Soft tissue sarcomas - the pitfalls in diagnosis and management!!

Prof. Chintamani
MS, FRCS(Ed.), FRCS(Glas.),FRCS (Irel.), FACS, FICS(Surg. Oncol.), FIMSA
President Association Of Breast Surgeons Of India
Tutor & Examiner
Royal College Of Surgeons Edinburgh UK
Vardhman Mahavir Medical College
Safdarjang Hospital New Delhi
• heterogenous group of tumours that are notorious for recurrence
• Paradigm shift in the management effort being more and more towards preservation of organ and function.
• Better understanding of the biological behaviour of these tumours.
• Still many grey areas that make these cancers unique.
• The cancers being rare and rarely diagnosed especially when deeply located in the organs, there are pitfalls in diagnosis and management
• As opposed to “errors”, pitfalls refer to unexpected or unforeseen difficulty or danger and do not include errors due to ignorance or lack of competence

• Underestimation of both malignant potential and suboptimal surgical treatment of primary tumour are the major pitfalls that can be lethal in these cancers
Biological behaviour-putting all eggs in one basket!!

- most significant pitfall as this heterogeneous group behaves differently at different sites, the presentation and diagnosis could be confusing.
- most pathologists have limited experience of studying these cancers on account of low incidence.
- With more than 20 histological subtypes it is nearly impossible for a single pathologist to encounter all subtypes in adequate numbers in his life time.
- Mis/suboptimal diagnosis are a rule rather than an exception.
• Inadequate or inappropriate biopsies (FNAC vs. Core needle biopsies vs. incision biopsies) and sampling errors

• FNA is mostly used to either make the initial diagnosis of exclusion or to ascertain the status of regional lymph nodes that have a major impact on the stage and outcome

• the core needle biopsies (CNB) are usually optimum and the representative yield can be improved if these are done under image guidance especially in deep seated cancers. The only concern is the adequacy of tissue desired by the pathologist to make an optimum and detailed analysis.
in making wrong incisions (transverse incisions in the thigh or extremities etc.) that might influence the planning of incisions for the subsequent surgery.
Benign lesions that may be confused as malignant (potential pitfalls!)

- Cellular benign fibrous histiocytoma that shows central necrosis in as many as 10-15% of cases
- Atypical fibrous histiocytoma (so-called ‘dermatofibroma with monster cells’)
- Lipoma showing extensive microscopic areas of fat necrosis is very frequently mistaken for atypical lipomatous tumour—however, such lesions entirely lack true adipocytic or stromal nuclear atypia or hyperchromasia
Malignant lesions mistaken as benign

- Low-grade fibromyxoid sarcoma
- Histopathological interpretation [high vs. low vs. intermediate grade tumours]
Imaging [can we bet on PET?]

MRI is considered better than computed scan (CT scan) except perhaps in the abdomen (retroperitoneal sarcomas) as it shows the soft tissue planes better.
therapies (PERCIST-PET response evaluation criteria in solid tumours on the lines of RECIST -Response Evaluation Criteria in Solid Tumours) is being used in some studies to highlight this role of PET in assessing response
Pitfalls in the management

- Optimum Surgery
- Quality of life – a notion of happiness
- Radiotherapy and chemotherapy (pitfalls in timing and sequencing)
- Pre-operative radiotherapy
- Preoperative chemotherapy
- R1 resection although not desirable may be acceptable in this subset.
Site Specific presentation of Soft Tissue Sarcomas

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Site specific distribution

- Head & Neck: 11%
- Trunk: 10%
- Retroperitoneum + Intra-abdominal: 14%
- Visceral: 15%
- Upper extremities: 15%
- Lower extremities: 35%
Pattern of growth

• Most grow in expansile manner
• Flattening the normal tissues around them in concentric manner to form compression & reactive zone ::= pseudocapsule
• Extension sprouting small tentacles perforating the pseudocapsule
• Small finger like extensions grow to form clinical occult deposits at a variable distance from the parent lesions
• Continued growth => multinodular mass configuration of large, high grade deeply situated sarcomas
• “Barriers”
Skin

- Angiosarcoma/Lymphangiosarcoma
- Kaposi`s sarcoma
- Epitheloid sarcoma
- Dermatofibrosarcoma protuberans (on trunk)
Dermatofibrosarcoma Protuberans
Angiosarcoma (Blood/Lymph vessels tumours)

Hemangiosarcoma

- Elderly
- Aggressive, arises in head and neck, breast, liver
- Specially affects the skin and superficial soft tissue (most STS are deep)
- Differentiation from hemangioma difficult
- 5-year survival rate < 20%
Hemangiopericytoma

- Originates in blood vessels
- Affects all ages
- Develops in the tips (Glomus tumour)
- Lower extremity/pelvis
- Commoner in females
- 5-year survival rate about 50%
Glomus tumour
Lymphangiosarcoma

- Older patients
- Aggressive
- Arises in chronic lymphatic stasis (specially post mastectomy)
- 5 year survival rate 10%
Dermatofibrosarcoma protuberans

- Rare
- Develops in the skin of trunk/extremities
- Almost never metastasizes
- 10% poorly differentiated (high grade)
- Survival related to tumour grade
Dermatofibrosarcoma Protuberans (recurrent)
DFS protuberance inguinal lymph nodes
Iliac part of dissection
Ilio-inguinal block dissection
Kaposi`s sarcoma

