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CLINICAL SURGICAL CASE MANUAL

A Comprehensive Treatise for Students, Teachers and Practitioners



2nd Edition

Kanchana Sundaramurthy

JAYPEE

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CHAPTER 3

Soft Tissue Sarcoma

TYPES

- Fibrosarcoma.
- Malignant fibrous histiocytoma.
- Liposarcoma.
- Rhabdomyosarcoma.
- Kaposi's sarcoma.
- Lymphangiosarcoma.
- Synovial sarcoma.
- Mesothelioma.
- Malignant schwannoma.
- Neuroblastoma.
- Malignant paraganglioma.
- Extraskeletal osteosarcoma.
- Malignant mesenchymoma.
- Epithelioid sarcoma.

SITE

- Lower limb: 46%.
- Retroperitoneum, trunk and mediastinum: 32%.
- Upper extremity: 13%.
- Head and neck: 9%.
- Synovial sarcoma can present even in muscle plane where synovial elements are present; it need not be close to joint.

INCIDENCE

- Most common soft tissue sarcoma (STS) in adults: Malignant fibrous histiocytoma.
- Most common STS in children: Rhabdomyosarcoma.
- Most common STS in retroperitoneum: Liposarcoma.

LYMPH NODAL SPREAD IN STS

S: Synoviosarcoma.
C: Clear cell variety of rhabdomyosarcoma.
R: Reticulum cell sarcoma.
E: Epithelioid sarcoma.
A: Angiosarcoma.
M: Malignant fibrous histiocytoma.

Differential Diagnosis from Carcinoma

Sarcoma	*Carcinoma*
Mesodermal	Epidermal
Predominantly blood spread	Lymphatic and blood spread
Osteosarcoma in young age; rest in elderly	Elderly
Prognosis worse than carcinoma	-
Rapid increase in size	Slow or rapid
Vascular (may mimic inflammatory swelling)	-

PREDISPOSING FACTORS

- Soft tissue sarcoma (STS) mostly occurs de novo and rarely results from malignant degeneration of pre-existing benign tumors.
- Genetic:
  - Li-Fraumeni syndrome:
    - » STS.
    - » Breast carcinoma.
    - » Brain carcinoma.
    - » Leukemia.
    - » Adrenal cortical tumor.

  - von Recklinghausen's disease:
    - » Neurofibroma (of nerve sheath).
    - » Café-au-lait spots.
  - Retinoblastoma.
  - Gardner's syndrome: Familial adenomatous polyposis with desmoid tumor.
  - Tumor suppressor genes *p53, RB1*.
- Radiation:
  - Post-irradiation, especially radiation therapy (RT) to breast or post-mastectomy area giving rise to fibrosarcoma of chest wall.
  - Ortho- and mega-voltage therapeutic radiation.
- Lymphedema:
  - Lymphangiosarcoma from chronic lymphedema: Stewart-Treves syndrome (poor prognosis).
  - Postsurgical.
  - Postradiation.
  - Filarial.
- Trauma:
  - Post-parturition.
  - Extremity trauma.
- Chemical:
  - Exposure to phenoxyacetic acid in herbicides and chlorophenols in wood preservatives.
  - 2,3,7,8, tetrachlorodibnezodioxin (TCDD).
  - Polyvinyl chloride.
  - Hemochromatosis.
  - Arsenic.
- Multiple enchondromas.
- Immunodeficiency: HIV-associated Kaposi's sarcoma.
- Fibrosarcoma from Marjolin's ulcer and other chronic ulcers like venous ulcers, thermal and acid burns, etc.
  - Squamous cell carcinoma is more common.
- Bone sarcomas in abnormal bone, e.g. Paget's.
- Miscellaneous:
  - Chromosomal disorders, especially Ewing's, synovial sarcoma, liposarcoma, etc.

