WHO Classification of Tumours, 5th edition: Soft Tissue and Bone Tumours
Corrections made in the second print run

Summary of corrections:

Front matter **p. iv**
The WHO Classification of Tumours Editorial Board > Expert members: Soft tissue and bone tumours

Drs Gronchi and Messiou have been added to the list of WHO Classification of Tumours Editorial Board expert members:

Gronchi, Alessandro
Fondazione IRCCS Istituto Nazionale dei Tumori
Milan

Messiou, Christina
Royal Marsden Hospital
London

Chapter 1: Soft tissue tumours **p. 2–3**
WHO classification of soft tissue tumours

Several ICD-O labels have been corrected, as shown.

Original text	Corrected text
Fibroblastic and myofibroblastic tumours … 9160/0 Angiofibroma NOS … **So-called fibrohistiocytic tumours** … 9251/1 Giant cell tumour of soft parts NOS … **Peripheral nerve sheath tumours** … 9540/3 Melanotic malignant peripheral nerve sheath tumour	**Fibroblastic and myofibroblastic tumours** … 9160/0 Angiofibroma … **So-called fibrohistiocytic tumours** … 9251/1 Giant cell tumour of soft parts … **Peripheral nerve sheath tumours** … 9540/3 Malignant melanotic nerve sheath tumour

And the following footnote has been added at the end of the WHO classification table:

“Subtype labels are indented.”

Chapter 1: Soft tissue tumours **p. 54**
Myositis ossificans and fibro-osseous pseudotumour of digits

Text that had accidentally been deleted from this page during the layout process has been added back to this section.

A corrected, printable version of page 54 is included at the end of this document.

Original text	Corrected text
is extraskeletal osteosarcoma, which lacks zonation and shows malignant cytology. **Cytology** Cytology features …	**Etiology** Unknown **Pathogenesis** Most cases of myositis ossificans and FP harbour the fusion *COL1A1-USP6* {2980,3008,1036,260,1504}. *USP6* fusion genes are also encountered in aneurysmal bone cyst and nodular fasciitis, tumours with overlapping morphology.

continued on p.2

Original text (cont.)	Corrected text (cont.)
	Macroscopic appearance Myositis ossificans is well delineated and ovoid, has a glistening soft-tan haemorrhagic centre and a firm greyish-white gritty periphery, and is 2–12 cm (average: 5 cm). FP is tan/grey/pink, smaller, and less well demarcated. **Histopathology** Myositis ossificans is characterized by zonal architecture. Initially, it is densely cellular, composed of spindle cells oriented randomly or in short intersecting fascicles. The spindle cells have eosinophilic cytoplasm and plump vesicular nuclei with nucleoli. Normal mitoses are often numerous. The stroma is vascular and myxoid and contains fibrin, extravasated erythrocytes, scattered lymphocytes, and osteoclast-like giant cells. Blood-filled cysts may be present. Peripherally, the spindle cells merge with osteoblasts that rim and populate ill-defined trabeculae and sheets of unmineralized woven bone, which is surrounded by well-formed trabecular and cortical-type bone that remodels into lamellar bone. Occasionally, the matrix contains cellular hyaline cartilage or the zonal architecture is not well developed {12,802,2116,2335,2658,2987}. The bone is randomly distributed in FP. An important differential diagnosis is extraskeletal osteosarcoma, which lacks zonation and shows malignant cytology. **Cytology** Cytology features …

Chapter 1: Soft tissue tumours **p. 82**
Angiofibroma of soft tissue > ICD-O coding

The ICD-O label has been corrected as shown.

Original text	Corrected text
ICD-O coding 9160/0 Angiofibroma NOS	**ICD-O coding** 9160/0 Angiofibroma

Chapter 1: Soft tissue tumours **p. 141**
Giant cell tumour of soft tissue > ICD-O coding

The ICD-O label has been corrected as shown.

Original text	Corrected text
ICD-O coding 9251/1 Giant cell tumour of soft parts NOS	**ICD-O coding** 9251/1 Giant cell tumour of soft parts

Chapter 1: Soft tissue tumours **p. 147**
Arteriovenous malformation/haemangioma > Definition

Some of the content has been removed from the *Definition* subsection as shown (and moved to later in the section as detailed below).

