Retroperitoneum

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Objectives

• Describe the Different tumor Groups
• Discuss role of imaging
• Develop an approach to managing retroperitoneal masses

Non-Neoplastic

• Retroperitoneal Fibrosis
• Extramedullary hematopoiesis
• Fluid
  – Pseudocyst
  – Abscess
  – Urinoma

Primary Neoplasms

• Originate outside Major Organs
• Rare and Diverse
• Bulk typically creates symptoms
• 80% Malignant

Tumor Classification
( > 100 Tumor Types)

• Mesodermal
• Neurogenic
• Germ Cell

Primary Tumor Classification

• Mesodermal
  – Lipoma/sarcoma
  – Leiomyoma/sarcoma
  – Malignant Fibrous Histiocytoma
  – Lymphangioma
  – Angiosarcoma
  – Hemangiopericytoma
Primary Tumor Classification

- Neurogenic
  - Paraganglioma
  - Sympathetic
    - Ganglioneuroma
    - Ganglioneuroblastoma
    - Neuroblastoma
- Nerve Sheath
  - Neurofibroma
  - Schwannoma

Primary Tumor Classification

- Germ Cell
  - Teratoma
  - Malignant Germ Cell Tumor

Aspiration

- Definitively Localize
- Determine Invasion and Spread
- Characterize specific Tumor Type

Fundamentals

- Confirm Location
- Organ of Origin?
- Density and Signal
- Specific tumor components
- Enhancement
- Pattern of Spread

Retroperitoneal Fibrosis

- Histo: Replacement by Fibrotic tissue and Inflammatory cells
- Encases Aorta and to a lesser degree the IVC
- Typical distribution: below Kidneys to pelvic brim
- Occasionally Extends into chest and along mesenteric vessels

Retroperitoneal Fibrosis

- Etiology
  - 2/3 idiopathic (Ormand’s disease)
  - 1/3 Secondary
    - Methysergide toxicity
    - Aortic aneurysm
    - Surgery
    - Hemorrhage
    - Inflammatory bowel disease
    - Collagen vascular disease
    - Radiation/surgery
    - Fibrosing conditions elsewhere
**Retroperitoneal Fibrosis**

- IVP, retrogrades
  - Medial deviation of mid ureters
  - Hydronephrosis

**Retroperitoneal fibrosis**

- CT
  - Mantle of soft tissue from renal hilum to iliacs
  - Laterally bounded by psoas
  - Variable enhancement
  - Aorta is encased but not deviated

**Retroperitoneal fibrosis**

- MR
  - Fibrotic phase
    - Low on T1 and T2
    - No enhancement
    - Highly Specific
  - Active phase
    - High on T2
    - Enhancement
    - Can not rule out malignancy
    - Must biopsy

**Treatment**

- Medical
  - Immunosuppression
  - Steroids
- Non Medical
  - Stents
  - Surgery
**Neurogenic Tumors**

- < 1/4th of RP malignancies
- Paraspinal masses
- Tubular
- Smooth or mildly lobular
- Typically benign
- Rapid growth, pain, infiltrative margins suggest malignancy

**Neurogenic Tumors Myxoid Matrix**

- Low density on CT
- Low signal on T1WI
- Myxoid matrix prolongs T2 (Bright)
- May have calcifications

**Nerve Sheath Tumors**

- Often psoas or para-psoas mass
- Assess neuroforamen
- Consider Neurofibromatosis if multiple

**T2 Target Sign**

**NF and PET FDG**

25 year old female with disseminated neurofibromatosis.

Multiple foci of moderate uptake of FDG are seen in neurofibromatosis lesions throughout the body.

