Histological tumour type (Core)

Histologic diagnosis is based on the 2020 World Health Organization (WHO) Classification of Soft Tissue and Bone Tumours, 5th edition (Table 1).[1] The WHO classification is based on microscopic morphologic findings, variably combined with immunohistochemical and/or molecular findings.[1] 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.

Soft tissue tumours are most often first sampled by biopsy. In some cases, the biopsy is suboptimally centred on the area(s) of interest leaving the pathologist with tissue that can be under-representative or misrepresentative of the lesion based on the imaging studies. Molecular testing may be required to achieve a full/correct diagnosis, but the small tissue size, tissue processing issues, or suboptimal targeting of biopsy materials may make this further testing impossible. The pathologist should specify any, and all, limitations of the tissue in achieving optimal diagnosis.

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

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
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

Descriptor	ICD-O codes[a]
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
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

Descriptor	ICD-O codes[a]
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
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

Descriptor	ICD-O codes[a]
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).[2] 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.

References

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

2 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.