Original text	Corrected text
Definition Arteriovenous malformation/haemangioma (AVM/H) is a fast-flow, mostly congenital vascular anomaly characterized by the presence of arteriovenous shunts. There are two distinctive forms: deep-seated and cutaneous (cirsoid aneurysm or acral arteriovenous tumour). Most cases of extracranial AVM/H harbour *MAP2K1* mutations, resulting in upregulated MAP2K1 (MEK1) activity. When these lesions involve multiple tissue planes, they are termed angiomatosis.	**Definition** Arteriovenous malformation/haemangioma (AVM/H) is a fast-flow, mostly congenital vascular anomaly characterized by the presence of arteriovenous shunts.

Chapter 1: Soft tissue tumours p. 147
Arteriovenous malformation/haemangioma > Subtype(s)

The text has been corrected as shown.

Original text	Corrected text
Subtype(s) None	**Subtype(s)** Deep-seated AVM/H; cutaneous AVM/H (also called cirsoid aneurysm or acral arteriovenous tumour); angiomatosis (involving multiple tissue planes)

Chapter 1: Soft tissue tumours p. 147
Arteriovenous malformation/haemangioma > Etiology

The text has been corrected as shown.

Original text	Corrected text
Etiology Unknown	**Etiology** Most are solitary and sporadic. Inherited lesions occurring as part of the rare capillary malformation–AVM syndrome are associated with germline *RASA1* mutations, which are probably causative {3064A}.

Chapter 1: Soft tissue tumours p. 147
Arteriovenous malformation/haemangioma > Pathogenesis

The text has been corrected as shown.

Original text	Corrected text
Pathogenesis Unknown	**Pathogenesis** Most cases of extracranial AVM/H harbour *MAP2K1* mutations, resulting in upregulated MAP2K1 (MEK1) activity.

Chapter 1: Soft tissue tumours p. 169
Pseudomyogenic haemangioendothelioma > Related terminology

The text has been corrected as shown.

Original text	Corrected text
Related terminology *Acceptable*: epithelioid sarcoma–like haemangioendothelioma.	**Related terminology** *Not recommended*: epithelioid sarcoma–like haemangioendothelioma.

Chapter 1: Soft tissue tumours p. 201
Embryonal rhabdomyosarcoma > Subtype(s)

The text has been corrected as shown.

Original text	Corrected text
Subtype(s) Embryonal rhabdomyosarcoma, pleomorphic	**Subtype(s)** Embryonal rhabdomyosarcoma, anaplastic

Chapter 1: Soft tissue tumours — p. 226
Schwannoma > Related terminology

The text has been corrected as shown.

Original text	Corrected text
Related terminology *Acceptable*: neurilemmoma.	**Related terminology** *Not recommended*: neurilemmoma.

Chapter 1: Soft tissue tumours — p. 258
Malignant melanotic nerve sheath tumour > ICD-O coding

The ICD-O label has been corrected as shown.

Original text	Corrected text
ICD-O coding 9540/3 Melanotic malignant peripheral nerve sheath tumour	**ICD-O coding** 9540/3 Malignant melanotic nerve sheath tumour

Chapter 3: Bone tumours — p. 338
WHO classification of bone tumours

Several ICD-O labels have been corrected, as shown.

Original text	Corrected text
Chondrogenic tumours … 9220/1 Chondromatosis NOS … **Osteogenic tumours** *Benign* 9180/0 Osteoma NOS 9191/0 Osteoid osteoma NOS … **Osteoclastic giant cell–rich tumours** … 9250/1 Giant cell tumour of bone NOS … **Notochordal tumours** … 9370/3 Chordoma NOS … **Other mesenchymal tumours of bone** … 8990/1 Mesenchymoma NOS	**Chondrogenic tumours** … 9220/1 Synovial chondromatosis … **Osteogenic tumours** *Benign* 9180/0 Osteoma 9191/0 Osteoid osteoma … **Osteoclastic giant cell–rich tumours** … 9250/1 Giant cell tumour of bone … **Notochordal tumours** … 9370/3 Conventional chordoma … **Other mesenchymal tumours of bone** … 8990/1 Fibrocartilaginous mesenchymoma

And the following footnote has been added at the end of the WHO classification table:

“Subtype labels are indented.”

