Retroperitoneal Sarcoma

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1mc. Liposarcoma - well diff/ De diff.
2mc. LMS
3mc. malignant Solitary fibrous tumor
4 - Undiff. pleomorphic sarcoma

- Diagnosis & staging:

* MRI vs CT AFP → preferred
  fat density to be compared w/ abdominal wall fat.

↓

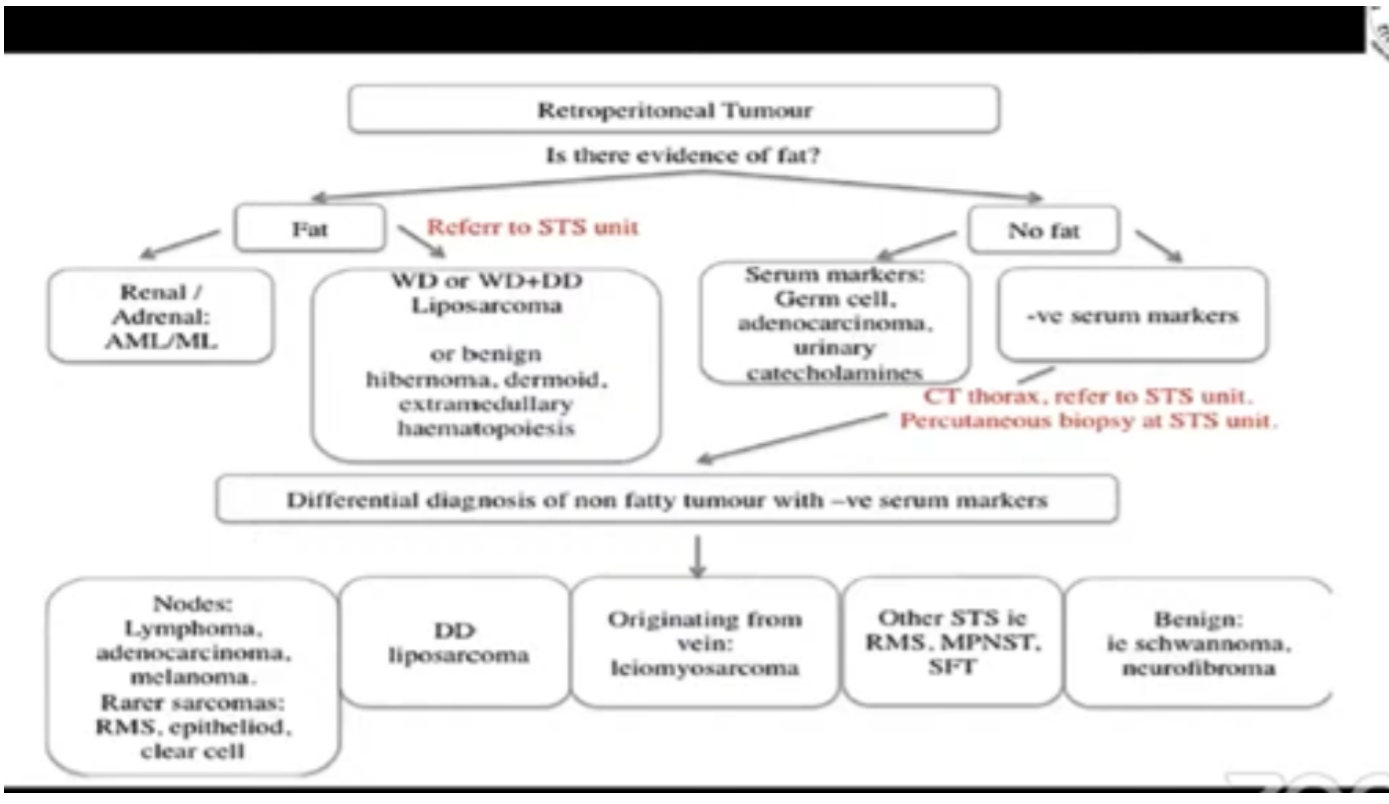
high motion artefacts in abdomen d/t increased sequence time → For tumors occupying pelvis / close to vertebra. / intravenival tumor thrombus.

* CT chest - staging. especially Lms.

* Role of PET-CT.

. indicated for staging in
  - GIST
  - Ewings.

Otherwise not indicated.



Source: IASO masterclass by Dr. Shraddha Patkar on YouTube

* Role of pre-op biopsy:

- When imaging is uncertain of pathology (young individual w/ emings suspicion)
- When suspecting lymphoma.
- Central (aortocaval) tumor.

Avoid transperitoneal Bx.

* No role of FNAC
* Core needle bx - 6 cores
  - 14/16 G needle.
  - posterior/lateral approach w/ co-axial technique.

Risk of needle track seeding < 2%

- IHC markers for sarcoma:

Liposarcoma - MDM 2

LMS - SMA, Desmin.

RP mass - DD:

- RP sarcoma.
- mass from Kidney, Pancreas. adrenal, duodenum
- metastatic nodal mass
- Lymphoma.
- AGT from undescended testis.

