



CORR Insights

CORR Insights®: Do Orthopaedic Oncologists Agree on the Diagnosis and Treatment of Cartilage Tumors of the Appendicular Skeleton?

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Where Are We Now?

Hyaline cartilage tumors are so common that some orthopaedic oncologists view them as the “low back pain” of orthopaedic oncology. Though

observed frequently, distinguishing between benign enchondroma and a low-grade chondrosarcoma is a challenge for orthopaedic oncologists. Which ones need treatment? Which ones can be observed?

These questions are difficult to answer because orthopaedic oncologists lack a gold standard for diagnosing this common tumor. Radiology and histology are not foolproof. Numerous papers have shown difficulty in distinguishing these tumors by

radiologic means alone [1, 5], and unlike other neoplasms, biopsy of hyaline cartilage tumors rarely provide a conclusive diagnosis, making it difficult for pathologists and radiologists to recommend a definitive treatment option. This was eloquently demonstrated by the Skeletal Lesions Interobserver Correlation among Expert Diagnosticians (SLICED) study group [4], which showed that even experienced pathologists and radiologists had low reliability in distinguishing benign versus malignant hyaline cartilage tumors, let alone the grade of malignant ones.

In the current study, Zamora and colleagues show that expert orthopaedic oncologists using clinical information and radiology studies barely have fair interobserver agreement in grading intramedullary extremity hyaline cartilage tumors.

With this as background, the argument could be made that radiologists, pathologists, and orthopaedic oncologists do not have a great handle on what to do with these tumors.

This CORR Insights is a commentary on the article “Do Orthopaedic Oncologists Agree on the Diagnosis and Treatment of Cartilage Tumors of the Appendicular Skeleton?” by Zamora and colleagues available at: DOI: 10.1007/s11999-017-5276-y.

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Where Do We Need To Go?

No consensus has been developed because the histologic interpretation of these tumors is flawed. Few would argue with Zemora and colleagues' definition of a benign enchondroma based on radiographic stability over a 3-year period. However, for the majority of surgically treated tumors, the final diagnosis was based on the pathology results. Similarly, an effort by a group from Spain to: (1) Distinguish between enchondroma and low-grade chondrosarcoma using an aggressiveness scale, and (2) develop a management algorithm also relied upon a final pathology diagnosis in 67% of cases [3]. Since the SLICED study showed that histology is unreliable in defining these tumors, any attempt to construct management schemes based on histology is building on shifting sands.

In a recent natural history study of these tumors, Deckers and colleagues [2] found that only 6% of their patients with hyaline cartilage tumors required operation for "medical reasons" (tumor growth or unexplained pain). Why are so many of these lesions in need of biopsy? We need a more precise means of histologically distinguishing enchondroma from low-grade chondrosarcoma, as well as an improved grading system for chondrosarcomas in general. In order

to develop better approaches, a paradigm shift in how we think about these tumors is warranted. Perhaps we need to steer clear of predicting histology and focus on predicting behavior based on prebiopsy decision-making analysis.

How Do We Get There?

Only a small percentage of long bone hyaline cartilage tumors require surgery [2]. If we could agree to use behavior as the gold standard, the key in developing a standardized approach to these tumors would then be to develop a nomogram based on clinical and radiologic criteria. In order to do that, we need to develop prospective natural history studies examining possible behavioral predictive variables for these tumors with long-term non-operative followup. Ideally, this will require hundreds to even thousands of patients and the cooperation of multiple centers over a long period of time and patience in not operating except when there is a medical reason such as progression or unexplained pain. As an organization, the members of the Musculoskeletal Tumor Society are perfectly suited to tackle this challenge. We have a new generation of young, bright, enthusiastic members who have demonstrated the ability to organize and accomplish large

prospective studies, develop predictive nomograms for other situations, and obtain federal research funding. Now we need these members to concentrate their focus on this problem. Anyone interested?

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