

Chapter 4

Classification of Primary Bone Tumors

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Primary bone tumors are named and classified based upon their histologic differentiation, as reported in the following Table 4.1. The term “tumor” refers in general to a growing mass and is also used by clinicians for lesions of nonneoplastic nature, such as histiocytic fibroma and osteochondroma, because their basic tissue expresses a recognizable differentiation, such as fibrohistiocytic and cartilaginous. Other growing lesions are usually defined as “reactive” and do not show a recognizable pattern of histologic differentiation. Lymphomas and myeloma are not considered primary bone tumors, deriving from hematopoietic system.

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Table 4.1 Classification of primary bone tumors

Histogenesis	Benign	Intermediate (locally aggressive and/or rarely metastasizing)	Malignant [(*) low-grade lesions if not dedifferentiated]
Fibrogenic and fibrohistiocytic	Histiocytic fibroma	Desmoplastic fibroma	Fibrosarcoma
Chondrogenic	Benign fibrous histiocytoma		
	Osteochondroma	Chondromyxoid fibroma	Chondrosarcoma grade 2
	Hemimelic epiphyseal dysplasia		Chondrosarcoma grade 3
	Enchondroma	Chondrosarcoma grade 1	Dedifferentiated chondrosarcoma
	Periosteal chondroma	Chondroblastoma	Mesenchymal chondrosarcoma
	Osteochondromyxoma		Peripheral chondrosarcoma (*)
	Subungual exostosis		Periosteal chondrosarcoma (*)
	Bizarre parosteal osteochondromatous proliferation	Fibrocartilaginous mesenchymoma	Clear cell chondrosarcoma (*)
	Synovial chondromatosis		
	Osteoma	Osteoblastoma	Low-grade central osteosarcoma (*)
Osteoclastic			Conventional osteosarcoma
			Telangiectatic osteosarcoma
			Small cell osteosarcoma
			Secondary osteosarcoma
			Parosteal osteosarcoma (*)
			Periosteal osteosarcoma (*)
			High-grade surface osteosarcoma
			Malignancy in giant cell tumor
	Giant cell rich	Giant cell reparative granuloma	Giant cell tumor of bone
	Vascular	Hemangioma	Epithelioid hemangioma
Lymphangioma			Epithelioid hemangioidothelioma (*) Angiosarcoma

Nervous	Schwannoma		
	Neurofibroma		
Lipogenic	Lipoma		MPNST
Myogenic	Leiomyoma		Liposarcoma
Notochordal	Benign notochordal cell tumor (BNCT)		Leiomyosarcoma
Tumors of undefined neoplastic nature/ miscellaneous tumors	Simple bone cyst	Aneurysmal bone cyst	Chordoma (*)
	Fibrous dysplasia	Langerhans cell histiocytosis	Adamantinoma (*)
	Osteofibrous dysplasia	Erdheim-Chester disease	Ewing sarcoma
	Chondromesenchymal hamartoma		Undifferentiated pleomorphic sarcoma
	Rosai-Dorfman disease		