Sponsored by

Malignant Odontogenic Tumours Histopathology Reporting Guide

Family/Last name

Given name(s)

Date of birth DD – MM – YYYY

Patient identifiers

Date of request DD – MM – YYYY

Accession/Laboratory number

Elements in **black text** are CORE. Elements in **grey text** are NON-CORE.

SCOPE OF THIS DATASET

SPECIMENS SUBMITTED (select all that apply)

- ○ Not specified
- ☐ Debulking/curettage
- ☐ Biopsy (excisional, incisional), *specify*
- ☐ Surgical resection, *specify*
- ☐ Neck (lymph node) dissection*, *specify*
- ☐ Other, *specify*

* *If a neck dissection is submitted, then a separate dataset is used to record the information.*

TUMOUR SITE (select all that apply)

- ○ Cannot be assessed

Laterality

- ☐ Left
- ☐ Right
- ☐ Midline
- ○ Laterality not specified

- ○ Mandible
  - ☐ Ramus
  - ☐ Condyle
  - ☐ Coronoid process
  - ☐ Body
  - ☐ Anterior
- ○ Maxilla
  - ☐ Nasal cavity/paranasal sinus (maxillary sinus)
  - ☐ Molar region alveolar process
  - ☐ Premolar region alveolar process
  - ☐ Incisor/canine region alveolar process
  - ☐ Zygomatic process
- ☐ Extraosseous, *specify site*
- ☐ Other, *specify including laterality*

TUMOUR DIMENSIONS (Note 1)

Maximum tumour dimension mm

Additional dimensions (largest tumour) mm x mm

- ○ Cannot be assessed, *specify*

HISTOLOGICAL TUMOUR TYPE (select all that apply) (Note 2)

(Value list from the World Health Organization Classification of Head and Neck Tumours (2017))

- ☐ Odontogenic carcinomas
  - ○ Ameloblastic carcinoma
  - ○ Primary intraosseous carcinoma, not otherwise specified (NOS)
  - ○ Sclerosing odontogenic carcinoma
  - ○ Clear cell odontogenic carcinoma
  - ○ Ghost cell odontogenic carcinoma
- ☐ Odontogenic carcinosarcoma
- ☐ Odontogenic sarcomas
- ☐ Other (hybrid etc.), *specify*
- ○ Cannot be assessed, *specify*

HISTOLOGICAL TUMOUR GRADE (Note 3)

(For primary intraosseous cell carcinoma only)

- ○ Not applicable
- ○ GX: Cannot be assessed
- ○ G1: Well differentiated
- ○ G2: Moderately differentiated
- ○ G3: Poorly differentiated
- ○ Cannot be assessed, *specify*

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NECROSIS (Note 4)

- ◯ Not identified
- ◯ Present
- ◯ Cannot be assessed, *specify*

EXTENT OF INVASION (Note 5)

- ◯ Not identified
- ◯ Entirely intraosseous
- ◯ Cortex perforated but extent limited by periosteum
- ◯ Infiltration into soft tissue beyond the periosteum
- ◯ Other, *specify*

PERINEURAL INVASION (Note 6)

- ◯ Not identified
- ◯ Present
- ◯ Cannot be assessed, *specify*

LYMPHOVASCULAR INVASION

- ◯ Not identified
- ◯ Present
- ◯ Cannot be assessed, *specify*

MARGIN STATUS (Note 7)

- ◯ Involved by tumour
  - Specify margin(s)/anatomical site, if possible
- ◯ Not involved by tumour
  - Distance from closest margin ____ mm
  - ◯ Distance not assessable
  - Specify site closest margin, if possible
- ◯ Cannot be assessed, *specify*

ANCILLARY STUDIES (Note 8)

- ◯ Not performed
- ◯ Performed, *specify*

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Scope

The dataset has been developed for the reporting of biopsy and resection specimens for malignant primary odontogenic tumours. Malignant neoplasms arising in the nasal cavity and paranasal sinuses, oral cavity, salivary glands, trachea, pharynx and larynx are dealt with in separate datasets. Bone, soft tissue and lymphoma protocols will be separately listed. In addition, neck dissections and nodal excisions are dealt with in a separate dataset, and this dataset should be used in conjunction, where applicable.

Dataset items should be completed taking into account all relevant information including clinical, pathological and radiological.

