Soft Tissue Sarcoma
Histopathology Reporting Guide
Resection Specimens

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.

☐ indicates multi-select values ◯ indicates single select values

SCOPE OF THIS DATASET

CLINICAL INFORMATION (select all that apply) (Note 1)

- ◯ Information not provided
- ☐ Familial syndrome, *specify*
- ☐ Multifocal disease, *specify*
- ☐ Other, *specify*

NEOADJUVANT THERAPY (Note 2)

- ◯ Information not provided
- ◯ Not administered
- ◯ Administered
  - ☐ Neoadjuvant chemotherapy
  - ☐ Neoadjuvant radiotherapy
  - ☐ Other, *describe*

OPERATIVE PROCEDURE (select all that apply) (Note 3)

- ◯ Not specified
- ☐ Resection, *specify if known*
- ☐ Amputation, *specify type*
- ☐ Other, *specify*

TUMOUR SITE (select all that apply) (Note 4)

- ◯ Not specified
- ☐ Cutaneous, *specify deeper extension if known*
- ☐ Head and neck, *specify site if known*
- ☐ Trunk, *specify site and depth if known*
- ☐ Extremities, *specify site and depth if known*

  Specify laterality
  - ◯ Left
  - ◯ Right
  - ◯ Not specified
- ☐ Abdominal/pelvic visceral organ(s), *specify site if known*
- ☐ Thoracic visceral organ(s), *specify site if known*
- ☐ Thoracic soft tissue (including mediastinum), *specify site if known*
- ☐ Retroperitoneum (including paratesticular), *specify site if known*
- ☐ Pelvis, *specify site if known*
- ☐ Other somatic or visceral site, *specify site if known*

Version 1.0 Published April 2021 ISBN: 978-1-922324-19-1

TUMOUR DEPTH – TISSUE PLANE (select all that apply) (Note 5)

- ◯ Cannot be assessed
- ◯ Not known
- ☐ Cutaneous
- ☐ Subcutaneous
- ☐ Subfascial/muscle
- ☐ Bone
- ☐ Abdominal/retroperitoneal
- ☐ Other, *specify*

TUMOUR DIMENSIONS (Note 6)

Maximum tumour dimension ____ mm

Additional dimensions ____ mm x ____ mm

OR

- ◯ No identifiable tumour (e.g., after preoperative therapy)
- ◯ Cannot be assessed, *specify*

HISTOLOGICAL TUMOUR TYPE (Note 7)

(Value list based on the World Health Organization Classification of Soft Tissue and Bone Tumours (2020))

- ◯ No residual tumour
- ◯ Atypical lipomatous tumour
- ◯ Liposarcoma, well-differentiated, *specify type*
- ◯ Dedifferentiated liposarcoma
- ◯ Myxoid liposarcoma
- ◯ Pleomorphic liposarcoma
- ◯ Dermatofibrosarcoma protuberans NOS
  - ◯ Dermatofibrosarcoma protuberans, fibrosarcomatous
- ◯ Solitary fibrous tumour NOS
- ◯ Inflammatory myofibroblastic tumour
  - ◯ Epithelioid inflammatory myofibroblastic sarcoma
- ◯ Myxoinflammatory fibroblastic sarcoma
- ◯ Infantile fibrosarcoma
- ◯ Fibrosarcoma NOS
- ◯ Myxofibrosarcoma
  - ◯ Epithelioid myxofibrosarcoma
- ◯ Low grade fibromyxoid sarcoma
- ◯ Sclerosing epithelioid fibrosarcoma
- ◯ Plexiform fibrohistiocytic tumour
- ◯ Giant cell tumour of soft parts
- ◯ Haemangioendothelioma, *specify type*[a]
- ◯ Kaposi sarcoma, *specify epidemiologic type*
- ◯ Epithelioid haemangioendothelioma NOS
  - ◯ Epithelioid haemangioendothelioma with *WWTR1-CAMTA1* fusion
  - ◯ Epithelioid haemangioendothelioma with *YAP1-TFE3* fusion
- ◯ Angiosarcoma
- ◯ Glomus tumour, malignant
- ◯ Leiomyosarcoma NOS
- ◯ Embryonal rhabdomyosarcoma NOS
  - ◯ Embryonal rhabdomyosarcoma, pleomorphic
- ◯ Alveolar rhabdomyosarcoma
- ◯ Pleomorphic rhabdomyosarcoma NOS
- ◯ Spindle cell rhabdomyosarcoma
- ◯ Osteosarcoma, extraskeletal
- ◯ Malignant peripheral nerve sheath tumour NOS
  - ◯ Malignant peripheral nerve sheath tumour, epithelioid
- ◯ Malignant melanotic nerve sheath tumour
- ◯ Atypical fibroxanthoma
- ◯ Angiomatoid fibrous histiocytoma
- ◯ Ossifying fibromyxoid tumour NOS
- ◯ Synovial sarcoma, *specify type*
- ◯ Epithelioid sarcoma
  - ◯ Proximal or large cell epithelioid sarcoma
  - ◯ Classic epithelioid sarcoma
- ◯ Alveolar soft part sarcoma
- ◯ Clear cell sarcoma of soft tissue
- ◯ Extraskeletal myxoid chondrosarcoma
- ◯ Desmoplastic small round cell tumour
- ◯ Rhabdoid tumour of soft tissue
- ◯ Perivascular epithelioid tumour, malignant
- ◯ Myoepithelial carcinoma
- ◯ Mixed tumour, malignant, NOS
- ◯ Undifferentiated sarcoma
- ◯ Spindle cell sarcoma, undifferentiated
- ◯ Pleomorphic sarcoma, undifferentiated
- ◯ Round cell sarcoma, undifferentiated
- ◯ Ewing sarcoma
- ◯ Other round cell sarcoma, *specify*
- ◯ Sarcoma of uncertain type, *specify whether unclassifiable or requires additional testing*
- ◯ Soft tissue tumour of uncertain biologic potential, *specify type where known*
- ◯ Other, *specify*