Chapter 3: Bone tumours — p. 368
Synovial chondromatosis > ICD-O coding

The ICD-O label has been corrected as shown.

Original text	Corrected text
ICD-O coding 9220/1 Chondromatosis NOS	**ICD-O coding** 9220/1 Synovial chondromatosis

Chapter 3: Bone tumours p. 391
Osteoma > ICD-O coding

The ICD-O label has been corrected as shown.

Original text	Corrected text
ICD-O coding 9180/0 Osteoma NOS	**ICD-O coding** 9180/0 Osteoma

Chapter 3: Bone tumours p. 394
Osteoid osteoma > ICD-O coding

The ICD-O label has been corrected as shown.

Original text	Corrected text
ICD-O coding 9191/0 Osteoid osteoma NOS	**ICD-O coding** 9191/0 Osteoid osteoma

Chapter 3: Bone tumours p. 440
Giant cell tumour of bone > ICD-O coding

The ICD-O label has been corrected as shown.

Original text	Corrected text
ICD-O coding 9250/1 Giant cell tumour of bone NOS	**ICD-O coding** 9250/1 Giant cell tumour of bone

Chapter 3: Bone tumours p. 451
Conventional chordoma > ICD-O coding

The ICD-O label has been corrected as shown.

Original text	Corrected text
ICD-O coding 9370/3 Chordoma NOS	**ICD-O coding** 9370/3 Conventional chordoma

Chapter 3: Bone tumours p. 456
Poorly differentiated chordoma > ICD-O coding

The ICD-O label has been corrected as shown.

Original text	Corrected text
ICD-O coding 9370/3 Chordoma NOS	**ICD-O coding** 9370/3 Poorly differentiated chordoma

Chapter 3: Bone tumours — p. 470

Fibrocartilaginous mesenchymoma > ICD-O coding

The ICD-O label has been corrected as shown.

Original text	Corrected text
ICD-O coding 8990/1 Mesenchymoma NOS	**ICD-O coding** 8990/1 Fibrocartilaginous mesenchymoma

References — p. 591

A new reference has been added to the reference list:

3064A. Thiex R, Mulliken JB, Revencu N, et al. A novel association between RASA1 mutations and spinal arteriovenous anomalies. AJNR Am J Neuroradiol. 2010 Apr;31(4):775–9. PMID:20007727

