Sponsored by

Ear and Temporal Bone 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

OPERATIVE PROCEDURE (select all that apply) (Note 1)

- ○ Not specified
- ☐ Biopsy (incisional, excisional, diagnostic sampling)
- ☐ Resection, *specify*
  - ☐ Temporal bone resection
  - ☐ Sleeve resection (cartilaginous portion of canal, including tympanic membrane)
  - ☐ Lateral temporal bone resection (sleeve and middle ear)
  - ☐ Radical external auditory canal resection
  - ☐ Subtotal temporal bone resection
  - ☐ Radical temporal bone resection (mastoidectomy, petrousectomy)
- ☐ Parotidectomy
- ☐ Neck (lymph node) dissection*, *specify*
- ☐ Other, *specify*

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

SPECIMENS SUBMITTED (select all that apply) (Note 2)

- ○ Not specified
- ○ Biopsy only
- ☐ Sleeve resection of temporal bone
- ☐ Lateral temporal bone
- ☐ Subtotal temporal bone resection
- ☐ Partial mastoidectomy with middle ear contents
- ☐ Radical mastoidectomy
- ☐ Parotidectomy (whether superficial and/or deep lobes)
- ☐ Neck dissection, *specify extent*
- ☐ Other, *specify*

TUMOUR SITE (select all that apply) (Note 3)

- ○ Cannot be assessed
- ☐ External auditory canal (EAC) — ☐ Left ☐ Right ○ Laterality not specified
- ☐ Middle ear — ☐ Left ☐ Right ○ Laterality not specified
- ☐ Temporal bone (including mastoid, petrous) — ☐ Left ☐ Right ○ Laterality not specified
- ☐ Inner ear — ☐ Left ☐ Right ○ Laterality not specified
- ☐ Other, *specify including laterality*

TUMOUR FOCALITY (Note 4)

- ○ Unifocal
- ○ Bilateral
- ○ Multifocal, *specify number of tumours in specimen*
- ○ Cannot be assessed, *specify*

TUMOUR DIMENSIONS (Note 5)

Maximum tumour dimension (largest tumour) mm

Additional dimensions (largest tumour) mm x mm

- ○ Cannot be assessed, *specify*

HISTOLOGICAL TUMOUR TYPE (select all that apply) (Note 6)

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

- ☐ Squamous cell carcinoma
- ☐ Ceruminous adenocarcinoma
  - ○ Ceruminous adenocarcinoma, not otherwise specified (NOS)
  - ○ Ceruminous mucoepidermoid carcinoma
  - ○ Ceruminous adenoid cystic carcinoma
- ☐ Ceruminous adenoma
  - ○ Ceruminous adenoma (NOS)
  - ○ Ceruminous pleomorphic adenoma
  - ○ Ceruminous syringocystadenoma papilliferum
- ☐ Aggressive papillary tumour
- ☐ Endolymphatic sac tumour
- ☐ Middle ear adenoma (carcinoid)
- ☐ Middle ear adenocarcinoma
- ☐ Meningioma (ectopic or direct extension)
- ☐ Vestibular schwannoma
- ☐ Paraganglioma (jugulotympanic glomus tumour)
- ☐ Other, *specify*
- ○ Cannot be assessed, *specify*

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HISTOLOGICAL TUMOUR GRADE (Note 7)

- ○ Not applicable
- ○ Low grade (well differentiated)
- ○ Intermediate grade (moderately differentiated)
- ○ High grade (poorly differentiated)
- ○ Cannot be assessed, *specify*

EXTENT OF INVASION** (select all that apply) (Note 8)

- ○ Not identified
- ☐ Bone and/or cartilage invasion (EAC)
- ☐ Jugular bulb
- ☐ Carotid artery invasion
- ☐ Dura
- ☐ Brain parenchyma invasion
- ☐ Parotid gland
- ☐ Temporomandibular joint (TMJ)
- ☐ Soft tissue involvement
- ☐ Skin involvement
- ☐ Nerve invasion, *specify nerve if possible (e.g. facial nerve, tympanic nerve, glossopharyngeal nerve, lesser petrosal nerve, greater petrosal nerve)*
- ☐ Other, *specify*
- ○ Cannot be assessed, *specify*

