbosacral myelomeningocele and cystic SCT may show similar fi ndings on US.
  - meningoceles occur cephalad to the sacrum.
  - gentle pressure on a sacral meningocele reveals bulging of the fontanelle
- Other critical information gained from US includes
  - presence of abdominal or pelvic extension
  - bowel or urinary tract obstruction,
  - assessment of the integrity of the fetal spine
  - documentation of fetal lower extremity function
- Radiographs of the pelvis identify any sacral defects or tumor calcifications.
- CT with intravenous and rectal contrast material defines the intrapelvic extent. liver metastasis and periaortic lymph node enlargement.
- MRI is useful when spinal involvement is suspected if the diagnosis is in doubt.
- chest radiograph is useful in revealing obvious pulmonary metastases

Prenatal intervention

- percutaneous aspiration - Purely cystic teratomas occur in 10–15% of cases.
- intrauterine endoscopic laser ablation of the large feeding arteries has been described
- open fetal surgery – progressive hydrops and high-output cardiac failure
  - surgery prior to 27–32 weeks'
  - procedure is not designed to completely remove the teratoma, require a second operation postnatally

Delivery

- Early delivery : rapid tumor growth, hemorrhage, ominous changes in Doppler, progression of placentomegaly or polyhydramnios
- Immediate delivery – polyhydramnios and placentomegaly, with associated mirror syndrome (resemble preeclampsia -threat to mother)
- planned cesarean section delivery tumors larger than 5 cm

Delivery

- Obstetric complications: ruptured SCT

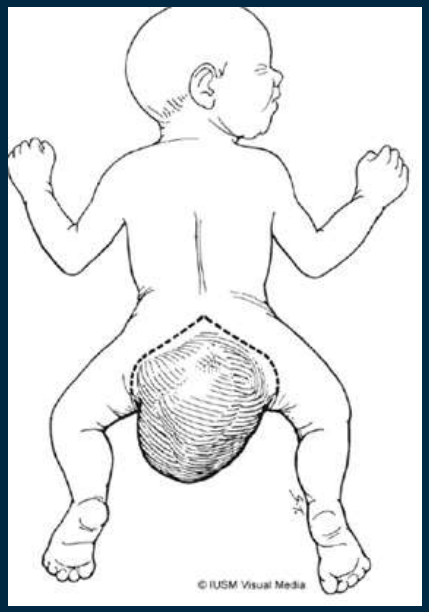
Postnatal intervention

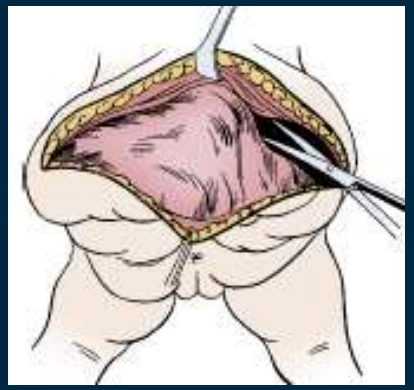
- Operative resection is usually carried out in the first week of life.
  - risk of malignant transformation
  - infection in the presacral cystic mass, which can diffuse to CSF (meningtis)
- Devascularization and staged resection may be considered for premature to avoid excessive blood loss.

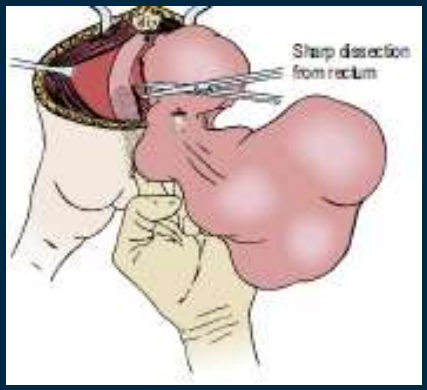
Surgery

- Approach
  - posterior sacral (Prone jackknife position) for type 1&2
  - abdominal for type 3&4
- Tumor excision
  - Raise skin flap and mobilize mass close to its capsule
- Resection of coccyx (37% recurrence if coccyx not removed)
  - control of the middle sacral or branches of hypogastric (after removing coccyx)
- Reconstruction of muscles for anal continence
- Restoration of normal perineal and gluteal appearance