CLINICAL FEATURES

- Swelling.
- Neurovascular deficit:
  - Referred pain.
  - Weakness.
  - Sensory impairment.
  - Pressure on vein causing edema.
- Vascular deficit is more in the veins than arteries because adventitia and thick arterial wall resist infiltration to some extent.
- Joint impairment:
  - In proximal and distal joints.
  - More due to neurological deficit rather than joint infiltration.
- Warmth, dilated veins over the swelling.
- Bone infiltration with local pathological fracture.
- Regional lymph node enlargement (SCREAM).
- Lung and bony secondary (bone—not very common).
- Variable consistency (cystic to hard).
- Head and neck STS may present with cranial nerve deficits.

Microscopic Features

- Compression zone of condensed and atrophic tissue immediately surrounding the STS.
- Edematous and neovascularized zone surrounding compression zone—most evident in rapidly growing STS, but present around all STS.
- These two zones form the pseudocapsule.
- Sarcoma expansion is stopped by:
  - Fascial boundaries.
  - Periosteal structures.
  - Vessel adventitia.
  - Nerve sheaths.

GRADING

- Grade I: Low grade.
- Grade II: Moderate grade.
- Grade III: High grade.
- Based on:
  - Degree of differentiation.
    - » Grade I: 75% differentiated.
    - » Grade II: 50% differentiated.
    - » Grade III: 25% differentiated.
    - » Grade IV: Undifferentiated.
  - Presence of necrosis.
  - Mitotic activity/Aneuploidy.
  - Cellularity.
  - Anaplasia/Pleomorphism.
  - Expansive/Infiltrative growth.
- High grade:
  - Rhabdomyosarcoma.
  - Extraskeletal Ewing's.
  - Synovial sarcoma.
  - Epithelioid sarcoma.
- Low grade:
  - Dermatomyofibrosis proliferans.
  - Myxoid liposarcoma.

TNM STAGING

- T1: Tumor less than 5 cm.
- T2: Tumor more than 5 cm.
- T3: Invasion to bone, vessel, nerve.
- N0: No nodes.
- N1: Nodal metastasis.
- M0: No metastasis.
- M1: Metastasis.

COMBINED STAGING

- IA: G1T1N0M0.
- IB: G1T2N0M0.
- IIA: G2T1N0M0.
- IIB: G2T2N0M0.
- IIIA: G3T1N0M0.
- IIIB: G3T2N0M0.
- IIIC: T1/T2N1/N2M0.
- IVA: T3M0.
- IVB: M1.

Stage Grouping

Stage	*Grading*	*A*	*B*
Stage I	G1	T1	T2
Stage II	G2	T1	T2
Stage III	G3	T1	T2
Stage IV		N1	M1

MSKCC Scoring System

Features	*Favorable factors*	*Adverse factors*
Size	<5 cm	≥5 cm
Site	Superficial	Deep
Histologic grade	Low	High

MSKCC Staging

Number of adverse factors	*Stage*
0	0
1	I
2	II
3	III
Metastases	IV

DIFFERENTIAL DIAGNOSIS

- Subfascial/Intramuscular lipoma.
- Chronic osteomyelitis.
- Organized hematoma.
- Myositis ossificans.
- Lymphangioma.
- Arteriovenous (AV) malformation.
- Other malignancies:
  - Osteosarcoma.
  - Lymphoma.
  - Solitary bone secondary.

INVESTIGATIONS

- Any soft tissue mass in an adult that is symptomatic or enlarging, any mass that is more than 5 cm, or any new mass that persists for more than 4 weeks should be biopsied.
- Tissue diagnosis:
  - Trucut biopsy × 2 vs Fine-needle aspiration cytology (FNAC):
    - » FNAC cannot subtype the tumor; it only shows sarcomatous elements.
    - » Only diagnoses malignancy; does not grade or classify.
    - » It is useful only in confirming recurrence after surgery and for lymph nodes to see if they are infiltrated.
    - » FNAC is done only if lymphoma is suspected.
    - » Trucut is not done on very soft (probably necrotic) or very hard (probably calcified) areas.
    - » 90% accuracy in recognition of recurrent sarcoma; grading is not possible.
  - Open biopsy:
    - » Done by the same surgeon who is going to perform radical surgery.
    - » If small and superficial: excision biopsy.
    - » Large lesion or deeply situated lesion: incision biopsy.
    - » Always done under general anesthesia (GA).
    - » Failure of repeated trucut (false negative, if necrotic or fibrotic): incision biopsy with Tilley-Henckel punch biopsy forceps and trap-door technique and site of biopsy is included in future resection.
    - » In extremities, longitudinal incision in its most superficial location without tissue flap, and meticulous hemostasis to prevent dissemination by hematoma and previous scar excised at the time of definitive surgery.
    - » No cautery (to avoid heat artifacts).
    - » Deep fascia is closed after biopsy.