Diagnosis based on (select all that apply)

- ◯ Not applicable
- ☐ Morphology
- ☐ Immunohistochemistry
- ☐ Molecular testing

[a] *e.g., Kaposiform, Retiform, Pseudomyogenic, Composite or Papillary Intralymphatic angioendothelioma.*

ISBN: 978-1-922324-19-1

HISTOLOGICAL TUMOUR GRADE[b] (Note 8)

- ◯ Grade 1
- ◯ Grade 2
- ◯ Grade 3
- ◯ Cannot be assessed, *specify*

[b] *Histological tumour grade is required only for specific histotypes – refer to Note, Table 3.*

MITOTIC COUNT[c] (Note 9)

/2 mm^2

◯ Cannot be assessed

[c] *10 HPFs approximates to 2 mm^2 on most modern microscopes, but the number of fields to be counted to encompass 2 mm^2 should ideally be calculated on individual microscopes – refer to Note 8, Table 3.*

NECROSIS[d] (Note 10)

- ◯ Not identified
- ◯ Present

%

[d] *Necrosis is required for those sarcomas that are gradable – refer to Note 8, Table 3.*

LYMPHOVASCULAR INVASION (Note 11)

- ◯ Not identified
- ◯ Present
- ◯ Indeterminate

RESPONSE TO NEOADJUVANT THERAPY (Note 12)

- ◯ No prior treatment
- ◯ No response
- ◯ Response
  - % viable tumour %
  - % necrosis %
  - % therapy-induced tissue changes (e.g., fibrosis or hyalinization) %
  - % of cell differentiation (e.g., myxoid liposarcoma) %
- ◯ Cannot be assessed, *explain reasons*

MARGIN STATUS (Note 13)

- ◯ Cannot be assessed
- ◯ Not involved (R0)
  - Distance of tumour from closest margin mm
  - Specify closest margin, if possible
  - Specify distance to other margin(s), if relevant
- ◯ Microscopically involved (R1)
  - Specify margin(s), if possible
- ◯ Macroscopically involved (R2)
  - Specify margin(s), if possible

LYMPH NODE STATUS (Note 14)

- ◯ Cannot be assessed
- ◯ No nodes submitted or found

Number of lymph nodes examined

- ◯ Not involved
- ◯ Involved
  - Number of involved lymph nodes
  - ◯ Number cannot be determined

COEXISTENT PATHOLOGY (Note 15)

- ◯ None identified
- ◯ Present
  - ☐ Neoplastic pathology, *specify*
  - ☐ Non-neoplastic pathology, *specify*
  - ☐ Other, *specify*

Version 1.0 Published April 2021 ISBN: 978-1-922324-19-1

ANCILLARY STUDIES (Note 16)

- ◯ Not performed
- ◯ Performed
  - ☐ Immunohistochemistry, *specify test(s) and result(s)*
  - ☐ Molecular findings, *specify test(s) and result(s)*
  - ☐ Other, *specify test(s) and results*

HISTOLOGICALLY CONFIRMED DISTANT METASTASES (Note 17)

- ◯ Not identified
- ◯ Present, *specify site(s)*

PATHOLOGICAL STAGING (UICC TNM 8th edition)[e] (Note 18)