TYPES OF RP TUMORS

Tissue of Origin	Benign	Malignant
Mesodermal		
Adipose	Lipoma	Liposarcoma
Smooth muscle	Leiomyoma	Leiomyosarcoma
Striated muscle	Rhabdomyoma	Rhabdomyosarcoma
Connective	Fibroma	Fibrosarcoma
Lymphatic vessels	Lymphangioma	Lymphangiosarcoma
Primitive mesenchyme	Myxoma	Myxosarcoma
Vessels	Hemangioma	Angiosarcoma
	Benign hemangiopericytoma	Malignant hemangiopericytoma
Histiocytic	Benign fibrous histiocytoma	Malignant fibrous histiocytoma
Uncertain origin	Xanthogranuloma	
Nervous		
Nerve fiber	Neurofibroma	Neurofibrosarcoma
	Benign neurilemoma	Malignant neurilemoma
Sympathetic nervous system	Benign ganglioneuroma	Malignant ganglioneuroma
	Sympathicoblastoma	
	Benign neuroblastoma	Malignant neuroblastoma
	Chemodectoma	
	Ependymoma extra-adrenal	
Heterotopic adrenal cortical tissue and chromaffin tissue		Carcinoma of cortical tissue Malignant non-chromaffin paraganglioma
Embryonic rests and notochord		
	Benign teratomas	Malignant teratomas
	Chordomas	

Source: IASO masterclass by Dr. Shraddha Patkar on YouTube

- Liposarcoma
  - mc
  - ubiquitous occurance
  - worse prognosis in RP compared to extremity

- Leiomyosarcoma (LMS)
  - 2mc
  - 50% metastatic at presentation
  - arise from major vessel - mc from IVC

* Prognostic factors
  - Complete microscopic excision - No role for frozen section
  - Tumor grade
  - multifocality.
  - Histological subtype.

→ Surgery in RP sarcoma :

- Unresectability in RP mass :
  1) B/L kidney involvement — Auto transplant kidney after resection.
  2) Diffuse sarcomatosis.
  3) Greater vessel encasement — Aorta encasement unresectable.
  4) Root of mesentery / SMA / celiac axis involvement
  5) Spinal cord involvement.

IVC reconstruction | infrarenal ligation can be done. Bowel drains through portal V and lower limbs develop collaterals.

* multivisceral resection decreases LR, but no outcome on OS.

- Compartment excision :
  - En bloc removal of the mass along with adjacent organs irrespective of involvement by tumor.
    * 3x decrease in LR of 10% at 3 years
  - By Sylvie Bonvalot & Gronchi.
  - Drawbacks — increased morbidity; No clear ur anatomical boundaries in RP; No survival benefit.

Important to avoid R2 resection

- Concept of Histopathological organ invasion (HOI)

- By Fairweather.
- The histological subtype influences the likelihood of local invasion, distant mets & guide multivisceral resection.

  eg: LMS has high risc of Distant mets, hence a less aggressive approach to reseet primary tumor.

Classification for Rationale for organ resection

1- suspected organ invasion
2- tumor involving end organ vascular supply
3- encasement of organ
4- adherent to organ
5- Adjacent to organ, required for R0/R1 resection
6- Iatrogenic injury to organ. requiring resection.

- Role of Radiotherapy in RP sarcoma:

1. STRASS trial - Preop RT + Sx vs Sx alone:

   - small subgroup of LPS showed benefit.
   - overall study showed No benefit for NART.

Scientific rationale for Preop RT

PRE-OPERATIVE RT (NART)	POST-OPERATIVE RT
• Tumor (Target) is clearly demarcated	• Can select patients based on grade and margin status
• Displaces radiosensitive organs	• Radiosensitive organs will move into the tumor bed
• Lower equivalent therapeutic dose	
• Shorter course of treatment	
• Treatment delivery not impacted by post op healing issues	
• Potential down-staging	

Source: IASO masterclass by Dr. Shraddha Patkar on YouTube

- IMRT | IGRT to minimise adverse effects.

2. Intra operative RT (IORT)

- study from Massachusetts General – slight benefit.
- difficult to give RT to wide area

3. Adjuvant RT (60 - 66 Gy | 1.8 - 2 Gy per #)

- SEER data - contradictory.
- Radiation induced enteritis
- doubtful areas of margin / stalk of the lesion to be marked ☆

Radiopaque clips which can be irradiated.

- Role of Chemotherapy in RP STS :

* Adjuvant chemotherapy:
  - for high grade, chemosensitive histology.
  - Not recommended for low grade tumors.

* Neoadjuvant chemo:

STRASS - II trial – ongoing

- Upfront Sx vs NACT f/b Sx
  - only for high grade LPS
  - EpiADM regime.

- management of Recurrence:
  - Sx wherever resectable.
  - Candidate for metastatectomy (mc-lung)
    - Long DFS
    - Good PS
    - oligometastatic