- Tissue of origin not clearly known
- Older patients
- Extremely indolent lesion
- Lower extremity (Mediterranean Jews)
- The epidemic & aggressive variety is associated with AIDS and in Bantu tribes
Epitheloid Sarcoma

- Rare & tissue of origin unknown, young adults
- Aggressive, typically appears on distal extremities
- Amongst the most common tumors of the hand and foot
- Spreads to non contiguous areas of skin, subcutaneous tissue, fat, draining lymph nodes and the bone [differentiates it from other STS]
- 5 year survival rate 30%
Lymph nodes

Spindle cell Sarcoma neck and chest wall
Head and neck

- Rhabdomyosarcoma

- Angiosarcoma (elderly)

- Osteogenic sarcoma (Jaw)
Spindle cell sarcoma palate
Rhabdomyosarcoma

- Tissue of origin: striated muscle
- In G-TNM staging all are grade – III
- All types in any age group
- **Embryonal**: teenagers, head and neck (70%), genitalia
- **Pleomorphic**: >30 years, rare, develops in extremities, highly anaplastic
Rhabdomyosarcoma (recurrent)
Distal extremity

- Epithelial sarcoma
- Synovial sarcoma
- Clear cell sarcoma
- Osteogenic sarcoma (femur)
Clear cell sarcoma

- Rare & tissue of origin unknown
- Now recognized as a form of malignant melanoma
- Adults <40 years
- Painless, firm, spherical masses on tendon sheaths & aponeurotic structures of distal extremities, head and neck
- 5 year survival rate is about 50%
CLEAR CELL SARCOMA

Trachea
Extremities

- Liposarcomas
- Malignant fibrous histiocytoma
- Tendosynovial sarcoma
- Fibrosarcoma
Malignant Fibrous histiocytoma
Liposarcoma

- Arises from fat tissue
- 15-18%
- Middle aged and older men
- Thigh, groin, buttocks, shoulder, retro-peritoneum
- Does not arise from benign lipomas !!
- 5 year survival rate is 80% for low grade, 20% for high grade liposarcomas
Liposarcoma Thigh
Fibrosarcomas

• Tissue of origin: fibrous tissue
• Incidence 5-10%
• Affects all age groups
• Arises in mesenchymal sites
• Usually involves abdominal wall and extremities
• 90% well differentiated (desmoid)
DESMOID TUMOUR
Malignant fibrous histiocytoma (MFH)

- Tissue of origin: fibrous tissue and histocyte
- Incidence 10-23%
- Age > 40 years
- Most common STS in some series
- Develops in extremities (specially legs), retroperitoneum
Mesothelium/retroperitoneum/mesentery

- Mesothelioma
- Leiomyosarcoma
- Liposarcoma
- Malignant fibrous histiocytoma
Mesothelioma

- Tissue of origin: mesothelium
- > 50 years
- Asbestos exposure
- Involves pleura, peritoneum
- Highly lethal
- 5 year survival rate < 10%
Retroperitoneal sarcomas

• Liposarcomas

• Leiomyosarcomas

Retropertitoneal sarcomas
Gastrointestinal tract

- Gastrointestinal stromal tumours (GIST) don’t express markers of myogenic differentiation
- Similar to leiomyosarcomas but different IHC staining
- GIST don’t stain for actin (Leiomyosarcomas do!)
- GIST express SC 117 (c-lat protein)
- Gastrointestinal autonomic nerve tumours (GANT) exhibit neural differentiation
- The pattern of recurrence is intra-abdominal including liver metastasis
Leiomyoma Peritonealis Disseminata (LPD)

- Women in reproductive years
- Asymptomatic, benign looking leiomyomas scattered throughout the peritoneal cavity ranging from 1-10 cm in size
- Stimulated by estrogen
- LPD causes occasional mechanical problems with bowel or pain
- No treatment except estrogen or anti-estrogen drugs required
Genitourinary tract

- Rhabdomyosarcoma

- Leiomyosarcoma (in adults): most common

- Arises in the bladder, kidneys, prostate (older patients)
Testes

Rhabdomyosarcoma arising in the para testicular tissue
Uterus

Three major types

• **Leiomyosarcoma** tumour of myometrium
• **Mesodermal mixed tumour** > malignant mixed Mullerian tumour

=> Elements of both carcinoma and sarcoma

• **Endometrial Stromal sarcoma**: arises from endometrial stroma and has an aggressive behaviour
Leiomyosarcoma vulva/Vagina
Chest wall

- Desmoids
- Liposarcomas
- Myogenic sarcomas
Neurofibrosarcoma [MPNST, Schwanoma]

- Origin: Nerve sheaths (thickening nerves, without anatomic predilection)
- Young and middle aged
- Patients with Von-Recklinghausen`s disease (10% develop sarcomatous changes during life)
- Histologically resembles fibrosarcoma
- Presents with **Superficial variety**: low grade, spreads extensively along nerve sheaths without metastasizing, survival rate > 90%
- **Penetrating variety**: nodular growth, vascular invasion/lung metastasis 5 year survival < 20%
Synovial sarcoma

- Originates from tenosynovial mesothelium
- Young adults 2\textsuperscript{nd} to 4\textsuperscript{th} decade
- Hard masses near tendons & painful
- Synovial and epitheloid sarcomas are the most common tumours of hands, knee and feet
- G-TNM stage: all are grade –III.
- High rate of local recurrence