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

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

Primary tumour (pT)

- ◯ Inadequate specimen for assessment
- ◯ TX Primary tumour cannot be assessed
- ◯ T0 No evidence of primary tumour

EXTREMITY AND SUPERFICIAL TRUNK

- ◯ T1 Tumour 5 cm or less in greatest dimension
- ◯ T2 Tumour more than 5 cm but no more than 10 cm in greatest dimension
- ◯ T3 Tumour more than 10 cm but no more than 15 cm in greatest dimension
- ◯ T4 Tumour more than 15 cm in greatest dimension

RETROPERITONEUM

- ◯ T1 Tumour 5 cm or less in greatest dimension
- ◯ T2 Tumour more than 5 cm but no more than 10 cm in greatest dimension
- ◯ T3 Tumour more than 10 cm but no more than 15 cm in greatest dimension
- ◯ T4 Tumour more than 15 cm in greatest dimension

HEAD AND NECK

- ◯ T1 Tumour 2 cm or less in greatest dimension
- ◯ T2 Tumour more than 2 cm but no more than 4 cm in greatest
- ◯ T3 Tumour more than 4 cm in greatest dimension
  - ◯ T4a Tumour invades the orbit, skull base or dura, central compartment viscera, facial skeleton, and or pterygoid muscles
  - ◯ T4b Tumour invades the brain parenchyma, encases the carotid artery, invades prevertebral muscle or involves the central nervous system by perineural spread

THORACIC AND ABDOMINAL VISCERA

- ◯ T1 Tumour confined to a single organ
  - ◯ T2a Tumour invades serosa or visceral peritoneum
  - ◯ T2b Tumour with microscopic extension beyond the serosa
- ◯ T3 Tumour invades another organ or macroscopic extension beyond the serosa
  - ◯ T4a Multifocal tumour involving no more than two sites in one organ
  - ◯ T4b Multifocal tumour involving more than two sites but not more than five sites
  - ◯ T4c Multifocal tumour involving more than five sites

Regional lymph nodes (pN)

- ◯ No nodes submitted or found
- ◯ NX Regional lymph nodes cannot be assessed
- ◯ N0 No regional lymph node metastasis
- ◯ N1 Regional lymph node metastasis

[e] *Reproduced with permission. Source: UICC TNM Classification of Malignant Tumours, 8th Edition, eds by James D. Brierley, Mary K. Gospodarowicz, Christian Wittekind. 2016, Publisher Wiley (incorporating any errata published up until 6th October 2020).*

Version 1.0 Published April 2021 ISBN: 978-1-922324-19-1

Definitions

CORE elements

CORE elements are those which are essential for the clinical management, staging or prognosis of the cancer. These elements will either have evidentiary support at Level III-2 or above (based on prognostic factors in the National Health and Medical Research Council (NHMRC) levels of evidence).[1] In rare circumstances, where level III-2 evidence is not available an element may be made a CORE element where there is unanimous agreement in the expert committee. An appropriate staging system e.g., Pathological TNM staging would normally be included as a CORE element.

The summation of all CORE elements is considered to be the minimum reporting standard for a specific cancer.

NON-CORE elements

NON-CORE elements are those which are unanimously agreed should be included in the dataset but are not supported by level III-2 evidence. These elements may be clinically important and recommended as good practice but are not yet validated or regularly used in patient management.

Key information other than that which is essential for clinical management, staging or prognosis of the cancer such as macroscopic observations and interpretation, which are fundamental to the histological diagnosis and conclusion e.g., macroscopic tumour details, may be included as either CORE or NON-CORE elements by consensus of the Dataset Authoring Committee.

⬆ **Back**

Scope

The dataset has been developed for the pathology reporting of resection specimens for soft tissue sarcomas. Adult rhabdomyosarcoma is also included in this dataset. A separate International Collaboration on Cancer Reporting (ICCR) dataset is available for reporting of biopsy specimens for soft tissue sarcomas.[2]

Some soft tissue tumours which rarely arise primarily in bone and in this case should be reported using the ICCR primary tumour in bone datasets.[3,4]

Lymphoma, uterine sarcoma, paediatric rhabdomyosarcoma and metastases are excluded from this dataset. Gastrointestinal Stromal Tumour (GIST) are also not included in this dataset as GIST displays a number of unique features which warrant its separate consideration; separate ICCR datasets for GIST are available.[5,6]

The authors of this dataset can be accessed here.

⬆ **Back**

Note 1 – Clinical information (Non-core)

It is the responsibility of the clinician requesting the pathological examination of a specimen to provide information that will have an impact on the diagnostic process or affect its interpretation. The use of a standard pathology requisition/request form including a checklist of important clinical information is strongly encouraged to help ensure that important clinical data is provided by the clinician with the specimen.