Surgery

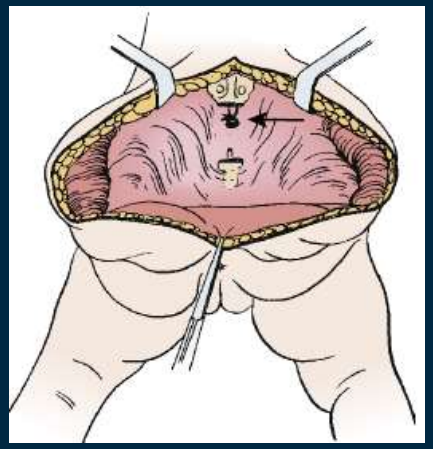


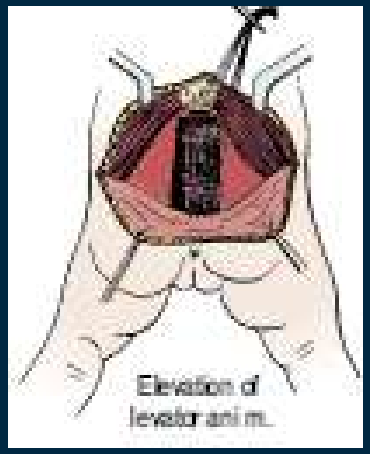


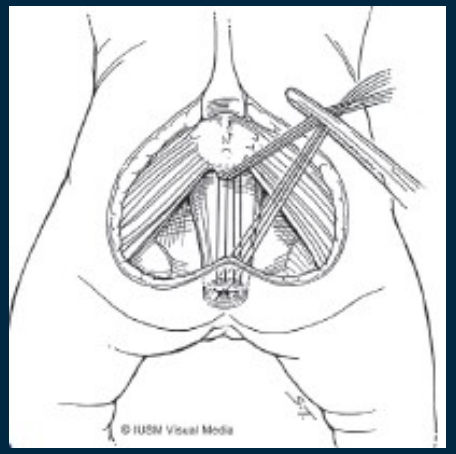


- Excision via inverted chevron incision: good exposure, keeps wound away from anal orfice. tumor excised along with coccyx taking care not to injure rectum

Surgery

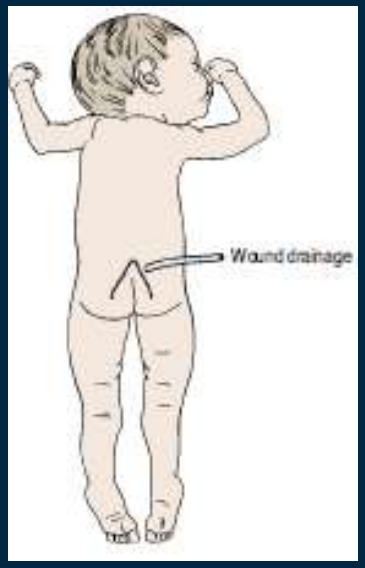






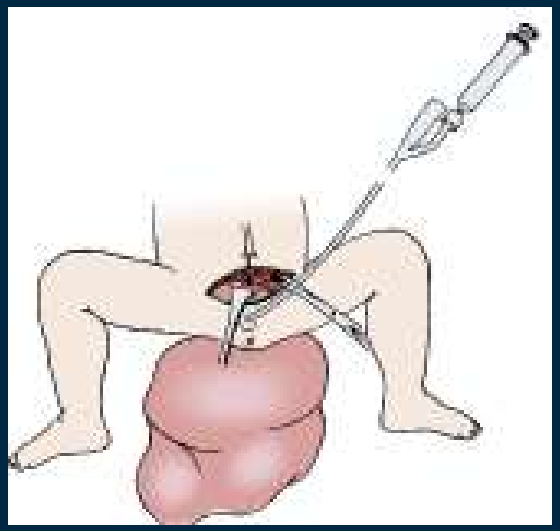
- Excision via chevron incision: Middle sacral artery ligated after removing coccyx. Placement of sutures between the anal sphincter and the presacral fascia and When the sutures are tied, the anal sphincter is pulled upward to the sacrum to form a gluteal crease.

Surgery



- Excision via chevron incision: Skin closure may be vertical or transverse closure. Rather than leaving “dog ears”, excess skin can be de-epidermized and buried to improve buttock contour

Surgery



- If tumor extends into the retroperitoneum, a urinary bladder catheter is inserted to facilitate suprapubic dissection. Lower abdominal transverse incision allows interruption of the middle sacral artery and dissection of the tumor from the sacrum and pelvis, which is eventually removed from the perineum.