  - » Drainage is brought out close to one end of incision and not through incision.
  - » In the majority of patients with intra-abdominal or retroperitoneal STS, laparotomy with intent to resect should be performed, and the diagnosis made at operation, unless the patient is clearly unresectable or will be undergoing pre-operative investigational treatment.
- – Frozen section:
  - » To ensure adequate sampling during open biopsy.
  - » To ensure tumor-free margins.
- – Aspiration cytology:
  - » 80–90% accuracy.
- – Radiologically guided grill biopsies.

- Imaging:
  - – Plain X-ray:
    - » To know the:
      - Anatomical site.
      - Borders of tumor.
      - Bone destruction.
      - Matrix formation.
    - » Findings:
      - Soft tissue mass.
      - Specifications of calcification.
      - Bone infiltration.
      - Periosteal reaction.
    - » Differential diagnosis – osteogenic sarcoma invading soft tissues.
  - – Computed tomography (CT):
    - » Position and extent of tumor.
    - » Compartmental involvement.
    - » Muscle group involvement.
    - » Neurovascular infiltration:
      - Obliteration of perineural fat.
      - Major vessel involvement and relationship to major vessels.
      - Seen better with IV contrast.
    - » Intra- and extra-osseous extension.
  - – Chest X-ray for secondaries chest; for extremity lesions, lung is the principal site of metastasis of high-grade lesions.
  - – CT chest, if CXR is negative.
  - – Liver scan for visceral lesions since liver is the principal site of metastases.
  - – Isotope bone scan:
    - » Periosteal infiltration is picked up early especially if symptomatic—bone pain or pathological fracture.
    - » Metastases and intraosseous bone spread is seen better.
  - – Magnetic resonance imaging (MRI):
    - » Tumor-muscle interface and tumor-vessel interface is seen better than CT.
  - – Angiogram:
    - » May or may not be done
    - » If CT with IV contrast is done, no need for angiogram.
  - – DSA:
    - » Principle:
      - One scout film is done.
      - One film with contrast is done.
      - Using computer, first film is subtracted from the second film.
    - » Advantages:
      - Clarity is better.
      - Less contrast.
      - Contrast can be used IV also (instead of intra-arterial).
    - » Disadvantages:
      - Expensive.
      - Clarity is less in IV than in intra-arterial angiogram.
      - May cause hypotension.
  - – Imaging of lymph nodal metastatic sites.

TREATMENT

- Principles:
  - – Surgery is the mainstay of treatment.
  - – Marginal excision is done at the level of false capsule:
    - » It is not a safe margin.
    - » So it is combined with RT.
  - – Intralesional excision:
    - » Tumor opened and curettage is done (curative), e.g. in Ewing's sarcoma.
    - » Adjuvant CT/RT alone or in combination is given.
  - – Radical wide excision with 5–10 cm margin is preferred to compartectomy, i.e. tumor with one intact tissue plane on all sides.
  - – No cautery (margins cannot be evaluated correctly).
  - – Surface of the specimen painted with India ink.
  - – Dead space if large, transposed muscle flap, and if small, suction drains are placed.
  - – Extremity should be immobilized postoperatively.
  - – If wound breaks down, myocutaneous flap is done.