It is the responsibility of the pathologist to verify that all clinical information necessary for an accurate diagnosis is available to ensure that diagnosis is made within the appropriate clinical/imaging context. This can often be achieved through discussion at a multidisciplinary tumour board meeting.

As an example, the coexistence of systemic disorders such as immunosuppression, which would be relevant in the evaluation of specific lesions such as Epstein-Barr virus (EBV)-related smooth muscle neoplasms and Kaposi sarcoma, should be reported.

⬆ **Back**

Note 2 – Neoadjuvant therapy (Core)

Neoadjuvant therapy may have a profound effect on the morphology of the tumour. In particular, knowledge of such prior therapy may help to interpret changes such as tumour differentiation, necrosis, vasculature changes, cellular atypia and presence of inflammatory cells. For this reason, information about any previous therapy is important for the accurate assessment of soft tissue tumour specimens.

⬆ **Back**

Note 3 – Operative procedure (Core)

It is important that the type and intent of the operative procedure is clearly stated by the surgeon, as this impacts accurate pathologic assessment.

⬆ **Back**

Note 4 – Tumour site (Core)

Primary anatomic site is an important prognostic parameter. The anatomic location often impacts on the risk of aggressive behaviour. As an example, atypical lipomatous tumour/well differentiated liposarcoma arising superficially has a risk of local recurrence around 10%, whereas when occurring in the retroperitoneum the risk approaches 80%.

⬆ **Back**

Note 5 – Tumour depth - tissue plane (Core)

Depth is also important. For example, the risk of distant spread of leiomyosarcoma varies from virtually 0% for purely dermal lesions to approximately 50% for deep seated tumours. For this reason, it is critical to specify anatomic location and depth as accurately as possible.

The 'not known' designation may be necessary if tumour is excised without any surrounding normal tissue or in the absence of any information from the surgeon.

⬆ **Back**

Note 6 – Tumour dimensions (Core and Non-core)

Tumour size is a critical parameter for assessment of the risk of malignant behaviour in selected histotypes such as solitary fibrous tumour.[7] Size is also part of some staging systems if/when used.

⬆ **Back**

Note 7 – Histological tumour type (Core)

Histological diagnosis is based on the 2020 World Health Organization (WHO) Classification of Soft Tissue and Bone Tumours, 5th edition (Table 1).[8] The WHO classification is based on microscopic morphologic findings, variably combined with immunohistochemical and/or molecular findings.[8] If further testing is not available, then the possible diagnostic options should be described. The histopathologic report should include the supporting ancillary testing if performed.

Table 1: World Health Organization classification of soft tissue tumours.[8]

Descriptor	ICD-O codes[a]
Adipocytic tumours	
Intermediate (locally aggressive)	
Atypical lipomatous tumour	8850/1
Malignant	
Liposarcoma, well-differentiated, not otherwise specified (NOS)	8851/3
Lipoma-like liposarcoma	8851/3
Inflammatory liposarcoma	8851/3
Sclerosing liposarcoma	8851/3
Dedifferentiated liposarcoma	8858/3
Myxoid liposarcoma	8852/3
Pleomorphic liposarcoma	8854/3
Epithelioid liposarcoma	
Myxoid pleomorphic liposarcoma	8859/3*
Fibroblastic and myofibroblastic tumours	
Intermediate (rarely metastasizing)	
Dermatofibrosarcoma protuberans NOS	8832/1
Pigmented dermatofibrosarcoma protuberans	8833/1
Dermatofibrosarcoma protuberans, fibrosarcomatous	8832/3

