PRINCIPLES OF LIMB SALVAGE IN PAEDIATRIC MALIGNANCY

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Leukaemias 33,8%
CNS tumours 20,3%
Lymphomas 12,5%
Sympathetic nervous system tumours 8,6%
Soft tissue sarcomas 6,5%
Renal tumours 6,1%
Bone tumours 4,6%
Germ cell tumours 3,4%
Other diagnoses 4,2%
The last few decades have seen rapid strides in the field of musculoskeletal oncology. Amputation no longer remains the only option to achieve local control in malignant bone and soft tissue tumors of the extremity.
This dramatic alteration in patient care is the result of significant advances along many fronts, including:

1. improved understanding of tumor biology
2. effective induction chemotherapy
3. technical advances in surgical techniques
4. better characterization of the biomechanics of the human skeleton
5. advanced material engineering and manufacturing techniques;
6. the development of a reliable, stable modular prosthesis for reconstruction.
In treating extremity sarcomas, the major therapeutic goals are:

Survival

Avoidance of a local recurrence

Maximizing function, and

Minimizing morbidity
The **MULTIDISCIPLINARY TEAM APPROACH** — it’s importance:

1. Optimizes treatment planning
2. Minimizes duplication of diagnostic studies
3. Reduces the time to implementation of the definitive therapeutic protocol.
Limb Sparing Surgery has now replaced amputation as the primary form of surgical intervention.

95% of patients with extremity sarcomas managed by limb conservation surgery

5% of these patients still require amputation. Common indications for amputation are:

- major neurovascular involvement and
- large primary tumor where resection would leave a functionally useless limb.
LIMB SPARING SURGERY — CRITERIA

• Tumour resection occurs with oncologically sound margins

• Reconstruction leads to a functional limb

• Soft tissue defects can be closed primarily or with soft tissue transfers.
Limb sparing surgery—indications

- Tumour meets all the criteria for LSS
- Survival is long enough to justify complex surgery
- Patient refuses amputation and accepts the risk of local recurrence of disease from inadequate margins
- Palliating patients with limb disease that is easily and safely operated on to improve quality of life e.g. impending fractures, fungation.
LSS- CONTRAINDICATIONS

- Survival is not expected to exceed three months
- Gross contamination.
- Local or systemic sepsis.
- Patient co-morbidities do not permit safe anaesthesia or surgery.
- NV involvement (relative contraindication)
A successful limb-sparing procedure can be divided into three stages, each of which directly affects patient outcome and survival.

- Tumor resection must spare significant structures.
- A stable, painless skeletal reconstruction must be accomplished.
- Third, the surrounding and supporting soft tissue is required to restore function and skeletal reconstruction.
The goal of the surgical resection should be to widely resect the tumor with negative margins, meaning a cuff of normal tissue completely surrounding the tumor.

Local recurrence rates after simple excision are 60 to 90 percent.

After wider local resection reduces to between 8 and 30 percent.

Following radical surgery, such as a compartmental resection, local failure rates are between 10 and 20 percent.
Wide surgical resection of the primary tumor (total en bloc excision of the primary tumor without cutting into tumor tissue and having a normal cuff of tissue completely surrounding the tumor) is the essential component of treatment for virtually all patients. The resection should take place through tissues outside of the tumor pseudocapsule, if one exists, through normal uninvolved tissue.

It should include en bloc the biopsy site with primary surgical specimen
• Arterial invasion

- Major arteries can be resected

- Reconstruction——reverse saphenous vein grafts or artificial materials
RECONSTRUCTIVE OPTIONS FOR SKELETAL DEFECTS

• Resection arthrodesis.
• Osteoarticular or massive allograft
• Allograft–prosthetic composite (APC)
• Endoprosthesis.
• Expandable Implants for the Skeletally Immature Patients
In these early days of limb salvage, resection arthrodesis was the main method of reconstruction.

Its primary advantages to restore skeletal stability and produce a long-term, durable reconstruction.
Osteoarticular or massive allografts

- A biologic solution to restoring a segmental defect of the skeleton.

- Complications; such as infection, nonunion and joint instability, and late complications such as instability and allograft fracture.
Allograft–prosthetic composite (APC).