- Nowadays, limb or function preserving surgery is done to avoid mutilating amputation
  - » If sciatic nerve is involved, only the nerve sheath is removed leaving behind the nerve, and postoperative radiation therapy (RT) is given.
  - » If the nerve has to be removed, no reconstruction is done, but kneebrace/footdrop splint is given.
  - » Tumor should not be seen during the procedure.
  - » Muscles should be resected from origin to insertion.
  - » Large arteries are freed in the subadventitial plane. If encased, excision and autologous vein graft.
  - » Veins: Except for common femoral, common iliac and subclavian, others can be sacrificed without reconstruction. These veins are reconstructed with autologous veins.
  - » Nerves: Epineurium is removed.
  - » Postoperative RT is given.
- For patients with limb sparing option, an adjuvant multimodality approach combining limb-sparing surgery with chemoradiation or radiation therapy yields local control and disease-specific survival rates comparable to amputation.
- Therefore, more than 90% of patients with primary extremity sarcomas should undergo limb-sparing surgery.
- Major vascular, nerve or bone involvement is a relative indication for amputation.
- For amputation, always go one joint above.

- For extremity/trunk STS (Fig. 3.1):
- Treatment modalities:
  - If intracapsular:
    - » Wide local excision with 2–3 cm clearance.
    - » In less than 5 cm growth, clearance is given all around.
    - » Frozen section to confirm negative margin.
  - If >5 cm:
    - » If low grade: Wide local excision.
    - » If high grade: Compartmental excision leaving behind major vessels and nerves.
    - » Complete excision with negative margin and PORT.
  - If >10 cm and high grade:
    - » Preoperative CT
    - » Compartmental excision.
    - » PORT.
  - Amputation:
    - » For recurrent tumors that are not resectable.
    - » Do not amputate through compartment of origin. It will lead to stump recurrence.
    - » Below knee (BK) for foot lesions, above-knee (AK) for calf lesions, hip disarticulation for large proximal thigh, and groin tumors and hemipelvectomy for pelvic tumors.
  - For head and neck, mediastinal or retroperitoneal tumors, wide excision is not possible and dosage of RT is limited.
  - For chest and abdominal wall tumors, wide excision and reconstruction.
  - For metastasis:
    - » No surgery (no role for debulking).
    - » Palliative CT—single drug:
      - I-phosphamide alone.
      - Adriamycin alone.
      - Dacarbazine alone.
      - Combination:
  - 50% response rate.
  - 10% complete cure rate.
    - » Isolated pulmonary metastases:
      - Surgery for primary.
      - Resection of secondary if patient is fit.
    - » Extrapulmonary or unresectable pulmonary metastases: Palliative CT.
  - Recurrence:
    - » If resectable, surgery.
    - » If not, RT.

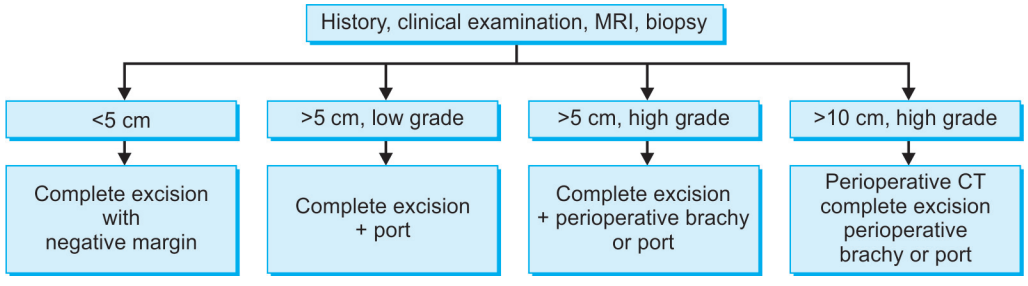


Fig. 3.1: Treatment options

– Limb-preserving surgery:
  » Tikhoff-Linberg procedure in upper limb:
    - Shoulder dislocation, remove tumor and bone, and replace with prosthesis, leaving behind uninvolved soft tissue and close.
    - + PORT.
  » Internal hemipelvectomy in lower limb.
    - Similar procedure + PORT.
– Reconstruction:
  » Groin defect:
    - Tensor fascia lata flap.
    - Inferiorly based rectus abdominis myocutaneous flap.
    - Pedicled greater omentum covered by meshed skin graft.
  » Trunk and neck defect:
    - Latissimus dorsi
    - Pectoralis major
    - Trapezius