**Sympathetic Origin**

- Form from primitive neural crest cells
- Sympathoblast
  - Neuroblastoma
  - Ganglioneuroblastoma
  - Ganglioneuroma
**Ganglioneuroma**
- Benign
- More common in mediastinum
- Generally asymptomatic
- Elongated low-density masses
- May be hyperintense on T2WI

**Paraganglioma (Extra-adrenal pheochromocytoma)**
- \(\simeq \) 10% of pheochromocytomas
- Majority (60-80%) symptomatic due to catecholamine excess
- More Malignant potential than pheochromocytomas (as high as 36% in one study)

**Paraganglioma**
- Organs of Zuckerkandl
- CT non-specific
  - Enhance avidly
  - Contrast safe but "contraindicated"
- High T2 signal helpful
- MIBG positive

**Paraganglia**
- Organs of Zuckerkandl
- CT non-specific
  - Enhance avidly
  - Contrast contraindicated
- High signal on T2 is suggestive but not universally seen
- Uptake on MIBG scan
Primary Tumor Classification

- Mesodermal
  - Lipoma/sarcoma
  - Leiomyoma/sarcoma
  - Malignant Fibrous Histiocytoma
  - Lymphangioma
  - Angiosarcoma
  - Hemangiopericytoma

Liposarcoma

- Most common Primary Retroperitoneal tumor
- 85% have Fat detected by CT or MR (i.e. 15% have no detectable fat)
- Well-differentiated, de-Differentiated Pleomorphic, Myxoid
- Malignant from inception

Liposarcoma

- Clinical presentation
  - Often grow slow, present late
  - Pain is most common complaint
- Infiltrative margins
- Complete excision usually impossible
- Local recurrence common (nearly 100%)
- Nodes rare.

Well Differentiated

- Local, contiguous growth
- No ability to Metastasize
- Challenge to resect
- Often recur

De-Differentiated

- Non-lipogenic transformation of well-differentiated tumor
- 75% occur within retroperitoneum due to ability to evade detection
- 15% at 7-8 years
- More aggressive
- Ability to metastasize
Dedifferentiated Liposarcoma
- More common in extremity, esp. deep muscles of thigh and buttocks
- Rare in retroperitoneum
- Note Well--Myxoid Material may be found in well differentiated tumors

Myxoid Liposarcoma
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Pleomorphic Liposarcoma
- Equal distribution in Extremity and Retroperitoneum
- Least common subtype overall
- Extensive adipogenic areas

Hibernoma
- Benign rare tumor of brown fat
- Thermoregulatory function
- Multiple mitochondria which accumulates excessive FDG
- Imaging suggestive but not specific
- Amenable to biopsy

Leiomyosarcoma
- More common than benign counterpart
- Necrosis is a clue
- May have intravascular invasion
- Often not separable from IVC

Malignant Fibrous Histiocytoma (MFH)
- Generally large masses
- Most common adult sarcoma-15% Abd/Retroperitoneal
- Necrosis uncommon
- T2WI helpful – “Bowl of fruit sign”
  - Mosaic of low and high signal
    - Fibrous tissue – hypointense
    - Soft tissue - intermediate
    - Myxoid stroma – hyperintense
Hyperintense T2WI
Myxoid Matrix

- Neurogenic Tumors
- Malignant Fibrous Histiocytoma
- Myxoid Liposarcoma

Lymphangioma

- Benign
- Fluid/Chyle-filled
- Multilocular
- Insinuates itself and may not respect intrinsic boundaries
- Can be huge

Germ Cell Tumors

- Teratoma
  - Mature
  - Immature
- Malignant Germ Cell Tumors

Teratoma

- Embryonal neoplasms with all 3 germ cell layers
- Most (80%) are mature (benign) and are cured by surgery
- Children (less than 6 months) and young adults (15-25 years)
- Female: male = 4:1

Teratoma

- Triad of
  - Fat (sebum or adipose tissue)
  - Fluid/Cystic
  - Calcification (may be clump-like)
Fat containing retroperitoneal masses:
- Liposarcoma: large, heterogeneous
- Teratoma: young patients, calcification, cystic area
- Myelolipoma: usually arising from adrenal
- Angiomyolipoma: arises from kidney, but attachment may be hard to find

Extra Gonadal GCT
- Very Rare
- Implies NO primary malignancy
- Evaluate ovary and testicle to r/o metastatic source