Descriptor	ICD-O codes[a]
Myxoid dermatofibrosarcoma protuberans	
Dermatofibrosarcoma protuberans with myoid differentiation	
Plaque-like dermatofibrosarcoma protuberans	
Solitary fibrous tumour NOS	8815/1
Fat-forming (lipomatous) solitary fibrous tumour	
Giant cell–rich solitary fibrous tumour	
Inflammatory myofibroblastic tumour	8825/1
Epithelioid inflammatory myofibroblastic sarcoma	
Myofibroblastic sarcoma	8825/3
Superficial CD34-positive fibroblastic tumour	8810/1
Myxoinflammatory fibroblastic sarcoma	8811/1
Infantile fibrosarcoma	8814/3
Malignant	
Solitary fibrous tumour, malignant	8815/3
Fibrosarcoma NOS	8810/3
Myxofibrosarcoma	8811/3
Epithelioid myxofibrosarcoma	
Low grade fibromyxoid sarcoma	8840/3
Sclerosing epithelioid fibrosarcoma	8840/3
So-called fibrohistiocytic tumours	
Intermediate (rarely metastasizing)	
Plexiform fibrohistiocytic tumour	8835/1
Giant cell tumour of soft parts	9251/1
Malignant	
Malignant tenosynovial giant cell tumour	9252/3
Vascular tumours	
Intermediate (rarely metastasizing)	
Retiform haemangioendothelioma	9136/1
Papillary intralymphatic angioendothelioma	9135/1
Composite haemangioendothelioma	9136/1
Neuroendocrine composite haemangioendothelioma	
Kaposi sarcoma	9140/3
Classic indolent Kaposi sarcoma	
Endemic African Kaposi sarcoma	
AIDS-associated Kaposi sarcoma	
Iatrogenic Kaposi sarcoma	
Pseudomyogenic (epithelioid sarcoma–like) haemangioendothelioma	9138/1
Malignant	
Epithelioid haemangioendothelioma NOS	9133/3
Epithelioid haemangioendothelioma with *WWTR1-CAMTA1* fusion	
Epithelioid haemangioendothelioma with *YAP1-TFE3* fusion	
Angiosarcoma	9120/3
Pericytic (perivascular) tumours	
Malignant	
Glomus tumour, malignant	8711/3

Descriptor	ICD-O codes[a]
Smooth muscle tumours	
Malignant	
Leiomyosarcoma NOS	8890/3
Skeletal muscle tumours	
Malignant	
Embryonal rhabdomyosarcoma NOS	8910/3
Embryonal rhabdomyosarcoma, pleomorphic	8910/3
Alveolar rhabdomyosarcoma	8920/3
Pleomorphic rhabdomyosarcoma NOS	8901/3
Spindle cell rhabdomyosarcoma	8912/3
Congenital spindle cell rhabdomyosarcoma with *VGLL2/NCOA2/CITED2* rearrangements	
MYOD1-mutant spindle cell/sclerosing rhabdomyosarcoma	
Intraosseous spindle cell rhabdomyosarcoma (with *TFCP2/NCOA2* rearrangements)	
Ectomesenchymoma	8921/3
Chondro-osseous tumours	
Malignant	
Osteosarcoma, extraskeletal	9180/3
Peripheral nerve sheath tumours	
Malignant	
Malignant peripheral nerve sheath tumour NOS	9540/3
Malignant peripheral nerve sheath tumour, epithelioid	9542/3
Malignant melanotic nerve sheath tumour	9540/3
Granular cell tumour, malignant	9580/3
Tumours of uncertain differentiation	
Intermediate (rarely metastasizing)	
Atypical fibroxanthoma	8830/1
Angiomatoid fibrous histiocytoma	8836/1
Ossifying fibromyxoid tumour NOS	8842/0
Mixed tumour NOS	8940/0
Mixed tumour, malignant, NOS	8940/3
Myoepithelioma NOS	8982/0
Malignant	
Phosphaturic mesenchymal tumour, malignant NTRK-rearranged spindle cell neoplasm (emerging)	8990/3
Synovial sarcoma NOS	9040/3
Synovial sarcoma, spindle cell	9041/3
Synovial sarcoma, biphasic	9043/3
Synovial sarcoma, poorly differentiated	
Epithelioid sarcoma	8804/3
Proximal or large cell epithelioid sarcoma	
Classic epithelioid sarcoma	
Alveolar soft part sarcoma	9581/3
Clear cell sarcoma of soft tissue	9044/3

Descriptor	ICD-O codes[a]
Extraskeletal myxoid chondrosarcoma	9231/3
Desmoplastic small round cell tumour	8806/3
Rhabdoid tumour of soft tissue	8963/3
Perivascular epithelioid tumour, malignant	8714/3
Intimal sarcoma	9137/3
Ossifying fibromyxoid tumour, malignant	8842/3
Myoepithelial carcinoma	8982/3
Undifferentiated sarcoma	8805/3
Spindle cell sarcoma, undifferentiated	8801/3
Pleomorphic sarcoma, undifferentiated	8802/3
Round cell sarcoma, undifferentiated	8803/3
Undifferentiated small round cell sarcomas of bone and soft tissue	
Ewing sarcoma	9364/3
Round cell sarcoma with *EWSR1*–non-ETS fusions	9366/3*
CIC-rearranged sarcoma	9367/3*
Sarcoma with *BCOR* genetic alterations	9368/3*

[a] These morphology codes are from the International Classification of Diseases for Oncology, third edition, second revision (ICD-O-3.2).[9] Behaviour is coded /0 for benign tumours; /1 for unspecified, borderline, or uncertain behaviour; /2 for carcinoma in situ and grade III intraepithelial neoplasia; /3 for malignant tumours, primary site; and /6 for malignant tumours, metastatic site.
*Codes marked with an asterisk were approved by the International Agency for Research on Cancer /WHO Committee for ICD-O at its meeting in January 2020. Incorporates all relevant changes from the 5th Edition Corrigenda October 2020.