    } Myocutaneous flap.
  » For full thickness chest/abdominal wall defect – Marlex mesh covered by appropriate myocutaneous flap.
  » For nerve deficit, braces and physical therapy or tendon transfer if possible.
– Radiotherapy:
  » PORT: 40–50 Gy initially followed by 10–15 Gy.
  » Preoperative RT:
    - For downstaging.
    - Large and fixed tumor may become small and mobile.
    - To decrease tumor-spreading effects of surgery.
    - Disadvantage: impaired healing.
  » Brachytherapy:
    - Implantation of tumor bed with radioactive sources.
    - Excellent local control, but serious wound problems.
    - Not practical in large tumor volumes.
  » Complications:
    - Fibrosis
    - Skin necrosis
    - Fracture
    - Nerve damage
    - Arterial stenosis

    } Can be reduced by lower doses.
– Chemotherapy:
  » For unresectable and recurrent tumors.
  » Adjuvant:
    - Preoperative IV or IA.
    - No conclusive evidence as to benefit.
  » Treatment:
    - Vincristine, Adriamycin, cyclophosphamide (VAC).
    - Doxorubicin, ifosfamide and dacarbazine (DTIC).
    - Cyclophosphamide, vincristine, adriamycin and DTIC (CYVADIC).
    - Ifosfamide ± Adriamycin.
    - Mesna, ifosfamide, doxorubicin and dacarbazine (MAID).
    - Doxorubicin is the most effective single agent.
  » Leiomyosarcoma and liposarcoma: Better response to CT than malignant fibrous histiocytoma (MFH) or rhabdomyosarcoma.
– Locoregional adjuvant therapy:
  » Isolated limb perfusion with CT agents
  » Combined chemoradiation with hyperthermia
  » Hyperthermia alone or in combination

  } Experimental may allow limb salvage surgeries.

Combination Therapy for STS

- Case study:
  – A 26-year-old woman with Grade III 6 cm MFH of left gastrocnemius muscle.
  – If surgery alone is contemplated, posterior compartment resection or AK amputation should be done.
  – Ideal treatment:
    » PORT with 4 field technique 2 Gy/day for 45 Gy.
    » Resection by 3–5 cm margin.
    » PORT 15–20 Gy to highest risk region (brachy) for a total of 60–65 Gy.
    » This is as effective as amputation for local control.

Advantages of PORT

- Radiation field is confined to tumor.
- RT can begin immediately (PORT is always delayed by weeks).
- After RT, tumor regresses and develops peritumor edema that marks the border of infiltrative STS.
- Well-oxygenated periphery of tumor is sterilized thereby diminishing chances of tumor implantation.

FACTORS AFFECTING LOCAL RECURRENCE

- Status of the surgical margin.
- Size of the primary.
- Grade of the lesion.
- Deep location.
- Recurrent lesion.

Follow-up

Primary extremity STS	*Clinical follow-up*	*Chest X-ray*
<5 cm, excised completely	Every 6 months for 5 years Every year thereafter	Every year
>5 cm, low grade	Same	Same
High grade, 5–10 cm	Every 3 months for 5 years Every 6 months thereafter	Every 6 months
High grade, 10 cm or more	Every 3 months	Every 3 months
Visceral or retroperitoneal (RP) STS	Every 3 months for 3 years Every 6–12 months thereafter	Abdominopelvic CT every 6 months for 3 years Every 12 months thereafter

PROGNOSIS

- 80% local recurrence within 2 years.
- Lung recurrence: If solitary, surgery.
- Adverse prognostic factors:
  - High grade.
  - Size >10 cm.
  - Tumors of head and neck, retroperitoneum, proximal limb.
  - Age >53.
  - Lymph nodal involvement.
  - Primary treatment by amputation.
  - Inadequate surgical margins.

