Corrigenda

WHO Classification of Tumours, 5th edition: Soft Tissue and Bone Tumours

Corrigenda updated: January 2022 (after 2nd print run)

Summary of corrections:

The WHO Classification of Tumours Editorial Board (p. iv)

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

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WHO classification of soft tissue tumours: ICD-O coding (p. 2–3)

The following footnote has been added at the end of the WHO classification (ICD-O coding) table:

Subtype labels are indented.

And several ICD-O labels have been corrected as shown below.

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

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Myositis ossificans and fibro-osseous pseudotumour of digits (p. 54)

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

A corrected, printable version of page 54 is included at the end of the PDF version of this corrigenda document (https://publications.iarc.fr/588).

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. **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 …

Updated online: n/a – This error was present in the print version only
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Angiofibroma of soft tissue (p. 82)

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

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Giant cell tumour of soft tissue (p. 141)

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

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Arteriovenous malformation/haemangioma (p. 147)

Some of the content has been moved from the *Definition* subsection to later in the section as shown 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.
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)
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).

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

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Pseudomyogenic haemangioendothelioma (p. 169)

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.

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Embryonal rhabdomyosarcoma (p. 201)

The text has been corrected as shown.

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

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Schwannoma (p. 226)

The text has been corrected as shown.

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

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Malignant peripheral nerve sheath tumour (p. 256)

In Table 1.04 (Table #5193 online), in the row for ANNUBP (atypical neurofibromatous neoplasm of uncertain biological potential), the proposed definition has been corrected as shown, in order to provide a lower limit as well as an upper limit.

Original text	Corrected text
ANNUBP: Proposed definition Schwann cell neoplasm with ≥ 2 of the following 4 features: cytological atypia, loss of neurofibroma architecture, hypercellularity, and < 1.5 mitoses/mm^2 (< 3 mitotic figures per 10 HPFs[a])	**ANNUBP: Proposed definition** Schwann cell neoplasm with ≥ 2 of the following 4 features: cytological atypia, loss of neurofibroma architecture, hypercellularity, and a mitotic count of > 0.2 mitoses/mm^2 (> 1 mitotic figure per 50 HPFs[a]) and < 1.5 mitoses/mm^2 (< 3 mitotic figures per 10 HPFs[a])

Updated online: Update pending
Updated in print: No (pending next print run)

Malignant melanotic nerve sheath tumour (p. 258)

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

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WHO classification of bone tumours: ICD-O coding (p. 338)

The following footnote has been added at the end of the WHO classification (ICD-O coding) table:

Subtype labels are indented.

And several ICD-O labels have been corrected as shown below.

Original text	Corrected text
Chondrogenic tumours … 9220/1 Chondromatosis NOS	**Chondrogenic tumours** … 9220/1 Synovial chondromatosis
Osteogenic tumours *Benign* 9180/0 Osteoma NOS 9191/0 Osteoid osteoma NOS	**Osteogenic tumours** *Benign* 9180/0 Osteoma 9191/0 Osteoid osteoma
Osteoclastic giant cell–rich tumours … 9250/1 Giant cell tumour of bone NOS	**Osteoclastic giant cell–rich tumours** … 9250/1 Giant cell tumour of bone

Original text	Corrected text
Notochordal tumours … 9370/3 Chordoma NOS	**Notochordal tumours** … 9370/3 Conventional chordoma
Other mesenchymal tumours of bone … 8990/1 Mesenchymoma NOS	**Other mesenchymal tumours of bone** … 8990/1 Fibrocartilaginous mesenchymoma

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WHO classification of bone tumours: ICD-O coding (p. 338)

Under the heading "Haematopoietic neoplasms of bone", the following code has been added for Rosai–Dorfman disease.

Original text	Corrected text
Haematopoietic neoplasms of bone … 9751/1 Langerhans cell histiocytosis NOS 9751/3 Langerhans cell histiocytosis, disseminated 9749/3 Erdheim–Chester disease Rosai–Dorfman disease	**Haematopoietic neoplasms of bone** … 9751/1 Langerhans cell histiocytosis NOS 9751/3 Langerhans cell histiocytosis, disseminated 9749/3 Erdheim–Chester disease 9749/3 Rosai–Dorfman disease

Updated online: Update pending
Updated in print: No (pending next print run)

Synovial chondromatosis (p. 368)

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

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Osteoma (p. 391)

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

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Osteoid osteoma (p. 394)

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

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Giant cell tumour of bone (p. 440)

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

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Conventional chordoma (p. 451)

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

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Poorly differentiated chordoma (p. 456)

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

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Fibrocartilaginous mesenchymoma (p. 470)

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

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Rosai–Dorfman disease (p. 498)

The *ICD-O coding* subsection has been updated as shown.