Version 3.2 of the ICD-O codes is finalised and available at: http://www.iacr.com.fr/index.php?option=com_content&view=article&id=149:icd-o-3-2&catid=80&Itemid=545. However, changes made to the histological entities during the 5th edition update will only be formally incorporated into a subsequent version of ICD-O once the 5th edition is complete. There are, therefore, some issues of concordance between the histological entities listed in the chapters of the WHO Classification of Tumours and the ICD-O Tables.

Back

Note 8 – Histological tumour grade (Core)

Histologic tumour grade offers important prognostic information. While several different grading systems exist, the French[10] grading system is the most widely used (see Table 2). This system is based on the assessment of differentiation, mitotic count, and necrosis.[10] Importantly, the system only applies to specific histotypes (see Table 3). Many other histotypes are not gradable. Reliable tumour grading is not possible after neoadjuvant therapy.

Table 2: Tumour Differentiation Score According to Histologic Type in the Updated Version of the French Federation of Cancer Centers Sarcoma Group System.[10]

Histologic type	Score
Atypical lipomatous tumour/Well-differentiated liposarcoma	1
Well-differentiated leiomyosarcoma	1
Malignant neurofibroma	1
Well-differentiated fibrosarcoma	1
Myxoid liposarcoma	2
Conventional leiomyosarcoma	2
Conventional fibrosarcoma	2
Myxofibrosarcoma	2
High-grade myxoid (round cell) liposarcoma	3
Pleomorphic liposarcoma	3
Dedifferentiated liposarcoma	3
Pleomorphic rhabdomyosarcoma	3
Poorly differentiated/pleomorphic leiomyosarcoma	3
Biphasic/monophasic/poorly differentiated Synovial sarcoma	3
Mesenchymal chondrosarcoma	3
Extraskeletal osteosarcoma	3
Extraskeletal Ewing sarcoma	3
Malignant rhabdoid tumour	3
Undifferentiated pleomorphic sarcoma	3
Undifferentiated sarcoma, not otherwise specified	3

Table 3: Guidelines for grading soft tissue sarcomas.

Tumours which are by definition high grade • Ewing sarcoma • Rhabdomyosarcoma (all types) • Angiosarcoma • Pleomorphic liposarcoma • Soft tissue osteosarcoma • Mesenchymal chondrosarcoma • Desmoplastic small cell tumour • Extra-renal rhabdoid tumour • Intimal sarcoma	**Tumours of varying behaviour for which grading or tumour-specific risk assessment may be prognostically useful** • Myxoid liposarcoma • Leiomyosarcoma • Malignant peripheral nerve sheath tumour • Solitary fibrous tumour • Myxofibrosarcoma • Dedifferentiated liposarcoma[a]
Tumours which are by definition low grade • Well differentiated liposarcoma/atypical lipomatous tumour • Dermatofibrosarcoma protuberans[b] • Infantile fibrosarcoma	**Tumours of varying behaviour for which grading parameters are not yet well defined** • Epithelioid hemangioendothelioma • Extraskeletal myxoid chondrosarcoma
Tumours which are not gradable but which often metastasize within 10-20 years of follow-up • Alveolar soft part sarcoma • Clear cell sarcoma • Epithelioid sarcoma • Synovial sarcoma[a] • 'Low-grade' fibromyxoid sarcoma • Sclerosing epithelioid fibrosarcoma	

[a] Some studies have shown prognostic difference between Grades 2 and 3 using the French grading system.
[b] Fibrosarcomatous Dermatofibrosarcoma Protuberans (DFSP) is usually regarded as intermediate grade.

⬆ **Back**

Note 9 – Mitotic count (Core)

Mitotic count is a key parameter for histologic grading of malignancy as well as a factor used in risk assessment schemes (refer to **Note 8 HISTOLOGICAL TUMOUR GRADE**, Table 3). The mitotic count should be determined in the most mitotic area of the tumour. The mitotic count should be reported per 2 mm^2. Ten high power fields (HPFs) approximates to 2 mm^2 on most modern microscopes, but the number of fields to be counted to encompass 2 mm^2 should ideally be calculated on individual microscopes.

