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).[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.

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
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

Descriptor	ICD-O codes[a]
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
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

Descriptor	ICD-O codes[a]
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
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.