No staging elements are included because there is no staging system for malignant odontogenic tumours recommended by the Union for International Cancer Control (UICC) or American Joint Committee on Cancer (AJCC), although staging based on size criteria has been suggested.[1]

Evidence to support this dataset

This dataset is based almost exclusively on professional judgement because there is no high level evidence to support individual data items. Malignant odontogenic tumours are rare and published series are often not homogeneous by tumour type, stage or treatment, making conclusions about the value of individual items impossible. In general, those tumours that show aggressive histological features are more likely to be associated with poor survival, but this tumour group is characterised by unpredictability of behaviour; low grade tumours may recur or metastasize many years after excision. For all types, local recurrence and metastasis are poor prognostic features[2,3] and outcomes are relatively poor after local recurrence.[4-6] Published mortality rates are limited by short follow up. Ameloblastic carcinoma appears to carry a better prognosis than other types, for reasons that are unclear[7] though maxillary lesions behave worse than mandibular,[8] with up to one third of maxillary lesions seeding pulmonary metastases.

There are no validated grading systems for odontogenic tumours. For primary intraosseous squamous carcinoma, the conventional squamous carcinoma grade has some value.[9]

Margin status after surgical excision is thought to be the key prognostic feature[2,10-13] and the best evidence relates to ameloblastic carcinoma,[7] primary intraosseous carcinoma;[4,9] and clear cell carcinoma.[10] Tumour dimensions and localisation are important prognostic features. Carcinomas arising in or limited to cysts therefore carry a better prognosis than those with widespread infiltration.[14]

As with most head and neck surgical resections, clearances may be very small or inadequate and extension into soft tissues beyond the periosteum is usually associated with a significant risk of local recurrence. The prognosis is worse when incomplete excision is in the infratemporal fossa or base of skull areas and therefore the anatomical site of involved margins should be specified clearly.

The role for adjuvant or salvage radiotherapy remains to be defined. The literature does not provide useful information on radiotherapy indications or the intent when it has been used. Despite use to control incompletely excised malignant odontogenic tumours, its value often appears limited[10,11] but has support in large series[7] and is usually considered most effective as planned multimodality treatment.

Sclerosing odontogenic carcinoma is unusual. Despite extensive perineural spread, this carcinoma carries a relatively good prognosis.[15,16] Odontogenic sarcomas are overall of low grade and tend to show local recurrence rather than distant spread and thus carry a better prognosis than other types of sarcoma, but still have significant mortality and recurrence rates.[17,18]

Note 1 – Tumour dimensions (Core and Non-core)

Reason/Evidentiary Support

Due to the nature of odontogenic lesions, reference to any imaging or consultation with a radiologist is recommended and maximum tumour dimension may be determined by a combination of methods including macroscopy, specimen or clinical radiology and microscopy. Size criteria for possible staging have been suggested,[1] with smaller tumor size associated with a better overall survival.[7]

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Note 2 – Histological tumour type (Core)

Reason/Evidentiary Support

All odontogenic and maxillofacial bone tumours should be given a type based on the most recent edition of the World Health Organization (WHO) Classification of Head and Neck Tumours.[19]

WHO classification of odontogenic and maxillofacial bone tumours[a19]

Descriptor	ICD-O codes
Odontogenic carcinomas	
Ameloblastic carcinoma	9270/3
Primary intraosseous carcinoma NOS	9270/3
Sclerosing odontogenic carcinoma	9270/3
Clear cell odontogenic carcinoma	9341/3
Ghost cell odontogenic carcinoma	9302/3
Odontogenic carcinosarcoma	8980/3
Odontogenic sarcomas	9330/3

a The morphology codes are from the International Classification of Diseases for Oncology (ICD-O). Behaviour is coded /0 for benign tumours; /1 for unspecified, borderline, or uncertain behaviour; /2 for carcinoma in situ and grade III intraepithelial neoplasia; and /3 for malignant tumours.

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Note 3 – Histological tumour grade (Core)

Reason/Evidentiary Support

For primary intraosseous squamous carcinoma, the conventional squamous carcinoma grade is used.

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Note 4 – Necrosis (Core)

Reason/Evidentiary Support

Necrosis is not only a tool to aid in grading of tumors, but in many instances, the presence of necrosis helps to confirm a diagnosis of malignancy in odontogenic tumors in general. Thus, while large clinical series of these rare tumors are not available, there is strong support that reporting necrosis aids in diagnosis, grade and tumor classification.[2,20]

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Note 5 – Extent of invasion (Non-core)

Reason/Evidentiary Support

Use Figure 1 to define the sites involved. Extent of invasion is best assessed by a combination of macroscopic, microscopic and radiographic information.

