

A guide to Pheochromocytoma and Paraganglioma Histopathology Reporting



Includes the  International Collaboration on Cancer reporting dataset denoted by *

Clinical details		Macroscopic findings cont.	
S1.02	Clinical info. on request form (complete as narrative or use the structured format below)	Text OR Information not provided*	S2.09 * Tumour dimensions Note: Repeat tumour identification and dimension for each tumour identified
	* Hormone status	Biochemically functioning	Can't be assess'd, specify OR Text AND __ mm maximum dimension (largest tumour)
	* Imaging findings	Text	* Additional dimensions (largest tumour)
	* Relevant biopsy/ cytology results	Text	__ mm x __ mm
	* Previous therapy (including pre-operative embolization, chemotherapy, radiotherapy, targeted therapy, immunotherapy)	Text	S2.10 * Appearance of uninvolved adrenal gland
	* Relevant familial history	Text	Not applicable OR
	* Presence of endocrine or other tumours	Text	* Medullary nodules (microphaeochromocytoma) (<10 mm)
	* Germline mutation or familial syndrome	Text	Present Absent Indeterminate Can't be assess'd, specify
G1.01	Copy to doctor	Text	* Diffuse hyperplasia
S1.03	Pathology accession number	Text	Present Absent Indeterminate Can't be assess'd, specify
S1.04	Principal clinician	Text	S2.11 * Specimen integrity
G1.02	Other clinical information received	Text	Not applicable Specimen intact Fragmented specimen Can't be assess'd, specify
Macroscopic findings			S2.12 Block identification key
S2.01	Specimen labelled as	Text	Text
S2.02	* Clinical information	Text	G2.02 Other macro. comment
S2.03	* Operative procedure	See p2	Text
S2.04	* Specimen(s) submitted	See p2	
S2.05	Specimen weight	__ g	Microscopic findings
G2.01	Specimen dimensions	__ x __ x __ mm	S3.01 * Histological tumour type
S2.06	* Tumour focality	Indeterminate Can't be assess'd, specify Unifocal Multiple	See p2
	* Number of tumours in same organ	__	G3.01 * Adverse features
	* Number of tumours in separate organs	__	See p2
S2.07	* Tumour site	See p2	S3.02 * Extent of invasion
S2.08	* Macroscopic appearance of lesion(s)	Repeat for each primary tumour	See p2
	Location	Medulla Indeterminate Other	S3.03 Extension into adipose tissue (for phaeochromocytoma)
	Details	Text	Absent Present <i>Distance</i> __ mm
	Borders	Encapsulated Infiltrative	S3.04 * Lymphovascular invasion
	Description	Text	See p2
	Distance to nearest excision margin	__ mm	S3.05 * Mitotic count
			**Note: 2 mm ² approximates 10 high power fields on some microscopes.
			__ mitotic figures/2 mm ² ** Can't be assessed, specify AND/OR
			Atypical mitoses
			Can't be assess'd Absent Present
			Ki-67 proliferation index
			__ % OR Can't be assess'd, specify
			S3.06 Non-tumour appearance
			Unremarkable OR Not identified/not assessable OR Adrenal cortical atrophy Hyperplasia Cortical nodules Medullary hyperplasia/nodule

