WHO Classification Changes for Soft-Tissue and Bone Tumors

A recent review clarifies the major classification changes in the current 2013 WHO classification, and factors in new genetic data that has emerged since the publication of the current volume.

In 2013, the World Health Organization (WHO) published an updated classification of tumors of the soft tissue and bone. This new publication was published 11 years after the last publication dealing with this topic and provides updates predominantly based on the identification of new genetic findings in different tumors types.

In a review published recently in Cancer, Leona A. Doyle, MD, discussed the major classification changes in the current 2013 WHO classification, along with new genetic data that has emerged even since the publication of the current volume. “This refinement has led to more reproducible classifications of soft-tissue and bone tumors, thereby allowing for more effective treatment stratification,” Doyle told Cancer Network. “It has also allowed wastebasket diagnostic categories and obsolete tumor types such as hemangiopericytoma and so-called malignant fibrous histiocytoma to be removed from the 2013 WHO classification, with the recognition that these groups included many tumors which can now be accurately classified as specific sarcoma types.”

**Soft-Tissue Tumors**

In her review, Doyle first addresses soft-tissue tumors and notes that the most notable change to the WHO classification is the elimination of the term “round cell liposarcoma.” This term was used to describe the morphologic appearance of a subset of high-grade myxoid liposarcoma, which most often demonstrates spindle cell morphology. Research has demonstrated that spindle cell and round cell morphology both have the same genetic findings and the same prognostic information, eliminating the need to distinguish between the two.

In addition, mixed-type liposarcoma was removed as a classification with consensus opinion stating that tumors showing this pattern “most likely represent dedifferentiated liposarcoma.” Malignant fibrous histiocytoma was also removed as a classification because many of these tumors can now be classified as specific sarcoma types.

Among the new additions to the classification are dermatofibrosarcoma protuberans and giant cell fibroblastoma, which were previously described in the WHO volume on skin tumors. Also, for the first time, gastrointestinal stromal tumor (GIST) and nerve sheath tumors are now included in the soft-tissue classification.

“The most notable change in the classification of GIST is the recognition of the category of ‘succinate dehydrogenase (SDH)-deficient GIST,’ ” Doyle wrote. “Tumors in this group are wild-type for KIT and PDGFRA mutations, and demonstrate loss of expression of the SDH complex, subunit B (SDHB) protein immunohistochemically, which reflects dysfunction of the SDH enzyme complex of the Krebs cycle.”

The addition of the category of undifferentiated/unclassified sarcoma is also new to the 2013 WHO Classification, and recognizes those tumors that cannot be classified into any of the other categories, due to lack of a demonstrable line of differentiation or lack of distinguishing histologic, immunohistochemical, or genetic features.

“Examples of recently described distinct entities and new genetic findings described for the first time in this volume include pseudomyogenic hemangioendothelioma and phosphaturic mesenchymal tumor, the presence of MYH9-USP6 fusion gene in nodular fasciitis, and the WWTR1-CAMTA1 fusion gene in epithelioid hemangioendothelioma,” Doyle said.

**Bone Tumors**

Doyle also detailed many of the changes and updates made to the classification of bone tumors. Chondrosarcoma are now separated into two ICD codes, reflecting the prognostic difference based
on disease grade. Significant genetic findings for chondrosarcoma are also discussed in the updated classification, with the recognition of the role of mutations in the isocitrate dehydrogenase 1 and 2 genes, which code for the metabolic enzymes IDH1 and IDH2. Fibrosarcoma of bone is now classified as a diagnosis of exclusion. Tumors classified as fibrosarcoma of bone are intermediate- to high-grade spindle cell malignant neoplasms and lack any line of differentiation other than fibroblastic.

According to the review article this new classification addressed several issues. “First, although historically the classification of fibrosarcoma of bone was used relatively commonly, it is now recognized that a fascicular or ‘herringbone’ pattern of growth may be observed in many different tumors types that can be classified on other specific diagnostic categories,” Doyle wrote. “Second, this pattern of growth may also be seen in otherwise unclassified high-grade pleomorphic sarcomas, and if significant pleomorphism is present the tumor is best classified as the latter.”

The 2013 WHO Classification of Tumors of Soft Tissue and Bone include many other updates, deletions, and reclassification of these tumor types that reflect the recent and ongoing advances made in the field.

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