⬆ **Back**

Note 10 – Necrosis (Core)

Necrosis is a key parameter for histologic grading of malignancy. As the French grading system[10] is only applicable to untreated tumours, assessment of necrosis following neoadjuvant treatment should not be performed. True coagulative necrosis (with neutrophil polymorphs and cellular debris) should be distinguished from stromal hyalinisation or infarction.

⬆ **Back**

Note 11 – Lymphovascular invasion (Non-core)

Evaluation of lymphovascular invasion has emerged as a potential prognostic parameter, however it is not yet widely adopted.[11,12]

⬆ **Back**

Note 12 – Response to neoadjuvant therapy (Non-core)

Neoadjuvant systemic and/or local treatment of soft tissue sarcomas is gradually entering into clinical practice.[13] Descriptive assessment of the amount of residual viable tumour and type of histologic response may represent valuable information in terms of estimation of efficacy of treatment. Correlation of microscopic features with macroscopic findings is critical. A scientific publication from the European Organisation for Research and Treatment of Cancer (EORTC) suggests that response should be evaluated microscopically on at least one complete central slide of tumour through its largest dimension.[14]

⬆ **Back**

Note 13 – Margin status (Core and Non-core)

The status of the resection margins directly impacts patient outcome. However, there is no generally accepted way of reporting margins for soft tissue tumours. If margins are involved, a distinction is often made between microscopic involvement (R1) and resections in which it is evident macroscopically that the tumour is incompletely resected (R2). In the case of negative margins (R0), the minimum that should be documented is the distance of tumour to the closest margins. The type of tissue comprising the resection margin should also be recorded since it might be that specific tissue types (e.g., fascia) are more robust marginal tissues than others. In some cases margin status cannot be assessed for example, in liposarcomas in the retroperitoneum, or in the case of debulking, piecemeal excision or tumour rupture, in which assessment of margins is not feasible.

Correlation with the surgical findings is critical to ensure accurate reporting.

⬆ **Back**

Note 14 – Lymph node status (Core)

Regional lymph node metastasis is uncommon in adult soft tissue sarcomas. However, there are a few exceptions, for example epithelioid sarcoma and clear cell sarcoma of soft parts. Lymph nodes are not sampled routinely in soft tissue resections, and it is not necessary to undertake an exhaustive search for nodes. However, when present, regional lymph node metastasis has prognostic importance and should be reported.

⬆ **Back**

Note 15 – Coexistent pathology (Non-core)

Pathologists should report other microscopically identifiable abnormalities that are relevant to the diagnosis. For example, the presence of precursor lesions in malignant peripheral nerve sheath tumours (MPNSTs).

⬆ **Back**

Note 16 – Ancillary studies (Core)

All immunohistochemical staining and molecular tests that contributed to the diagnosis should be documented. This includes molecular testing performed on histological tumour types that are defined by specific genetic aberrations (i.e., *CIC*-rearranged sarcomas).

⬆ **Back**

Note 17 – Histologically confirmed distant metastases (Core)

The presence of distant metastases strongly influences outcome. The pattern of metastatic spread of soft tissue sarcomas often depends on the specific histologic type. For example, metastatic spread to the lungs is very common in leiomyosarcoma whereas myxoid liposarcoma can spread to soft tissues and bone without involving the lungs.

⬆ **Back**

Note 18 – Pathological staging (Non-core)

Pathological staging is frequently not applicable or useful in most sarcoma types and has therefore been included in this dataset as a non-core element. However, staging is required in many existing reporting systems (Union for International Cancer Control (UICC)[15] or American Joint Committee on Cancer (AJCC)[16] 8th edition staging systems), and in many cancer centres around the world it is mandated or used as a quality assurance indicator. Staging may also be required per local/institutional preference.

⬆ **Back**

References

1 Merlin T, Weston A and Tooher R (2009). Extending an evidence hierarchy to include topics other than treatment: revising the Australian 'levels of evidence'. *BMC Med Res Methodol* 9:34.

2 International Collaboration on Cancer Reporting (2021). *Soft Tissue Sarcoma Histopathology Reporting Guide - Biopsy Specimens*. Available from: http://www.iccr-cancer.org/datasets/published-datasets/soft-tissue-bone (Accessed 19th April 2021).

3 International Collaboration on Cancer Reporting (2021). *Primary Tumour in Bone Histopathology Reporting Guide - Biopsy Specimens*. Available from: http://www.iccr-cancer.org/datasets/published-datasets/soft-tissue-bone (Accessed 19th April 2021).