Microscopic findings cont.		Synthesis and overview	
G3.02	Malignant potential	Text	S5.01 *PATHOLOGICAL STAGING See p3
	Scoring system (e.g. PASS, Weiss etc.)		S5.02 Year and edition of staging system Text
	Score for malignant potential	—	G5.01 Diagnostic summary Text
S3.07	*Margin status	Not involved (R0), specify distance to closest margin Involved, specify R1 (microscopic) or R2 (macroscopic) extent and location of involvement if possible Cannot be assessed, specify	Include: Specimen submitted; Histological tumour type; Diameter of largest tumour; Completeness of excision; Tumour stage.
	*Distance to closest margin	— mm	S5.03 Overarching comment Text
	*Closest margin	Text	G5.02 Edition/version of RCPA protocol Text
	*Extent of margin involvement	— mm R1 (microscopic), specify if possible — mm R2 (macroscopic), specify if possible	S2.03 Operative procedure
	Location of involved margin	Text	Not specified
S3.08	*Lymph node status	No nodes submitted or found OR	OR (Select all that apply)
	Lymph node biopsy site(s)	Text	<ul style="list-style-type: none"> • Biopsy (core needle, incisional, excisional), specify • Open resection, specify procedure, including other organs if present (e.g. adrenal resection and liver biopsy) • Laproscopic • Organ-sparing • Other (e.g. conversion, laproscopic, open), specify
		Not involved Involved	S2.04 Specimen(s) submitted
	Number of lymph nodes examined	—	Not specified*
	Number of positive lymph nodes	—	OR (Select all that apply)
	*Extranodal extension (ENE)	Not identified Present Cannot be determined	<ul style="list-style-type: none"> • Adrenal tumour <ul style="list-style-type: none"> o Left o Right • Biopsy tissue, specify site(s) and laterality • Lymph nodes, specify biopsy/dissection, site(s) and laterality • Other (e.g. right neck mass, midline abdominal mass), specify site(s) and laterality
	*Number of nodes with ENE	—	S2.07 Tumour site
	Maximum dimension of largest lymph node metastasis	— mm	Not specified
S3.09	*Distant metastases	Not identified Not assessed Present, specify site(s)	OR (Select all that apply)
G3.03	Coexistent abnormalities	Absent Present, specify	<ul style="list-style-type: none"> • Adrenal tumour <ul style="list-style-type: none"> o Left o Right • Other abdominal or pelvic <ul style="list-style-type: none"> o Paraaortic o Urinary bladder o Other, specify • Head and neck <ul style="list-style-type: none"> o Carotid body <ul style="list-style-type: none"> • Left • Right o Middle ear (jugulotympanic) <ul style="list-style-type: none"> • Left • Right o Vagal <ul style="list-style-type: none"> • Left • Right o Laryngeal <ul style="list-style-type: none"> • Left • Right o Other, specify site(s) and laterality
S3.10	Distant metastases	Not identified Info not available Present, specify site(s)	S3.01 Histological tumour type
G3.04	Additional microscopic comment	Text	<ul style="list-style-type: none"> • Pheochromocytoma • Extra-adrenal paraganglioma • Composite pheochromocytoma <ul style="list-style-type: none"> o Neuroblastoma, specify o Ganglioneuroma, specify o Malignant peripheral nerve sheath tumour, specify • Composite paraganglioma <ul style="list-style-type: none"> o Neuroblastoma, specify o Ganglioneuroma, specify o Malignant peripheral nerve sheath tumour, specify • Other, specify
Ancillary findings			
G4.01	*Ancillary studies	See p3	

G3.01 Adverse features

Histological features

(Select all that apply)

- Growth pattern
 - o Large and irregular nests
 - o Diffuse
 - o Pseudorosette (even focal)
- Cellularity
 - o Moderate (150-250 cells/U)
 - o High (>250 cells/U)
 - o Indeterminate
- Cytological features
 - o Spindle cells
- Necrosis
 - o Comedonecrosis
 - o Other, *specify*
- Other, *specify*

Other features

- Extra-adrenal abdominal or mediastinal location
- Size >50 mm
- Negative staining for SDHB
- Biochemical test showing high level methoxytyramine

Other histopathological features

- Adenoma
- Hyperplasia
- Other, *specify*

Other

S3.02 Extent of invasion

- Cannot be assessed
- Capsular invasion not present

OR (Select all that apply)

- Microscopic transcapsular penetration of tumour capsule within an organ
- Microscopic transcapsular penetration of organ capsule
- Invasion into peritumoural soft tissue
- Invasion into adjacent structure(s)/organ(s), *specify*

S3.04 Lymphovascular invasion

Not specified*

OR (Select all that apply)

- Present
 - o Periadrenal or peritumoural for extra-adrenal tumours, *specify*
 - Intracapsular
 - Extracapsular
 - o Adrenal vein
 - o Vena cava
 - o Other (e.g. adrenal central vein and tributaries), *specify*

S5.01 Stage & stage group##

Suffixes

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

Primary Tumour (T)

- TX Primary tumour cannot be assessed
- T1 Pheochromocytoma <5 cm in greatest dimension, no extra-adrenal invasion
- T2 Pheochromocytoma ≥5 cm or paraganglioma-sympathetic of any size, no extra-adrenal invasion
- T3 Tumour of any size with invasion into surrounding tissues (e.g. liver, pancreas, spleen, kidneys)

Phaeochromocytoma: within adrenal gland

Paraganglioma sympathetic: functional

Paraganglioma parasympathetic: nonfunctional, usually in the head and neck region

*Note: Parasympathetic paraganglioma are not staged because they are largely benign.

Regional lymph node (N)

- NX Regional lymph nodes cannot be assessed
- N0 No lymph node metastasis
- N1 Regional lymph node metastasis

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G4.01 Ancillary studies

Not performed

OR

(Select all that apply):

- Immunohistochemistry performed
 - o Synaptophysin, *specify result*
 - o S-100, *specify result*
 - o SDHB, *specify result*
 - o Tyrosine hydroxylase, *specify result*
 - o Other, *specify*
- Molecular testing performed, *specify result(s) if available*
- Other, *specify*