Surgery

- Excision via a midline gluteal cleft incision: for patients with curarino syndrome posterior sagittal approach may be used

Complications

- Intraoperative hemorrhage
  - major cause of mortality is hemorrhagic shock.
  - tightly wrap the teratoma with an elastic bandage
  - emergent laparotomy and temporarily cross-clamping the distal abdominal aorta has been reported
- Wound complications
- Urinary and fecal incontinence / retention
  - Temporary or persistent
  - 40 % of premature infants with large SCTs and in whom the levator and gluteal muscles are severely attenuated
- Limb weakness

Adjuvant chemo

- completely excision of malignant does not require adjuvant therapy.
- For localized malignant recurrence, complete resection remains the cornerstone of salvage therapy
- Good survival after chemo (88 % with local and 75 % with metastases)
- Residual malignant YST lesions must be excised.

Follow up

- AFP levels and rectal examination every 3 months to 3 years of age.
  - AFP should return to normal by 9 months of age
- Recurrence
  - 11 % tumor recurrence with mature teratoma
  - 4 % recurrence with immature teratoma have been reported
  - 43–50 % of these occurrences are malignant

Prognosis

- Factors
  - prematurity and perinatal complications,
  - presence of malignancy (age and completeness of resection)
- mortality rate
  - 5 % diagnosis in neonate
  - 50 % for diagnosis in fetus (better prognosis if diagnosis was incidental)

2. Intrathoracic Teratomas

Mediastinal Teratomas

- Second most common extragonadal site for teratomas in children
- 7–10 % of all teratomas are mediastinal
- Commonly anterior mediastinum, cystic, associated Klinefelter syndrome
- orthopnea or a reduction in the tracheal diameter >50% on axial imaging is at a significant risk for airway collapse during general anesthesia.
- Immature elements are not of prognostic significance in children under 15 years, but associated with local and distant metastases above that age

Mediastinal teratoma

- Anterior mediastinal mass extending into the neck

Other thoracic Teratomas

- Intracardiac
  - From atrium or ventricle
- intrapulmonary teratomas
  - Few cases reported
- Intrapericardial
  - leading cause of massive pericardial effusion in the neonate

3. Abdominal Teratomas

Retroperitoneal Teratomas

- Third most common extragonadal site
- 4% of all childhood teratomas
- 50 % are identified in the first year of life
- F:M (2:1)
- CT or MR helps differentiate from neuroblastoma or Wilms tumor
- 24 % found to be malignant
- Operative excision is usually straight forward but occasionally the tumor may encase major vessels

Gastrointestinal Teratomas

- quite rare
- majority are gastric (1–2.5 % of all teratomas)
- mass effect and Intramural components may ulcerate or hemorrhage,
- GI teratomas are not felt to have the potential for malignant degeneration
- Recurrence following resection is quite rare
- Other rare sites of abdominal teratomas include liver, gallbadder, pancreas, kidney, intestine, bladder, prostate, uterus, mesentery, omentum, abdominal wall, and diaphragm.

4. Head and neck Teratomas

Head and Neck Teratomas

- 10 % of teratomas.
- no sex predilection.

Cervical Teratomas

- 8% of teratomas
- frequently cause significant airway and esophageal Obstruction
- cystic lymphatic malformations are the most likely differential
- Complete excision is accomplished through a wide collar incision.
  - often have a pseudocapsule, not difficult to separate from the strap muscles
  - pretracheal fascia is sometimes very adherent.
  - which facilitates gentle elevation of the tumor out of the neck.
  - If arise from thyroid gland, involved lobe is excised

Cervical teratoma

- **Large cervical teratoma**

Craniofacial teratoma

- Epiganthus
  - Protruding from the mouth (arise from the soft or hard palate)
  - An EXIT procedure is often required. Surgical excision is mandatory
  - high degree of organization often gives appearance of a parasitic fetus
- Nasopharyngeal
  - Some lesions may also extend intracranially
  - Additional craniofacial anomalies, such as cleft palate and mandibular deformity, can result from the growth of such tumors
- Other- intracranial, Oropharyngeal, orbit, middle ear, skull

Craniofacial teratoma

- **Large Epignathus :** Baby delivered by EXIT using tracheostomy and mass excised. Baby is on mandible molding device

Head and neck teratoma

- **EXIT (**Extrauterine intrapartum treatment**) :** Baby partially delivered by C/S but remains attached to the placenta, while the surgeon establishes an airway. The delivery is then completed. *(Originally developed to reverse tracheal occlusion done for CDH)*

Miscellaneous teratoma sites

- Skin
- Parotid
- Vulva
- Perianal region (away from the coccyx)
- spinal canal
- umbilical cord (possibly associated with omphalocele)
- Placenta

References

- Hollcomb and Aschcraft Pediatric surgery 7th edition, 2020
- Surgery of childhood tumors 3rd edition, 2016