** *Invasion into any of these anatomical structures may be a clinical/surgical and/or imaging observation and/or histology finding(s).*

BONE/CARTILAGE INVASION (Note 9)

- ○ Not identified
- ○ Present
  - ○ Clinical observation and/or imaging
  - ○ Histologic
- ○ Cannot be assessed, *specify*

PERINEURAL INVASION (Note 10)

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

LYMPHOVASCULAR INVASION (Note 11)

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

MARGIN STATUS (Note 12)

- ○ Involved by invasive carcinoma
  - Specify margin(s), if possible
- ○ Not involved by invasive carcinoma
  - Distance of tumour from closest margin ____ mm
  - ○ Distance not assessable
  - Specify closest margin, if possible
    - ☐ Skin
    - ☐ Soft tissue
    - ☐ Bone
    - ☐ Parotid gland
- ○ Cannot be assessed, *specify*

COEXISTENT PATHOLOGY (select all that apply) (Note 13)

- ○ None identified
- ☐ Chronic otitis media
- ☐ Cholesteatoma
- ☐ Osteomyelitis (acute, chronic)
- ☐ Other, *specify*

ANCILLARY STUDIES (Note 14)

- ○ Not performed
- ○ Performed, *specify*

PATHOLOGICAL STAGING (Note 15)

TNM Descriptors (only if applicable) (select all that apply)

- ☐ m - multiple primary tumours
- ☐ r - recurrent
- ☐ y - post-therapy

Primary tumour (pT)*

- ○ Not applicable
- ○ T1 Tumour limited to the EAC without bony erosion or evidence of soft tissue involvement
- ○ T2 Tumour with limited EAC bone erosion (not full thickness) or limited (<0.5 cm) soft tissue involvement
- ○ T3 Tumour eroding the osseous EAC (full thickness) with limited (<0.5 cm) soft tissue involvement, or tumour involving the middle ear and/or mastoid
- ○ T4 Tumour eroding the cochlea, petrous apex, medial wall of the middle ear, carotid canal, jugular foramen, or dura, or with extensive soft tissue involvement (>0.5 cm), such as involvement of TMJ or styloid process, or evidence of facial paresis

*** *Note that the results of lymph node/neck dissection are derived from a separate dataset.*

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Scope

The dataset has been developed for the reporting of resection and biopsy specimens of the ear and temporal bone. It includes ONLY primary tumours of the external auditory canal, middle and inner ear, including both benign and malignant entities (specifically due to anatomic confines and management alternatives which may require significant, destructive or disfiguring surgery).

By definition, all malignancies of the external ear (pinna, concha, scaphoid, lobe, etc., such as squamous cell carcinoma, basal cell carcinoma, atypical fibroxanthoma, Merkel cell carcinoma and melanoma) are separately covered by the dermatopathology datasets.

Neck dissections and nodal excisions are dealt with in a separate dataset, and this dataset should be used in conjunction, where applicable.

For bilateral tumours, a separate dataset should be completed for each tumour.

Note 1 – Operative procedure (Core)

Reason/Evidentiary Support

The anatomy and surgical interventions of the ear and temporal bone are complex, with unfamiliar terminology frequently used (see Figure 1). Thus, it is absolutely critical to maintain open communication with the treating surgeon, oncologist, dermatologist and radiologist with respect to exact anatomic site of involvement, tumour laterality, and specific operative procedures or landmarks identified to yield the most accurate information.[1-4]