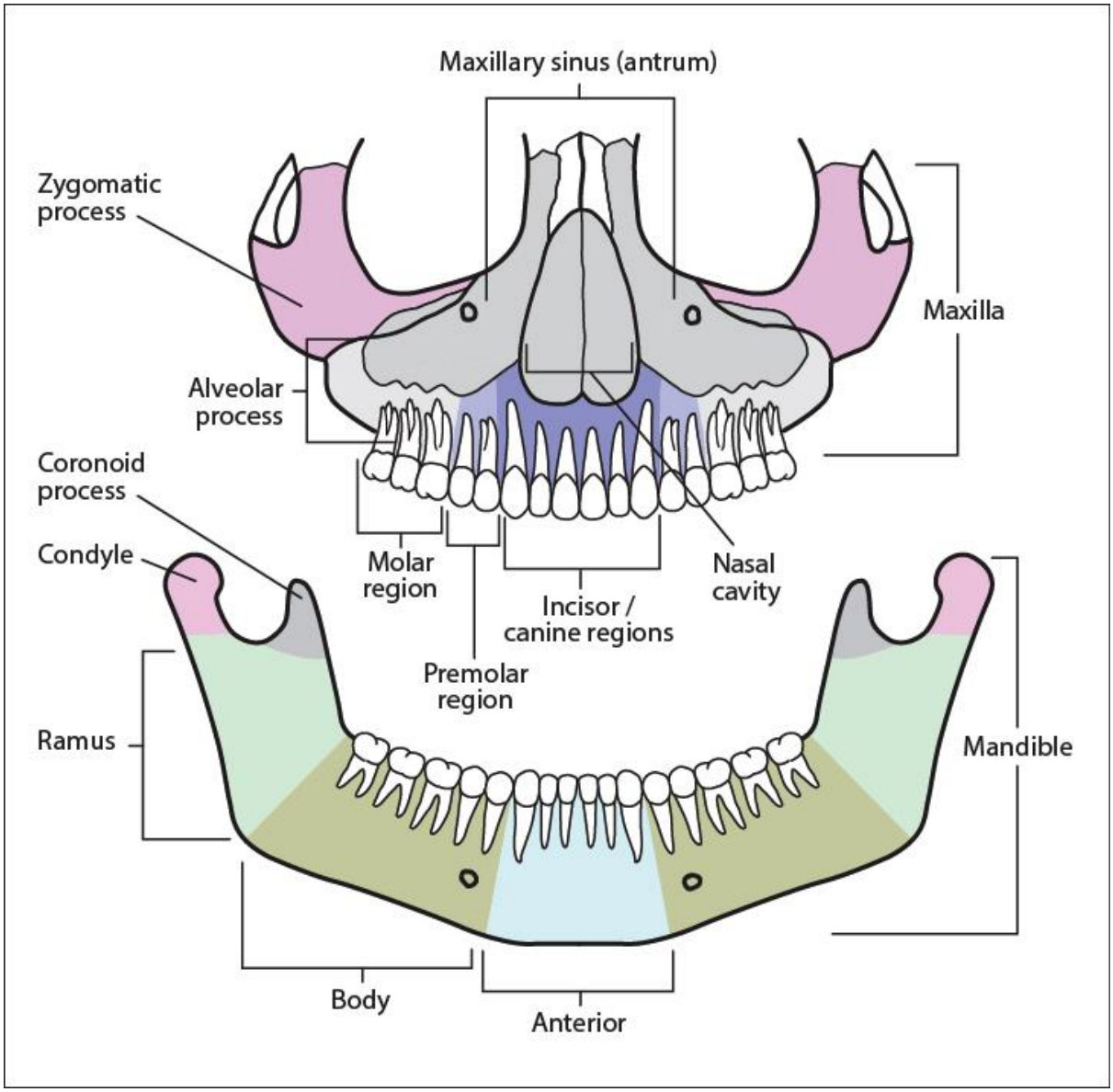

Figure 1. **Diagram showing anatomical sites for extent of involvement**

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Note 6 – Perineural invasion (Core)

Reason/Evidentiary Support

Note that the extensive perineural spread seen in sclerosing odontogenic carcinoma does not appear to be a poor prognostic feature.[15,16]

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Note 7 – Margin status (Core)

Reason/Evidentiary Support

Margin status is thought to be a key prognostic item. Surgical clearance is often by only a small margin and it is important to know whether the excision is marginal around a large part of the periphery of the tumour or just focally, as reoperation may be possible.

The prognosis is worse where an incomplete excision is located in the infratemporal fossa or base of skull areas and therefore the anatomical site of involved margins should be specified clearly.

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Note 8 – Ancillary studies (Non-core)

Reason/Evidentiary Support

There are a number of immunohistochemical and molecular studies that may be relevant. Some already have potential but unproven therapeutic benefit. Examples include; EWSR1 rearrangements in clear cell odontogenic carcinoma[21] and *BRAF* v600E mutation in ameloblastic carcinoma.[22] Such tests may also increase diagnostic certainty and, if performed, should be recorded.

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