Histological tumour type (Core)

Histologic diagnosis is based on the World Health Organization (WHO) Classification of Tumours, Soft Tissue and Bone Tumours, 5th edition, 2020 (Table 1). Many tumours of the bone are surgically assessed by biopsy. In some cases, the biopsy is suboptimally centred on the area(s) of interest or affected by the surgical process, leaving the pathologist with tissue that can be under-representative or misrepresentative of the lesion based on the imaging studies. In a few instances, more sophisticated testing (e.g., molecular) 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 testing impossible. The pathologist should specify any, and all, limitations of the tissue in achieving optimal diagnosis. In addition, comments can be made in case the diagnosis on biopsy is not certain for reasons other than limitations of the material, or when there is still a differential diagnosis.

<u>Table 1: World Health Organization classification of intermediate and malignant bone tumours</u> and undifferentiated small round cell sarcomas.¹

| Descriptor | ICD-O codes ^a |
|---------------------------------------|--------------------------|
| Chondrogenic tumours | |
| Intermediate (locally aggressive) | |
| Atypical cartilaginous tumour | 9222/1 |
| Malignant | |
| Chondrosarcoma, grade 1 | 9222/3* |
| Chondrosarcoma, grade 2 | 9220/3 |
| Chondrosarcoma, grade 3 | 9220/3 |
| Periosteal chondrosarcoma | 9221/3 |
| Clear cell chondrosarcoma | 9242/3 |
| Mesenchymal chondrosarcoma | 9240/3 |
| Dedifferentiated chondrosarcoma | 9243/3 |
| Osteogenic tumours | |
| Malignant | |
| Low-grade central osteosarcoma | 9187/3 |
| Osteosarcoma | 9180/3 |
| Conventional osteosarcoma | |
| Telangiectatic osteosarcoma | |
| Small cell osteosarcoma | |
| Parosteal osteosarcoma | 9192/3 |
| Periosteal osteosarcoma | 9193/3 |
| High-grade surface osteosarcoma | 9194/3 |
| Secondary osteosarcoma | 9184/3 |
| Fibrogenic tumours | |
| Malignant | |
| Fibrosarcoma NOS | 8810/3 |
| Vascular tumours of bone | |
| Malignant | |
| Epithelioid haemangioendothelioma NOS | 9133/3 |
| Angiosarcoma | 9120/3 |

| Descriptor | ICD-O codes ^a |
|---|--------------------------|
| Osteoclastic giant cell–rich tumours | |
| Intermediate (locally aggressive, rarely metastasizing) | |
| Giant cell tumour of bone | 9250/1 |
| Malignant | |
| Giant cell tumour of bone, malignant | 9250/3 |
| Notochordal tumours | |
| Malignant | |
| Conventional chordoma | 9370/3 |
| Chondroid chordoma | |
| Poorly differentiated chordoma | 9370/3 |
| Dedifferentiated chordoma | 9372/3 |
| Other mesenchymal tumours of bone | |
| Malignant | |
| Adamantinoma of long bones | 9261/3 |
| Dedifferentiated adamantinoma | |
| Leiomyosarcoma NOS | 8890/3 |
| Pleomorphic sarcoma, undifferentiated | 8802/3 |
| Haematopoietic neoplasms of bone | |
| Plasmacytoma of bone | 9731/3 |
| Malignant lymphoma, non-Hodgkin, NOS | 9591/3 |
| Hodgkin disease NOS | 9650/3 |
| Diffuse large B-cell lymphoma NOS | 9680/3 |
| Follicular lymphoma NOS | 9690/3 |
| Marginal zone B-cell lymphoma NOS | 9699/3 |
| T-cell lymphoma NOS | 9702/3 |
| Anaplastic large cell lymphoma NOS | 9714/3 |
| Malignant lymphoma, lymphoblastic, NOS | 9727/3 |
| Burkitt lymphoma NOS | 9687/3 |
| Langerhans cell histiocytosis NOS | 9751/1 |
| Langerhans cell histiocytosis, disseminated | 9751/3 |
| Erdheim-Chester disease | 9749/3 |
| Rosai–Dorfman disease | |
| Undifferentiated small round cell | |
| 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.
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