QUICK CLINICAL SUMMARY OF STS FEATURES

- Old age (except osteosarcoma).
- Short duration history.
- Large swelling, ill-defined, skin overswelling, stretched and shiny with dilated veins.
- Variable consistency.
- Warmth (increased vascularity).
- Distal neurovascular deficit may or may not be present.
- Restricted mobility.

IMPORTANT DIFFERENTIAL DIAGNOSIS

- Intramuscular lipoma.
- Chronic organized hematoma.
- Solitary bone secondary.

SUMMARY OF MANAGEMENT APPROACH

- Extremity or trunk:
  - <5 cm all grades, >5 cm low grade: Complete excision with 2–3 cm clearance and negative margin confirmed by frozen.
  - 5–10 cm high grade: Complete excision with negative margin and PORT (external beam).
  - >10 cm high grade: Preoperative CT + complete excision with negative margin + PORT (external beam).
  - Radical excision is compartmental excision; muscle removed from origin to insertion.
  - Postoperative RT for high-grade tumors and for unsatisfactory clearance.
- Retroperitoneal:
  - Exploratory laparotomy and complete resection or incomplete resection + PORT.
- Metastases:
  - Isolated pulmonary metastases: Surgery for primary + resection of lung metastases if patient is fit.
  - Unresectable pulmonary metastases or extrapulmonary metastases:
    - » CT: 47% response rate; 10% complete response rate.
    - » MAID, VAC, CYVADIC.
- Recurrence: Isolated local reresection.

AGGRESSIVENESS OF STS

- Maximum:
  - Synovial and rhabdomyosarcoma.
  - Trucut biopsy is important.
  - Always PORT.
- More the aggressiveness, more the response to CT, RT and more the chances of spontaneous regression due to:
  - Increased immune system stimulation.
  - Increased production of tumor necrosis factor.
  - So, immunotherapy can be given for better immune response.

  - Examples include choriocarcinoma, renal cell carcinoma (RCC), malignant melanoma—5% incidence of spontaneous regression.
- Immunotherapy:
  - For RCC:
    - » IL-2.
    - » Lymphokine-activated killer (LAK) cells.
    - » Interferon.
- For malignant melanoma:
  - LAK cells.
  - Interferon.
  - IL-2.
  - Bacillus Calmette-Guérin (BCG), levamizole: Immune modulators.