- Transitional step between allografts and endoprostheses.
- Provide the benefits of a biologic reconstruction along with the immediate stability achieved by a cemented endoprosthesis.
- Same high rate of early complications (i.e. infection and nonunion) as does standard allograft reconstruction.
Endoprosthesis

- Highly successful and durable method for the restoration of skeletal integrity and joint function.
- Cemented stem provides immediate fixation for early mobilization and rehabilitation.
- Advances in the use of local rotational flaps have improved joint stability and simultaneously reduced the risk of infection.
- Custom-manufactured endoprostheses replaced by modular systems with standard instrumentation and multiple reconstruction option.
Biopsy

- Osteosarcoma distal femur (conventional intramedullary)
Induction Chemotherapy

Three cycle of Adriamycin and high dose Methotrexate based chemo.
Post chemo.
Lab No. S14202/15 & S14203/15

Gross Description
S14202/15
Received for frozen, bone marrow tissue: Two bone marrow pieces together measuring 1x0.5x0.3 cm.

S14203/15
Distal femur specimen measures 15 cm in length. Outer surface shows attached soft tissues and tendons. The segment is already bisected. Cut surface shows a tumor in the metaphyses measuring 8x6 cm which is extending into epiphyses, cut surface of which is grey white and firm to hard with focal haemorrhagic and gelatinous areas. Tumor is involving the medullary cavity and breaching through the cortical bone to extend in the soft tissue in distal part of femur. Rest of the bone appears normal. Tumor is situated 5 cm away from the proximal resection margin of bone.

Microscopic Examination
S14202/15 and S14203/15
Sections from tumor in the medullary cavity of the lower end of femur show large areas of necrosis. Multiple irregular bony trabeculae of new bone formation, abundant fibrous tissue proliferation with neovascularization, focal granulation tissue and a few scattered bizarre cells possibly proliferating fibroblasts are seen. Also present are very tiny foci of residual tumor involving much less than 2-3% of the examined tissue suggestive of an osteosarcoma. The soft tissue in the periphery of tumor are free of tumor.

Diagnosis
Post chemotherapy resected lower end of right femur: Extensive necrosis with fibroblastic proliferation (>95%) and scanty residual osteosarcoma.

Note: Report on proximal resected margin will follow.
CASE 3
slicing reveals a partially well circumscribed tumor measuring 4.5x3.5x3 cm. Cut surface through which is grey white and firm. The tumor is situated 10 cm, 8 cm, 3.5 cm and 2 cm away from proximal, distal, medial and lateral margins respectively and it is 0.5 cm and 2 cm away from the resected base and skin respectively. The tumor is intermuscular in location and infiltration the muscle. Serial slicing reveals another tumor nodule measuring 1x1 cm which is situated 2 cm proximal to main tumor. This nodule is 1 cm away from the lateral margins (which is the nearest margin) and 1.5 cm from the skin.

Microscopic examination

Sections show a highly cellular spindle cell tumor. Tumor cells are arranged in sheets and interlacing bundles. There is moderate nuclear atypia with presence of scattered tumor giant cells. Increased mitotic activity is observed. Focal necrosis and myxoid change is present. At places the tumor is having nodular pattern. Tumor is focally extending into surrounding adipose tissue and skeletal muscle. Resected base, proximal, distal medial and lateral margins of the larger nodule are free. Overlying epidermis is also free. The smaller nodule also shows similar tumor. The tumor in this smaller nodule is reaching very close to the nearest margin (lateral) however the cut margin is free.

IHC: Tumor cells are positive for S-100.

Diagnosis

Left thigh mass: Malignant peripheral nerve sheath tumor.
Limbsparing surgery is the technique of choice for surgical management of limb sarcomas. In comparison to amputation, limb sparing surgery:

Has the same overall survival rate

Has higher patient satisfaction

Has a lower energy expenditure for walking

Has a lower cost to the community.

Limbsparing surgery is a complex procedure that requires expert knowledge of the requirements and criteria for its use. It is an important part of multidisciplinary management of sarcoma. The success of such surgery is maximised when conducted in centres with specific interest and expertise in this field.
THANK YOU