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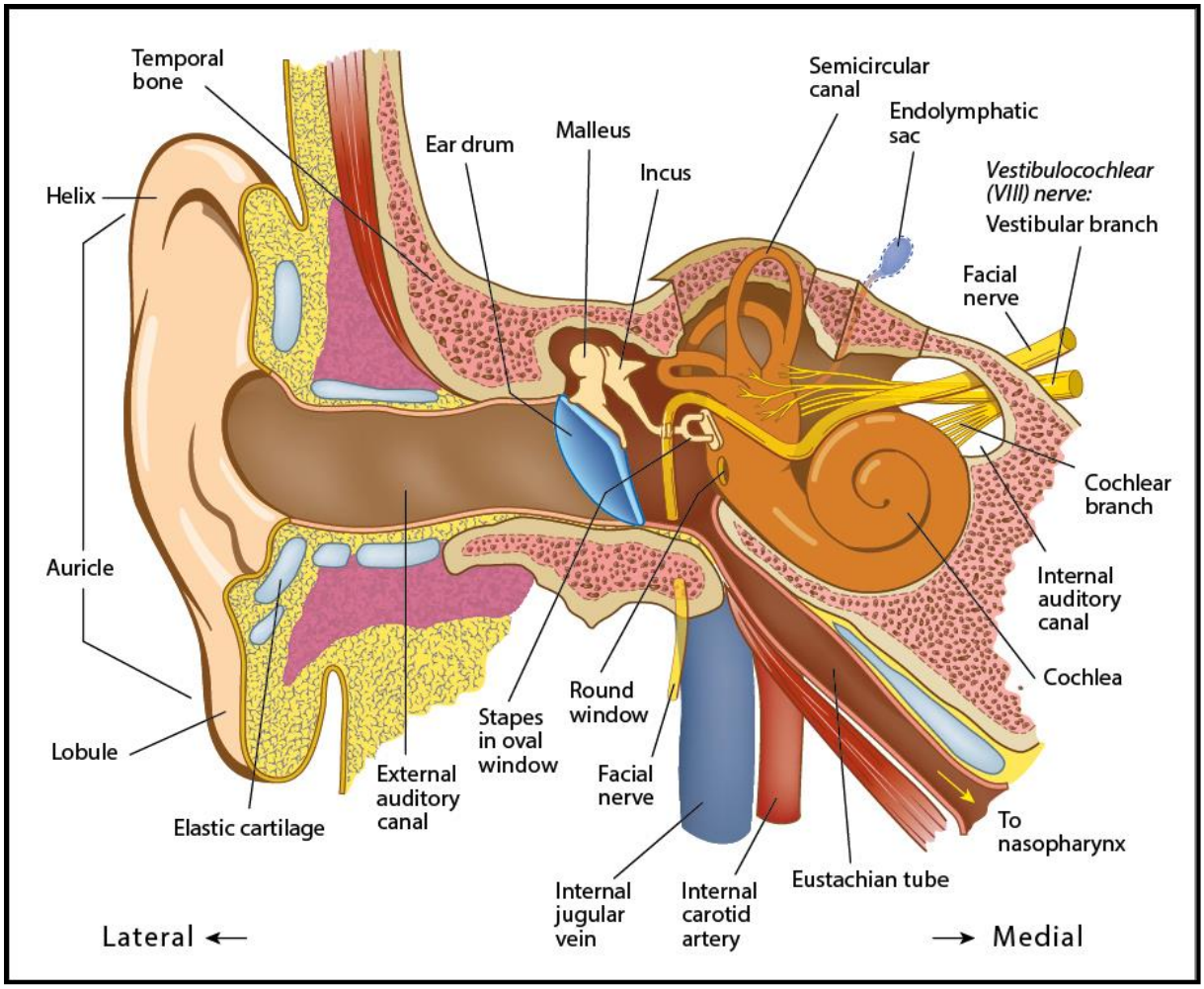

Figure 1. Diagram of ear and temporal bone anatomic landmarks

Note 2 – Specimens submitted (Core)

Reason/Evidentiary Support

In light of the complex anatomy and often unfamiliar surgical interventions of the ear and temporal bone, it is imperative to obtain information about the exact anatomic site of involvement, tumour laterality, and specific operative procedures or landmarks identified to yield the most accurate information.[5]

'Not specified' should be used rarely and only after good faith effort has been employed to obtain the requisite information.

