Provincial Nasopharyngeal Cancer Treatment Guidelines

Approved at the Provincial Head and Neck Cancer Guideline Meeting
May 8, 2015

Clinical practice guidelines have been developed after multi-disciplinary consensus based on best available literature. As the name suggests, these are to be used as a guide only. These guidelines do not replace physician judgment which is based on multiple factors including, but not limited to, the clinical and social scenario, comorbidities, performance status, age, available resources and funding considerations. The Saskatchewan Cancer Agency disclaims all liability for the use of guidelines except as expressly permitted by the Agency. No portion of these guidelines may be copied, displayed for redistribution to third parties for commercial purposes or any non-permitted use without the prior written permission from the Agency.

Recommendations for drug treatment presented in the Cancer Agency guidelines for a cancer site may not reflect provincial cancer drug funding. Please refer to the current Saskatchewan Cancer Agency drug formulary at www.saskcancer.ca for information on cancer drug listing and funding.

Benefits and risk of the proposed should be discussed with patient.

Participating in clinical trials is encouraged when available. Involvement of a multidisciplinary team is strongly recommended.

Background:
Nasopharyngeal carcinoma represents those carcinomas arising within the following borders: The posterior nasal aperture and septum (anterior), posterior pharyngeal mucosa (posterior), sphenoid (superior) and the oropharynx (inferior). While common some Asian countries, nasopharyngeal carcinoma is rare in North America, representing only a small fraction of head and neck cancers. [1] 65 -85% of patients present with lymph node metastases.[2] It also has a higher frequency of distant metastasis than other head and neck cancers. Etiology includes ethnicity, chemical exposures and EBV virus. More controversial is the association with smoking or alcohol. [3,4] The most common presentation is a neck mass (41%), followed by auditory symptoms (27%), nasal symptoms/bleeding (21%) and cranial nerve deficits (8%).[2]

Content:

WHO PATHOLOGIC CLASSIFICATION

- Type I: keratinizing squamous cell carcinoma - 20%
- Non-keratinizing carcinoma
  - Type II: differentiated non keratinizing carcinoma - 30-40%
  - Type III: undifferentiated non keratinizing carcinoma - 40-50%
- Basaloid squamous cell carcinoma

STAGING

AJCC 7th edition (2009):

Tumour

- T1 – Tumour confined to the nasopharynx or tumour extends to oropharynx and/or nasal cavity without parapharyngeal extension
- T2 – Tumour with parapharyngeal extension
- T3 - involves bony structures of skull base and/or paranasal sinuses
- T4 – Tumour with intracranial extension and/or involvement of cranial nerves, hypopharynx, orbit, or with extension to the infratemporal fossa/masticator space

Nodes

- N1 - Unilateral metastases in cervical lymph nodes, 6 cm or less in greatest dimension, above the supraclavicular fossa, and/or unilateral or bilateral retropharyngeal lymph nodes, 6 cm or less in greatest dimension
- N2 - Bilateral metastases in cervical lymph nodes, 6 cm or less in greatest dimension, above the supraclav fossa
- N3a – Metastases in a lymph node(s) greater than 6 cm
- N3b – Extension to the supraclavicular fossa

Overall stage

- I - T1 N0
- II - T1-T2 N1, T2 N0 (i.e. T2 or N1)
- III - T3 N0-2, or T1-3 N2 (i.e. T3 or N2)
- IVA - T4 N0-2
- IVB - N3
- IVC - M1

WORKUP

All patients with a suspected nasopharyngeal carcinoma should be seen in consultation and undergo a complete history and head & neck focused exam. An upfront multidisciplinary discussion including surgical, oncologic and diagnostic input is recommended. If available, office endoscopy performed on initial visit is useful to evaluate the entire upper aero-digestive tract.

Imaging includes CT and MRI from base of skull to clavicles, and chest x-ray. Chest CT is recommended for N2-3 disease. A PET scan may be necessary to further evaluate suspected regional or distant disease. With consideration of patient dentition, history of dental work and location of tumour; dental consultation may be indicated. Dental guards can be considered. A PEG tube prior to treatment is optional and not routinely necessary.

MANAGEMENT

T1N0M0

- Radiotherapy alone is associated with excellent outcomes in these patients [2]

T2-4N1-3M0

- Concurrent definitive chemo-radiotherapy (+/- adjuvant chemotherapy) [5,6]
  - Adjuvant chemotherapy is controversial [7]
- Induction chemotherapy followed by chemo-radiotherapy is an option, though not supported by robust level I evidence [10]. Please refer to systemic therapy guidelines.

M1

- Systemic chemotherapy [11-13]

IMRT technique using integrated boost can reduce toxicity and lead to improved salivary gland function[8], and is standard for head and neck cancers. Whether with chemotherapy or without, radiotherapy should include volumes defined by gross clinical and radiological disease and the bilateral neck. The nodal levels to cover and definition of intermediate vs low dose regions is at the discretion of the treating physician. Recommended neck coverage for the clinically node negative patient includes the bilateral levels II-V and retropharyngeal nodes. [8]
Example of standard RT prescription for nasopharyngeal carcinoma:

- 70Gy/33# to gross disease
- 59Gy/33# to intermediate risk areas
- 54Gy/33# to low risk areas

RECURRENT DISEASE

In cases of suspected recurrent disease, biopsy should be obtained when possible along with re-staging investigations including at minimum CT of the head and neck and chest x-ray or CT chest. The patient should be considered in a multidisciplinary setting. There is a paucity of rigorous randomized data on which to base any individual treatment decision.

Management decisions depend on the location (primary site, new primary site, neck or distant), the histology, time since initial diagnosis and prior therapies. The overall condition and functional status of the patient should also be carefully considered.
In general, surgical management is favored for patients previously treated with radiotherapy, and likewise those previously having undergone surgery alone should be considered for radiotherapy (+/- chemotherapy). Neck recurrence alone may be adequately treated with neck dissection if no previous surgical dissection. Various chemotherapeutic options have been investigated. [11-13] Cases involving previous treatment with surgery and radiotherapy present a more difficult clinical situation which can benefit from multidisciplinary consultation. Some patients may be eligible for repeat radiotherapy even to high doses. Small localized recurrences may be managed surgically in some cases. Recurrence at the base of skull or other difficult-to-access locations can benefit from external consultation at centers equipped with gamma knife or SBRT technologies. Other options include palliative radiotherapy or best supportive care.

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