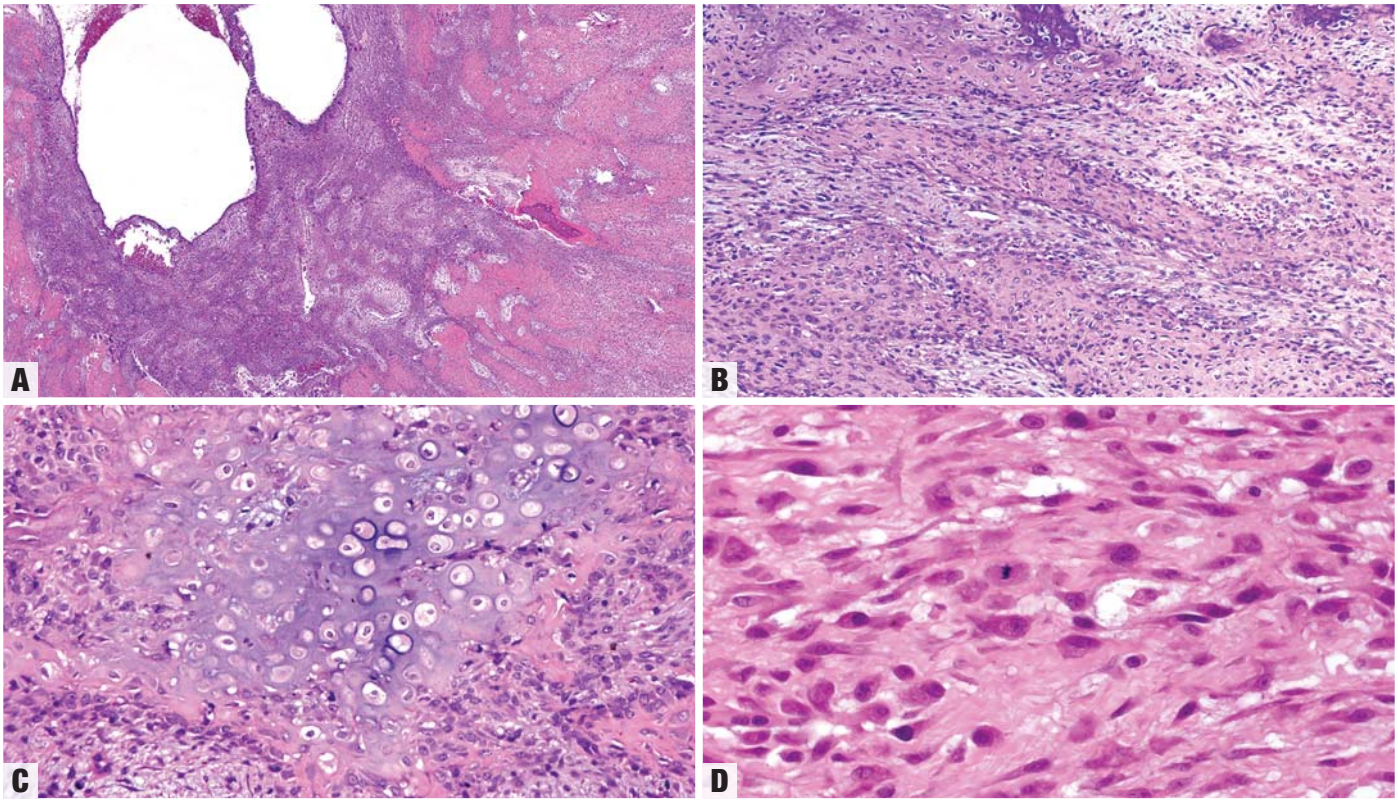

Fig. 1.58 Myositis ossificans. **A** Zonation pattern with focally cystic hypercellular centre surrounded by progressively maturing woven bone. **B** Poorly formed woven bone associated with osteoblasts merges with matrix that is well formed and trabecular in architecture. **C** Hypercellular hyaline cartilage undergoing enchondral ossification. **D** Fascicles of plump spindle cells with elongate nuclei that are mitotically active. The stroma is myxocollagenous with scattered extravasated red blood cells. Histological resemblance to nodular fasciitis is evident.

Etiology
Unknown

Pathogenesis
Most cases of myositis ossificans and FP harbour the fusion *COL1A1-USP6* {2980,3008,1036,260,1504}. *USP6* fusion genes are also encountered in aneurysmal bone cyst and nodular fasciitis, tumours with overlapping morphology.

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Histopathology
Myositis ossificans is characterized by zonal architecture. Initially, it is densely cellular, composed of spindle cells oriented randomly or in short intersecting fascicles. The spindle cells have eosinophilic cytoplasm and plump vesicular nuclei with nucleoli. Normal mitoses are often numerous. The stroma is vascular and myxoid and contains fibrin, extravasated erythrocytes, scattered lymphocytes, and osteoclast-like giant cells. Blood-filled cysts may be present. Peripherally, the spindle cells merge with osteoblasts that rim and populate ill-defined trabeculae and sheets of unmineralized woven bone, which is surrounded by well-formed trabecular and cortical-type bone that remodels into lamellar bone. Occasionally, the matrix contains cellular hyaline cartilage or the zonal architecture is not well developed {12,802,2116,2335,2658,2987}. The bone is randomly distributed in FP. An important differential diagnosis is extraskeletal osteosarcoma, which lacks zonation and shows malignant cytology.

Cytology
Cytology features a dual cell population of spindle cells and large ganglion-like cells set in a myxoid stroma {1649}.

Diagnostic molecular pathology
Molecular studies for *USP6* rearrangement may be useful in the appropriate clinicopathological context.

Essential and desirable diagnostic criteria
Essential: hypercellular fascicles of uniform spindle cells; admixed woven bone with zonation, being most mature at the periphery.

Staging
Not clinically relevant

Prognosis and prediction
Treatment of myositis ossificans and FP is usually simple excision. Prognosis is excellent; recurrence is uncommon.