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Note 3 – Tumour site (Core)

Reason/Evidentiary Support

It is important to document the exact site of the tumour, as tumour location is correlated with patient outcome. As an example, patients with middle ear squamous cell carcinomas have a worse outcome than patients with squamous cell carcinoma of the external auditory canal.[1,3,11,12]

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Note 4 – Tumour focality (Non-core)

Reason/Evidentiary Support

The identification of bilateral tumours, especially in the setting of endolymphatic sac tumours,[6,7] paraganglioma,[8,9] acoustic/vestibular Schwannoma[10] and meningioma[10] increases the potential discovery of inherited or syndrome associated disease.

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Note 5 – Tumour dimensions (Core and Non-core)

Reason/Evidentiary Support

The single greatest tumour dimension, using macroscopic and/or microscopic measurements, should be used to determine the most accurate extent of tumour. In biopsy samples, it may be underestimated. Thus, to be as thorough as possible, the documentation of the tumour dimension may require additional clinical or imaging information to yield this value.

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Note 6 – Histological tumour type (Core)

Reason/Evidentiary Support

The types of ear and temporal bone primary tumours are limited. Few cases have been reported for several specific tumour categories, and thus prognostication about each specific tumour type is limited, at best. Overall, the most common tumour type is squamous cell carcinoma, and it is known to have the worst patient outcome.[15,17,25,26] When adenoid cystic carcinoma and mucoepidermoid carcinoma are the ceruminous adenocarcinoma type, parotid gland evaluation is recommended to exclude origin from the parotid gland with secondary invasion into the external canal.[13,27]

World Health Organization (WHO) classification of tumours of the ear[a28]

Descriptor	ICD-O codes
Squamous cell carcinoma	8070/3
Ceruminous adenocarcinoma	8420/3
Ceruminous adenoid cystic carcinoma	8200/3
Ceruminous mucoepidermoid carcinoma	8430/3
Ceruminous adenoma	8420/0
Aggressive papillary tumour	8260/1
Endolymphatic sac tumour	8140/3
Vestibular schwannoma	9560/0
Meningioma	9530/0
Middle ear adenoma	8140/0

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 7 – Histological tumour grade (Core)

Reason/Evidentiary Support

Generally, grades are applied to squamous cell carcinoma or salivary gland primaries only, while other tumour types for the most part do not have tiered grading systems (such as ceruminous adenocarcinoma, middle ear adenoma, endolymphatic sac tumours or schwannoma). Poorly differentiated tumours portend a poor patient survival.[29] The same grading of central nervous system meningiomas is applied to ear and temporal bone, realising that >95% are WHO grade 1 tumours.

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Note 8 – Extent of invasion (Core)

Reason/Evidentiary Support

The extent of invasion may need to be evaluated by imaging or during intraoperative assessment, as histologic identification of these structures may not be feasible. If there is involvement of any of these recognized structures, documentation will provide prognosis and management information. For example, patients with primary ear and temporal bone carcinoma with parotid gland involvement have a worse prognosis than patients without parotid gland involvement.[13] If there is advanced disease clinically, then parotid gland resection is generally recommended.[13] Similarly, when there is destructive cartilage and/or bone invasion, the patients tend to have a worse prognosis.[14-18] The macroscopic and microscopic extent of tumour frequently overlap. Thus, invasion "microscopically" into any of these structures is for the most part not recognized, unless the part is specifically stated to be from the site. Thus, on histologic examination, you may not recognize the specific structure. Therefore, correlation between macroscopic and microscopic findings is encouraged to yield the most meaningful findings.[4,16,25,27,28] As an example, patients who exhibit dura involvement, will have a worse patient outcome.[18,28]

Due to the type of samples, tumour budding or tentacular pattern of invasion may not be histologically identified. However, if this type of growth is seen in squamous cell carcinoma, patients tend to have a shorter survival.[29-31]

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Note 9 – Bone/cartilage invasion (Core)