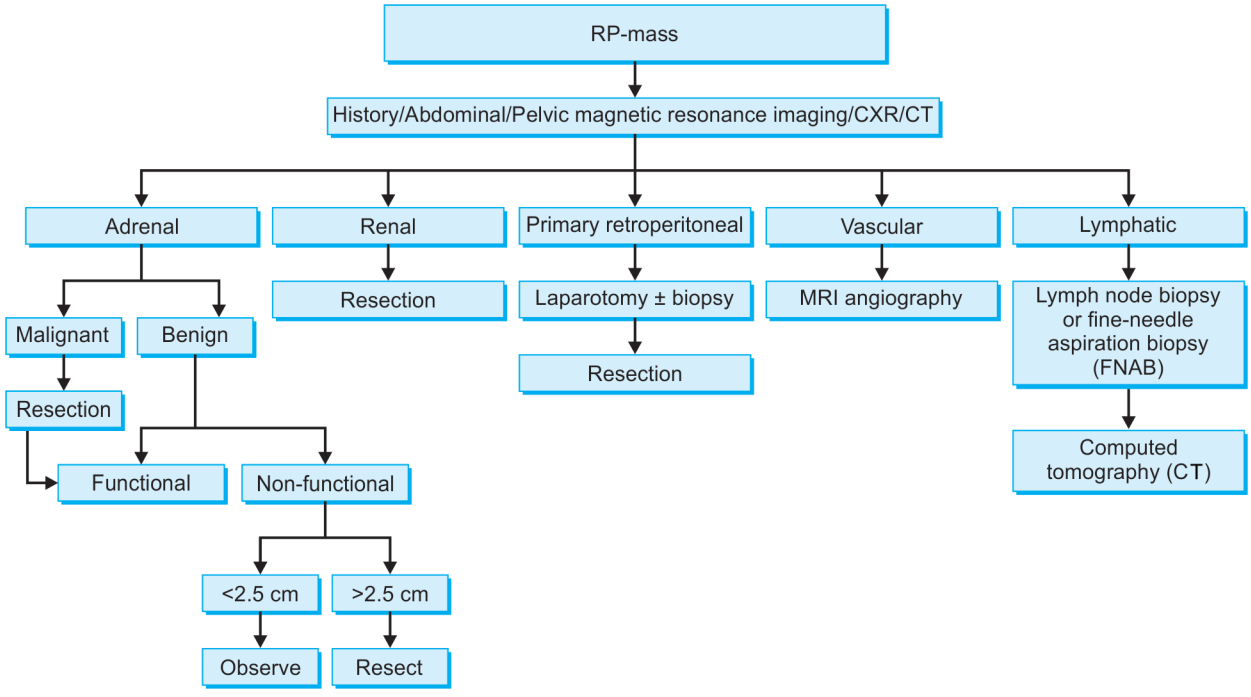
MALIGNANT FIBROUS HISTIOCYTOMA

- 2 types:
  - Myxoid.
  - Inflammatory: Poorer prognosis.
- In head and neck MFH, prophylactic block dissection of neck is advocated.

RETROPERITONEAL SARCOMA

- Associated with:
  - Gardner's syndrome.
  - Familial retinoblastoma.
  - Neurofibromatosis.
  - Li-Fraumeni syndrome.
  - Gene mutations of p53 on chromosome 17.
- Usually not fully resectable.
- Scrotum should always be examined for testicular malignancy causing retroperitoneal adenopathy.
- So PORT necessary depending on whether diagnosed early.
- Clinical features:
  - Big tumor.
  - Signs of local infiltration and pressure:
    - » Subacute intestinal obstruction.
    - » Hydronephrosis.
    - » Varicose veins.
- Treated always with surgery—either curative or palliative.
- Preoperative preparation always includes bowel preparation because bowel resection may be necessary for complete removal of tumor.
- Kidney, colon, pancreas and spleen may have to be removed (when aorta or inferior vena cava (IVC) is not involved).
- Tumor should never be enucleated; leads to local recurrence.
- Intraoperative RT gives a better prognosis.

Management Algoritham for Retroperitoneal Sarcoma



- Palliative RT given for:
  - Pain with unresectable tumor.
  - Recurrence.
  - Positive tumor margins after primary resection.
- CT given in patients with metastatic disease: multidrug CT with Adriamycin.

CHONDROSARCOMA

- Age group: 35–55 years.
- Site: Pelvis, ribs and proximal long bones.
- Origin: From benign tumors of cartilage.
- Microscopy: Speckled calcifications; no cortical destruction.
- Prognosis depends on grade:
  - 75% 5-year survival for well differentiated.
  - 10% 5-year survival for high grade.

FIBROSARCOMA

- Age and sex: male, 35–55 years.
- Site:
  - Metaphysis/Diaphysis of tibia, femur/soft tissue.
  - Highest incidence on thigh and arms.
  - From burns, scars, injection sites, ulcers, post-irradiation.
- Diagnosis:
  - Dilated veins overswelling demonstrated by infrared photography.
  - Firm rubbery nodule with slow and painless enlargement.
- Microscopy: spindle-shaped fibroblasts or pleomorphic.
- Spread:
  - Blood-borne: pulmonary, bone, liver secondaries.
  - No lymph node involvement.
- Treatment: Resection in one plane beyond involved tissue.
- Prognosis:
  - 30% 5-year survival.
  - Low grade: More differentiated, better prognosis.
  - High grade: Poorer prognosis.
  - Congenital fibrosarcoma: Better prognosis.

