PRINCESS MARGARET CANCER CENTRE
CLINICAL PRACTICE GUIDELINES

CENTRAL NERVOUS SYSTEM

SKULL BASE TUMOURS
CNS Site Group – Skull Base Tumours

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Skull Base Tumours

1. Introduction
   - chordoma of skull base, account for 0.2% of intracranial tumours
   - chondrosarcoma of skull base, account for 0.1% of intracranial tumors

This document is intended for use by members of the Central Nervous System site group of the Princess Margaret Hospital/University Health Network.

The guidelines in this document are meant as a guide only, and are not meant to be prescriptive. There exists a multitude of individual factors, prognostic factors and peculiarities in any individual case, and for that reason the ultimate decision as to the management of any individual patient is at the discretion of the staff physician in charge of that particular patient’s care.

2. Prevention
   - No specific prevention available

3. Screening and Early Detection
   - No screening available

4. Diagnosis and Pathology

Chordomas
   - chordomas arise from ectopic notochord remnants in skull base (40%), vertebrae (10%) and sacrum (50%)
   - generally indolent, slow growing, but do invade aggressively into bone and into adjacent soft tissue
   - metastases are rare, local recurrence is the main issue
   - on MRI, chordomas of the skull base are generally midline and are hypo-intense on T1 and hyperintense of T2 with minimal enhancement with gadolinium
   - CT imaging often demonstrates the bony lytic changes very exquisitely
   - chordoma histologic sub-types: conventional
     - chondroid
     - dedifferentiated
   - microscopically, conventional chordomas are composed of uniform cells with small oval or round eccentric nuclei and dense chromatin. The hallmark microscopic features of chordomas are the numerous, variably sized vacuoles located in the tumor cell cytoplasm, the physaliphorous cells
   - cells may be arranged in a diffuse or lobular pattern, or they may be clustered in groups or islands in a sheet like pattern
   - fibrous tissue surrounds the neoplasm and extends projections into the tumor, usually without forming a true capsule
   - chondroid variant contain a significant cartilaginous component with features of either chondrosarcoma or chondroma
   - the dedifferentiated variant of chordoma is rare, comprising 2-8% of chordomas. These can occur de novo, or as a sarcomatoid transformation in recurrences of conventional chordoma, sometimes following radiation therapy
specialized histochemistry, chordoma tumor cells tend to be periodic acid-Schiff (PAS) positive. The matrix stains diffusely with mucicarmine and Alcian blue, and it stains metachromatically with toluidine blue; it is negative with Sudan black

- electron microscopy, ultrastructural features in chordomas include desmosomal attachments and prominent mucinous vacuoles

- immunohistochemically, the tumor cells label with cytokeratins and epithelial membrane antigen (EMA), and are S-100 positive

- subject to TNM staging system for bone tumours, however in published data the TNM staging system has not been useful for prognostic purposes, with the degree of resection and local extent much more important

Chondrosarcomas
- Chondrosarcoma is a malignant cartilage tumor that originates from enchondral bone
- usually arises in the parasellar area, cerebellopontine angle, or paranasal sinuses and it may also arise in the clivus
- most common in men in the fourth decade of life
- on CT, chondrosarcomas of the skull base demonstrate matrix calcification, endosteal scalloping, cortical breach, soft tissue mass and heterogeneous contrast enhancement
- on MRI, chondrosarcomas of the skull base are generally lateralized and are low to intermediate signal on T1 and on T2 very high intensity in non mineralised / calcified portions, and on T1 gad most demonstrate heterogeneous moderate to intense contrast enhancement
- chondrosarcomas can be divided into conventional, clear cell, mesenchymal, and dedifferentiated
- conventional subtype is the most common in the skull base
- conventional chondrosarcomas have been further subdivided into histologic grades I-III
- these tumors do not stain for epithelial markers or oncofetal antigens, distinguishing them from chordomas
- subject to TNM staging system for bone tumours, however in published data the TNM staging system has not been useful for prognostic purposes, with the degree of resection and local extent and grade are much more important

5. Management

5.1 Management Algorithms
- all cases are reviewed at tumour board by a multidisciplinary team (neurosurgery, otolaryngology, radiation oncology, neuro-oncology, neuropathology, neuroradiology) for a recommendation on further management
- skull base chordomas and chondrosarcomas are generally approached with an attempt for gross total resection
- all chordomas, whether grossly totally resected or with an incomplete resection are offered high dose focal RT
• chondrosarcomas, which are generally low grade, are usually simply observed if a gross total resection is accomplished, and are considered for a course of high dose RT if an incomplete resection is done

5.2 Surgery
• 5 year survival and progression free survival is significantly improved with gross total resection
• maximal safe subtotal resection is to be done even when gross total resection is not possible
• various approaches and techniques are possible depending on the exact location and extent of the tumour

5.3 Medical Therapy
• there is no known effective chemotherapy agent in chordoma or chondrosarcoma
• imatinib mesylate has demonstrated some moderate activity in chordoma

5.4 Radiation Therapy
• historically proton beam therapy had been the standard of care, but recent innovations in photon therapy have allowed for the same doses, control rates and complication rates as seen with proton therapy
• Immobilization: thermoplastic mask
• Imaging: CT, MRI flair, T1 with gadolinium
• GTV: surgical cavity and enhancing disease
• Care is taken to not include surgical approach areas
• CTV: 0.5 cm
• PTV: 0.3 cm
• Technique: IGRT, IMRT, Stereotactic, arcs
• Dose: Chordoma 76-78 Gy/38-39
• Chondrosarcoma 70 Gy/35
• all patients treated on units with hexapod treatment couch
• IGRT: daily cone beam CT, correct for all displacements greater than 1 mm and angular displacements greater than 1 degree

6. Oncology Nursing Practice

Refer to general oncology nursing practices

7. Supportive Care

7.1 Patient Education

Driving
• possible restriction

Seizures
• education about seizures
• what to do when a seizure occurs
• how to take seizure medications
• possible side effects of seizure medications
• avoid heights, taking baths or swimming alone

Raised Intracranial Pressure: Steroids
• symptoms of raised intracranial pressure
• side effects of steroids
• titration of steroids for optimal dose

When to call multidisciplinary team
• change in seizure pattern
• new or progressive neurologic loss
• symptoms of raised intracranial pressure

7.2 Psychosocial Care
• assess family finances
• assess for possible disability applications
• assess possible depression/anxiety
• presence or absence of drug program, apply for provincial assistance if necessary
• possible need for assistive devices or services in the home

7.3 Symptom Management
• seizures
• raised intracranial pressure
• neurologic loss
• visual loss
• depression
• psychosis
• anger issues
• poor memory

7.4 Clinical Nutrition
• recommend normal diet as per recommendations of Canadian Cancer Society
• diabetic diet if elevation of blood glucose secondary to steroids

7.5 Palliative Care
• make referral in cases of progressive disease for which there is no further active therapy recommended
• management of uncontrolled symptoms

7.6 Rehabilitation
• in cases of neurologic loss, assess for possible rehabilitation OT/PT
• assess for supportive devices in the home

8. Follow-up Care
• q6 monthly with MRI brain for first 5 years
• q12 monthly with MRI brain between 5-10 years
• q24 monthly with MRI beyond 10 years