

Sarcomas

Sarcomas are malignant tumours that are named differently depending on which tissue they arose:

Rhabdomyosarcoma

Definition:

Tumor of the muscle that can arise in any part of the body

Incidence:

- The average age is six to eight years and also in teenagers
- Rhabdomyosarcoma is the most common soft tissue sarcoma of childhood and comprises approximately 50% of all soft tissue sarcomas

Histological subtypes:

Embryonal - more favorable and tend to occur in the younger age group

Alveolar - less favorable and tend to occur in adolescent age group

Signs and symptoms:

Usually present with a lump or swelling. The site is variable. Usually originates in one of three major regions of the body. Visible mass.

Head and Neck (40%) - Facial pain, nasopharyngeal obstruction, cranial nerve palsies

Genito-urinary tract (20%) - Vaginal bleeding, urinary obstruction, constipation

Extremity (20%) - arm, leg, shoulder or buttock

Staging & Prognosis:

Survival rates depend on site of tumor and extent of disease

STAGE	2 YEAR SURVIVAL RATE (%)
I	85-90
II	75
III	60
IV	20

Diagnostic Evaluation – at BCCH

- CT or MR of primary lesion
- Bone scan
- Bone marrow aspirate and biopsy
- Biopsy of mass after MR scan

Treatment

1 Surgery:

Surgical biopsy is usually essential. The tumour should be surgically resected if possible.

2 Radiotherapy:

Rhabdomyosarcoma is radiosensitive and radiotherapy is important for primary tumour control if surgical resection is not possible.

3 Chemotherapy:

All patients require adjuvant chemotherapy. This has resulted in a significant improvement in survival. Commonly used drugs: cyclophosphamide, dactinomycin, vincristine, doxorubicin, ifosfamide, etoposide

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Soft Tissue Sarcoma

Rhabdomyosarcoma

Fibrosarcoma

Neurofibrosarcoma

Synovial sarcoma

Malignant fibrous histiocytoma

Rhabdoid tumour

Cell of Origin

Embryonic mesenchyme that gives rise to skeletal muscle

Fibroblasts

Schwann cell

Mesenchyme

Fibroblast

Unknown