SYNOVIAL SARCOMA

- Age group: any age, extremely malignant.
- Site:
  - Close to a major joint.
  - May arise in connective tissue, muscle or subcutaneous tissue.
- Microscopy – elements simulating synovium and fibroblasts.
- Spread – through tissue planes, bloodstream and lymphatics.
- Prognosis – poor.

EWING'S SARCOMA

- Potential: Highly malignant.
- Site: Middle of metaphysic.
- Age: Childhood.
- Clinically: Fever, increased ESR.
- Microscopic: Onion peel appearance.
- Treatment: CT/RT.
- Prognosis: Poor.

KAPOSI'S SARCOMA

- Prevalent in immunosuppressed patients (especially HIV affected).
- Origin: From proliferating capillary vessels and perivascular connective tissue cells.
- Association: Acute or chronic immunosuppression.
- Macroscopic: Macular to nodular, brownish.
- 4 types:
  - Indolent nodules.
  - Nodules on lower limb in immunosuppressed postrenal transplant patients.
  - Epidemic: Associated with HIV.
  - Head and neck nodules.
- Site:
  - Feet, hands, ear, nose, GIT.
  - In AIDS-related Kaposi's, oral and perioral lesions are seen in 55% of cases; especially palate (commonest), tongue, pharynx and larynx.
  - May metastasize to salivary glands, or cervical lymph nodes.
- Treatment:
  - If small: Surgery.
  - If large: RT/CT/interferon.
  - Palliative:
    - » Single drug therapy with vinblastine or etoposide—systemic or intralesional.
    - » Interferon: systemic or intralesional.
    - » Local RT.

LYMPHANGIOSARCOMA

- Origin: Long-standing complication of lymphedema.
- Site: On brawny arm of postradical mastectomy

(Stewart-Treves syndrome) or on lower limb following groin resection.
- Spreads to chest wall and lungs.
- Treatment:
  - RT is the first line of treatment.
  - If it is resistant, surgery.
- Prognosis: usually fatal.

HEMANGIOSARCOMA

- Multifocal.
- If single, surgery.

RHABDOMYOSARCOMA:

- Embryonal rhabdomyosarcoma:.
  - 10% of malignancy in children <15 years.
  - 2–6 year age group: Head and neck, genitourinary (GU) tract.
  - 14–16 year age group: Head and neck, GU tract + trunk, extremities (thigh, shoulder, upper arm).
  - Better prognosis.
- Adult type: Thigh, shoulders, upper arm.
- Lymph node metastasis + blood-borne to lungs, bone and liver.
- Investigations: MRI to demonstrate spread along tissue planes.
- Treatment for head and neck: Biopsy followed by RT, CT.
- For trunk, extremity – surgery + CT.
- Prognosis:
  - In children, 5-year survival is >50%.
  - In adults, it is most malignant; CT/RT not useful.

LEIOMYOSARCOMA

- Common in gastrointestinal tract (GIT), uterus.
- May be from smooth muscle of major blood vessels, oral, nasal cavities, gallbladder.
- Treatment: Surgical.
- Recurrence is possible.
- Prognosis: Poor.

LIPOSARCOMA

- Site: Extremities, retroperitoneum.
- Incidence: Common in males and older age group.
- Orientated in the direction of underlying muscle.
- Types:
  - Well differentiated:
    - » Good prognosis.
    - » Local invasion only.
  - Myxoid:
    - » Groin, thigh, trunk, popliteal fossa and retroperitoneum.
    - » 50% risk of local recurrence after excision.
  - Fibroblastic, lipoblastic:
    - » Poor prognosis.
    - » 62% recurrence rate after excision.
  - Pleomorphic:
    - » Strong tendency to metastasize.
    - » Very poor prognosis.
    - » 77% mortality.
- Treatment: Wide excision for well-differentiated and RT for others.

DESMOID TUMORS

- Otherwise called musculoaponeurotic fibromatosis.
- Types:
  - Nodular fasciitis.
  - Plantar fibromatosis.
  - Desmoid.
- Sites:
  - Head and neck, thigh, shoulder in men.
  - In women, anterior abdominal wall is the commonest site.

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