Original text	Corrected text
ICD-O coding None	**ICD-O coding** 9749/3 Rosai–Dorfman disease

Updated online: Update pending
Updated in print: No (pending next print run)

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

Updated online: n/a – A standalone reference list is currently not included in WHO Classification of Tumours Online
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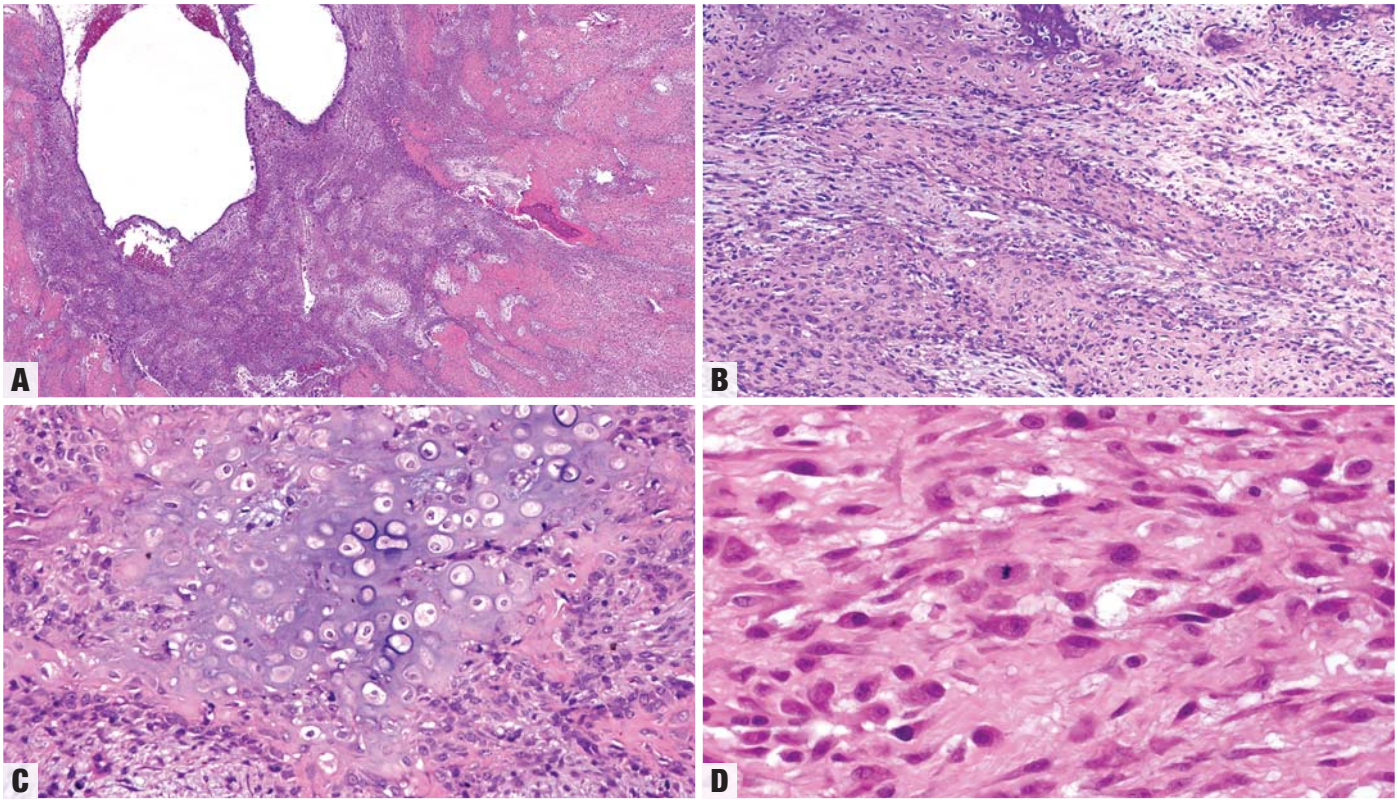

Fig. 1.58 Myositis ossificans. **A** Zonation pattern with focally cystic hypercellular centre surrounded by progressively maturing woven bone. **B** Poorly formed woven bone associated with osteoblasts merges with matrix that is well formed and trabecular in architecture. **C** Hypercellular hyaline 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.

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 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.