



PRINCESS MARGARET CANCER CENTRE CLINICAL PRACTICE GUIDELINES

CENTRAL NERVOUS SYSTEM

MENINGIOMA

CNS Site Group – Meningioma

Author: Dr. Norm Laperriere Date: April 2019

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Meningioma

1. Introduction

- 20% of all intracranial tumors
- 2 cases per 100,00 per year, increases with older age
- female/male ratio 2:1
- majority of cases are sporadic
- associated with NF2
- associated with prior RT to brain/head/scalp

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

- No TNM staging
- Typical appearance on CT/MRI: diffusely enhancing mass arising from meninges
- WHO 2007 classification: 3 grades
- Grade I, benign, approx. 85% of cases
- Grade II, atypical, approx. 10-12% of cases
- Grade III, malignant, approx. 3% of cases

- The best-characterized and most common genetic alteration is the loss of the NF2 gene (*NF2*) on chromosome 22q. *NF2* encodes a tumor suppressor known as merlin (or schwannomin)
- Up to 60% of sporadic meningiomas were found to harbor *NF2* mutations.
- Monosomy of chromosome 7 is frequently reported in radiation-induced meningiomas.
- The most consistent chromosomal abnormality isolated in meningiomas is on the long arm of chromosome 22.
- Meningiomas can also be associated with different genetic syndromes, namely NF2, Gorlin, Rubinstein-Taybi syndromes.

2007 WHO Grade	Histological Subtype	Histological Features
I	Meningothelial, fibroblastic, transitional, angiomatous, microcystic, secretory, lymphoplasmacytic	Does not fulfill criteria for grade II or III

	metaplastic, psammomatous	
II (Atypical)	Chordoid, clear cell	4 or more mitotic cells per 10 hpf and/or 3 or more of the following: increased cellularity, small cells, necrosis, prominent nucleoli, sheeting, and/or brain invasion in an otherwise Grade I tumor
III (Anaplastic or Malignant)	Papillary, rhabdoid	20 or more mitoses per 10 hpf and/or obviously malignant cytological characteristics such that tumor cell resembles carcinoma, sarcoma, or melanoma

5. Management

5.1 Management Algorithms

- all cases are reviewed at tumour board by a multidisciplinary team (neurosurgery, otolaryngology (skull base tumours), radiation oncology, neuro-oncology, neuropathology, neuroradiology) for a recommendation on further management

Prognostic Factors:

- Simpson resection grade
- age, older age worse
- histologic grade

Grade I

- Complete surgical resection is the treatment of choice for most non-skull based tumors.
- For newly diagnosed symptomatic meningioma for whom surgery is not being done (ie: cavernous sinus, optic nerve, other skull base), up front RT is usually recommended.
- For meningiomas where a partial resection has been done with residual disease, then upfront RT or delayed RT are possible options.
- For recurrent meningiomas, upfront RT is usually recommended.

Grade II

- Complete surgical excision wherever possible.
- For atypical meningiomas where a gross total resection has been done, then observation and RT for recurrence is usually recommended, as the 10 year relapse rate is approx. 50%, so possibly half of all patients with a complete resection will never recur.
- a randomized study of observation vs immediate RT is about to open for completely resected atypical meningiomas
- For gross residual or recurrent tumor, then RT is usually recommended at that time.

Grade III

- Complete surgical excision wherever possible.
- For malignant meningiomas, post-operative RT is recommended in all cases regardless of degree of resection.

5.2 Surgery

Complete surgical resection (Simpson's grade I) is the standard of care for all grades of meningioma, but is seldom possible in skull base tumors (the vast majority of which are grade I meningiomas).

Vertex parasagittal tumor locations are also difficult to achieve a Simpson's grade I resection in view of attachment of tumor to the superior sagittal sinus and residual or recurrent disease in that region is a common occurrence.

Most cases will be amenable to a microscopic resection, but certain anterior midline skull base locations may be amenable to an endoscopic resection trans-nasally through the sphenoid sinus or anterior skull base.

For tumors with invasion of the cranial bone, resection of the bone should be done with reconstitution of the skull with synthetic material.

Occasionally, preoperative embolization of feeding vessels may be done to reduce intraoperative blood loss.

5.3 Chemotherapy

There is no known chemotherapeutic or molecular agent known to be effective for any grade of meningioma at any stage of the disease. Patients may be treated with new experimental agents as they become available at the time of recurrence where they have exhausted any further surgical or radiotherapy option.

5.4 Radiation Therapy

Radiosurgery:

Indications:

- maximal tumor diameter should not exceed 3 cm
- skull base meningiomas at initial presentation
- residual/recurrent grade I meningiomas
- recurrent grade II/III meningiomas after prior fractionated RT

Immobilization:

- rigid radiosurgery frame

Imaging:

- CT and MRI T1 gadolinium, T2

GTV:

- enhancing tumor

CTV, PTV:

- none

Dose:

- 12 to 14 Gy based on location and aggressiveness of tumor

Fractionated RT:

Indications:

- all tumors with a maximal dimension greater than 3 cm
- all tumors with a prior partial resection
- all grade II and III tumors

Immobilization:

- thermoplastic frame
- stereotactic relocatable frame

Imaging:

- Pre-operative imaging (CT, MRI)
- planning CT
- planning MRI (T1 gad, T2)

GTV:

- surgical cavity and enhancing tumor
- care is taken to not include surgical approach areas

CTV:

- 0 for grade I meningiomas with no prior surgery, may use a 3-5 mm margin on adjacent dura
- 0 - 5 mm for grade I meningiomas with prior surgery
- 0.5 mm for all grade II and grade III tumors

PTV:

- 0.3 cm with daily IGRT

Technique:

- daily IGRT
- IMRT

Dose:

- grade I meningioma 50 Gy/25 or 54 Gy/30
- atypical or malignant meningioma 60 Gy/30 + optional boost to enhancing mass with 3mm margin (PTV) of 6-10 Gy/3-5 fractions, for total of 66-70 Gy/33-35 fractions

IGRT

- daily image guidance is provided with cone beam CT and all displacements greater than 1 mm are corrected daily
- for any angular displacement greater than 3 degrees, the patient will be taken off the treatment table and set up will be redone

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

- grade I meningioma at seen 6 months, then q12 monthly with MRI until 5 years, then once every 2 years
- Atypical/malignant meningiomas seen q 6 monthly with MRI until 5 years, then once yearly