Reason/Evidentiary Support

Bone and/or cartilage invasion may be a macroscopic feature, sometimes not seen on histology sections due to the nature of the clinical sampling performed. However, it is recommended that a histologic section through the involved bone should be performed to obtain histologic evidence of the extent of bone and/or cartilage involvement. In general, stage correlates with bone and/or cartilage invasion, with high stage patients more frequently showing bone invasion than low stage patients. Further, patients with bone and/or cartilage invasion will usually have a worse prognosis and require more extensive treatment than patients without bone invasion.[16,30]

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Note 10 – Perineural invasion (Core)

Reason/Evidentiary Support

If the biopsy is very small with only tumour included, it may be prudent to use "cannot be assessed" in order to alert the clinician that perineural invasion cannot be reliably excluded in the sampled

material. Patients who manifest perineural invasion, especially if it is identified in large or named nerves (such as lesser petrosal nerve, tympanic nerve), have a worse clinical outcome, irrespective of the tumour type or tumour grade.[19,31]

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Note 11 – Lymphovascular invasion (Core)

Reason/Evidentiary Support

By inference, lymphovascular invasion is thought to be associated with a worse clinical outcome. However, in ear and temporal bone tumours, this finding has not been independently evaluated in prospective or prognostic studies.

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Note 12 – Margin status (Core)

Reason/Evidentiary Support

The best overall outcomes for tumours of ear and temporal bone are achieved when margins are negative. In general, mucosal/epithelial margins are reported, but bone and soft tissue margins carry similar prognostic value, and thus should also be reported, especially as the deep margins (bone and soft tissue) are often more clinically significant than superficial margins (skin). Tumours which are meticulously debulked have the best long term outcome.[4,11,17,18,21,26,32-34]

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Note 13 – Coexistent pathology (Non-core)

Reason/Evidentiary Support

Management may be complicated by coexistent pathology. Patient with otitis media generally show a poor survival,[4] but if there is acute or chronic osteomyelitis, options for radiation and chemotherapy may be limited.[35,36]

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Note 14 – Ancillary studies (Non-core)

Reason/Evidentiary Support:

In most patients, further studies are not required for the diagnosis. However, additional molecular testing may be of benefit, especially in syndrome associated, bilateral, or uncommon tumour presentations. It is true that in most patients, "further studies" are not required. However, not infrequently adjuct immunohisotochemistry (IHC) is required to differentiate among tumour types especially in limited sampling, frequently affected by distortional changes that alter the "typical" histology, rendering the case problematic to diagnose without IHC. Ancillary tests rarely may be required to identify the primary site of metastatic disease.

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Note 15 – Pathological staging (Core)

Reason/Evidentiary Support

There is no standardised staging system for this anatomic site, although it has been suggested by several groups. However, staging is still of value in standardizing therapy for these various unusual tumours. The T staging is most significant for squamous cell carcinoma and for salivary gland-type tumours, particularly of the external auditory canal and middle ear.[11,12,17,18,20,37-39] Pathological staging has not been well developed for inner ear tumours, such as endolymphatic sac tumour, where clinical staging may be more appropriate.[40] In inner ear cases, it is probably more important to make certain that a clinical (c-stage) is accurately determined, than necessarily being definitive about a pathological (p-stage). The studies used as a guide are retrospective where the patient outcomes were not available, primarily used as a guide for therapy rather than prognosis.

Overall, there is a poor prognosis when lymph node metastases are identified, correlating to advanced stage, whether in the cervical lymph nodes or those of the parotid gland parenchyma.[4,12,24,27,39,41]

It is important with parotid gland lesions to interpret direction extension as part of the pT stage, being careful to interpret direct extension "into" a lymph node separately from metastasis "to" a lymph node that shows extracapsular extension. Tumour associated lymphoid proliferation is an important distinction to make, as this is a reaction to the neoplasm rather than representing a true lymph node (subcapsular sinus, lymph node capsule, sinus histiocytosis, and medullary zone). Metastases to an intraparotid lymph node that shows extranodal extension is associated with a worse outcome when compared to patient with extranodal extension in cervical lymph nodes only of cutaneous squamous cell carcinoma.[42,43]

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