4 International Collaboration on Cancer Reporting (2021). *Primary Tumour in Bone Histopathology Reporting Guide - Resection Specimens*. Available from: http://www.iccr-cancer.org/datasets/published-datasets/soft-tissue-bone (Accessed 19th April 2021).

5 International Collaboration on Cancer Reporting (2021). *Gastrointestinal Stromal Tumour (GIST) Histopathology Reporting Guide - Biopsy Specimens*. Available from: http://www.iccr-cancer.org/datasets/published-datasets/soft-tissue-bone (Accessed 19th April 2021).

6 International Collaboration on Cancer Reporting (2021). *Gastrointestinal Stromal Tumour (GIST) Histopathology Reporting Guide - Resection Specimens*. Available from: http://www.iccr-cancer.org/datasets/published-datasets/soft-tissue-bone (Accessed 19th April 2021).

7 Demicco EG, Griffin AM, Gladdy RA, Dickson BC, Ferguson PC, Swallow CJ, Wunder JS and Wang WL (2019). Comparison of published risk models for prediction of outcome in patients with extrameningeal solitary fibrous tumour. *Histopathology* 75(5):723-737.

8 WHO Classification of Tumours Editorial Board (2020). *Soft Tissue and Bone Tumours. WHO Classification of Tumours, 5th Edition, Volume 3*. IARC Publications, Lyon.

9 Fritz A, Percy C, Jack A, Shanmugaratnam K, Sobin LH, Parkin DM, Whelan SL and World Health Organization (2000). *International classification of diseases for oncology*, World Health Organization, Geneva.

10 Guillou L, Coindre JM, Bonichon F, Nguyen BB, Terrier P, Collin F, Vilain MO, Mandard AM, Le Doussal V, Leroux A, Jacquemier J, Duplay H, Sastre-Garau X and Costa J (1997). Comparative study of the National Cancer Institute and French Federation of Cancer Centers Sarcoma Group grading systems in a population of 410 adult patients with soft tissue sarcoma. *J Clin Oncol* 15(1):350-362.

11 Gustafson P, Akerman M, Alvegård TA, Coindre JM, Fletcher CD, Rydholm A and Willén H (2003). Prognostic information in soft tissue sarcoma using tumour size, vascular invasion and microscopic tumour necrosis-the SIN-system. *Eur J Cancer* 39(11):1568-1576.

12 Engellau J, Bendahl PO, Persson A, Domanski HA, Akerman M, Gustafson P, Alvegård TA, Nilbert M and Rydholm A (2005). Improved prognostication in soft tissue sarcoma: independent information from vascular invasion, necrosis, growth pattern, and immunostaining using whole-tumor sections and tissue microarrays. *Hum Pathol* 36(9):994-1002.

13 Gronchi A, Palmerini E, Quagliuolo V, Martin Broto J, Lopez Pousa A, Grignani G, Brunello A, Blay JY, Tendero O, Diaz Beveridge R, Ferraresi V, Lugowska I, Merlo DF, Fontana V, Marchesi E, Braglia L, Donati DM, Palassini E, Bianchi G, Marrari A, Morosi C, Stacchiotti S, Bagué S, Coindre JM, Dei Tos AP, Picci P, Bruzzi P and Casali PG (2020). Neoadjuvant chemotherapy in high-risk soft tissue sarcomas: final results of a randomized trial from Italian (ISG), Spanish (GEIS), French (FSG), and Polish (PSG) Sarcoma Groups. *J Clin Oncol* 38(19):2178-2186.

14 Wardelmann E, Haas RL, Bovée JV, Terrier P, Lazar A, Messiou C, LePechoux C, Hartmann W, Collin F, Fisher C, Mechtersheimer G, DeiTos AP, Stacchiotti S, Jones RL, Gronchi A and Bonvalot S (2016). Evaluation of response after neoadjuvant treatment in soft tissue sarcomas; the European Organization for Research and Treatment of Cancer-Soft Tissue and Bone Sarcoma Group (EORTC-STBSG) recommendations for pathological examination and reporting. *Eur J Cancer* 53:84-95.

15 Brierley JD, Gospodarowicz MK and Wittekind C (eds) (2016). *Union for International Cancer Control. TNM Classification of Malignant Tumours, 8th Edition*, Wiley, USA.

16 Amin MB, Edge SB, Greene FL, Byrd DR, Brookland RK, Washington MK, Gershenwald JE, Compton CC, Hess KR, Sullivan DC, Jessup JM, Brierley JD, Gaspar LE, Schilsky RL, Balch CM, Winchester DP, Asare EA, Madera M, Gress DM and Meyer LR (eds) (2017). *AJCC Cancer Staging Manual. 8th Edition*, Springer, New York.