DIAGNOSIS AND TREATMENT OF SOFT TISSUE TUMOURS

THE DUTCH NATIONWIDE-ACCEPTED CONSENSUS

AN VAN GEEL, JAM VAN UNNIK & RB KEUS

DEFINITION

A heterogenous group of uncommon tumours with histopathological aspects of connective, muscle, fatty tissue or peripheral nerve tissue.

Include nonepithelial tumours.

Exclude tumours haematopoietic system, lymph nodes, skeleton and central nervous system.

CLASSIFICATION
(Enzinger and Weiss)

- Fibrosarcoma
- Dermatofibrosarcoma protuberans
- Malignant fibrous histiocytoma
- Liposarcoma
  - well differentiated
  - mixoid
  - round cell
  - pleomarphic
- Leiomyosarcoma
- Rhabdomyosarcoma
- Rhabdoid sarcoma
- Angiosarcoma
- Malignant haemangiopericytoma

- Synovial sarcoma
- Malignant Schwannoma
- Peripheral primitive neurectodermal tumour
- Extraskeletal chondrosarcoma
  - myxoid
  - mesenchymal
- Extraskeletal osteosarcoma
- Malignant granular cell tumour
- Alveolar soft tissue sarcoma
- Clear cell sarcoma
- Extraskeletal Ewing's sarcoma

Most frequent localization :

- Lower extremity 40% (75% above knee)
- Upper extremity 15%
- Chest/abdominal wall 15%
- Head and neck 15%
- Retroperitoneum 10%

EPIDEMIOLOGY

- Soft tissue sarcoma < 1% all new malignancies (National Cancer Registration)

Incidency rate

- 2.7 in males per 100,000 per year
- 2.1 in females per 100,000 per year

DIAGNOSTICS

- Assesment of complaints & growth speed & local tumour growth
- Assesment of regional & distant metastases
- Involvement of surrounding structures

IMAGING

- X RAYS:
  - impression extension tumour
  - skeletal involvement
- USG:
  - impression of presence
  - localization
  - extension tumour

- CT
- MRI
  - exactly localization
  - macroscopic extension

SCREENING

- Metastasizes to lung → x ray

INCISIONAL BIOPSY

- Avoid contamination of other muscles
- Longitudinal incision
- Not undermine skin margins
- Biopsy from peripher
- Wash with sterile water

→ Pathology

Recording

- localization
- tumour size
- recurrence
- previous treatment

TREATMENT

- Surgery

  - direct primary excision : size < 3 cm, superficial
  - radical resection : tumour en bloc + normal tissue at least 2 cm

  Inadequate surgery → clearance margins show evidence of residual macroscopic tumour → re-resection entire contaminated area

  If re-resection is not possible → radiotherapy

- Radiation

  Indication :

  - after plan non radical resection
  - resection margin < 1 cm after fixation
  - incomplete resection of the original surgical area

  Patient with limited tumour process, post operative adjuvant radiotherapy reduce the risk of a local recurrence.

  In this combined therapy → pre-operative
  → post-operative

The advantages post-operative radiation over pre-operative radiation:

- No delay of surgery
- No increased risk of post-operative complications
- Optimal information on the extent, margins and histological aspects of tumour → treatment volume
- The radiation oncologist can inform himself exact tumour extent during surgery

Post-operative radiotherapy indicated for G3 tumours and recurrences

In all other cases post-operative radiotherapy indicated by the result of surgery :

- After non-radical surgery
- After reoperation because of non-radical surgery
- After marginal radical surgery (< 2 cm)
- Tumour contamination during surgery

Guidelines for determining the target area :

- Related compartment is target area
- Margins grades 2 and 3 are 7-10 cm longitudinally from original tumour. For low grade → the margin 5 cm.
- In subcutaneous tumours not infiltrating fascia → margin 5 cm.

Severe complication after radiotherapy :

- Function impairment – fibrosis muscles & subcutaneous tissues
- Joint ankylosis
- Lumph oedema
- Vascular insufficiency – vascular damage

→ physiotherapy

- Chemotherapy

The End

==========

==========