Enchondroma vs (low-grade) chondrosarcoma: where are we at, right now?

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Learning objectives

- to extensive review the literature and to attempt to identify most helpful imaging features to differentiate a benign from a malignant cartilage tumor, according to recent literature.

- to define "borderline" category.
Background

Enchondromas (EC) are benign cartilage neoplasms of the hyaline cartilage. They are common and occur as an incidental finding in 2.9% of routine knee MRI examinations [1]. Their benign behavior generally warrants no follow-up, except in cases of Ollier disease (multiple enchondromatosis) and Mafucci syndrome (when associated with soft-tissue or visceral hemangiomas) where the risk of secondary malignancy is reported to be as high as 25%.

Chondrosarcoma (CS) of bone is the third most common primary malignant bone tumor after multiple myeloma and osteosarcoma. The most common form of CS is central CS of long bone (conventional variety). CSs are histologically differentiated into grade 1 (low-grade), grade 2 (intermediate grade) and grade 3 (high-grade), dependent on cellularity, cellular atypia and mitosis. Dedifferentiated CS is a highly malignant variant, characterized by the development of a high-grade, non-cartilaginous sarcoma in association with a pre-existing low-grade CS.

Classically, any bone lesion is assigned a presumptive nature according to age of patient, affected bone(s) and location inside the bone, and its morphological features (described ahead). The diagnosis is primarily based in clinical and imaging criteria, with biopsy reserved for doubtful cases. However, the variability of tumors' biological behavior and overlapping morphologic characteristics do not allow for a definitive distinction between their benign or malignant nature. Furthermore, the histologic diagnosis is greatly dependent on the quality and quantity of the biopsy specimen, and intra-tumoral histologic heterogeneity frequently leads to inaccurate diagnosis caused by sampling errors [2].

These issues are mainly addressed when distinguishing between enchondroma and low-grade chondrosarcoma, as generally higher grade malignant tumors exhibit more overt aggressive characteristics. A combination of clinical, radiologic and histologic criteria have been used to strengthen the diagnosis but this has also led to incorporation and diagnostic review biases [1]. Also, interobserver variability occurs even when specific diagnostic criteria are applied, which itself casts doubt on the validation of the published criteria [1]. In some cases, it is simply not possible to predict the biologic behavior of cartilage lesions. In such cases, lesions are classified as "borderline". Fortunately, while the distinction of enchondroma and low-grade chondrosarcoma is difficult, it doesn't carry the prognostic significance attached to separating Grade 1 from Grade 2 tumors, given the fact that grade 1 chondrosarcomas are slowly growing and have a low metastatic potential. However, the fearful dedifferentiated chondrosarcoma, developing in association with a pre-existing low-grade CS, must be sought out.
Pain attributable to the lesion (not related to injury or arthritis), is an indication for surgery as it has been consistently associated with CS.
Imaging findings OR Procedure Details

Location

A lesion located in a flat bone (e.g., spine, ilium, ischium, scapula, rib) is in favor of a CS, as it is rare for EC to be found here, while lesions located in hands and feet favor EC. For other bones however, location doesn't serve much as a discriminator.

Tumoral morphological features

- **Size** - for most, size matters, with a tumor size in excess of 6 cm being suspicious for CS [4]. However, a CS is small at the beginning of its story and large benign lesions may be found, making size an inconsistent discriminator between EC and CS.

- **Margins** - a well-defined but nonsclerotic tumor margin occurs when trabecular or cortical bone abuts the tumor margin. In the diaphysis of long bones, trabeculae are scanty. Therefore, due to the paucity of adjacent trabeculae, the margin of an enchondroma in the diaphysis may appear ill-defined, which is why poorly defined margins cannot be strictly used as a feature of CS.

- **Pattern of calcifications** - several studies have established an association between the type of tumoral calcifications and the probability of malignancy, namely popcorn-calcifications and windblown calcifications [1], [3], but studies didn't find it to be a differential variable [4].

- **Soft tissue mass** - presence of a soft tissue mass is considered as almost pathognomonic for malignancy (fig. 1 and fig. 4). However it is most often encountered in high-grade than in low-grade CS. In Grade 1 CS, a soft tissue mass is seen in only 16% of patients. Therefore, the absence of tumor extension into the soft-tissues does not exclude a central Grade 1 CS [4].

- **MRI features**
  1. the value of the presence of low-signal intensity septae within the tumor on T2-weighted MR images, as well as septal or ring-and-arc enhancement on non-dynamic gadolinium-enhanced MR sequences are controversial;
  2. confluent enhancement was described as a predictor of malignancy [1] (fig. 3 B).
  3. detection of cartilage islands surrounded by fat has also been reported as reliable in differentiating between benign and malignant tumors, however in
the setting of a fracture this sign is unreliable (fat signal change secondary to a hematoma) [5].

The role of MRI remains mostly to determine the extent of the lesion and the presence of an associated soft tissue mass because the main questions are still answered upon a radiograph. Furthermore, in a recent study [1], the number of lesions which were borderline on MRI evaluation was very similar to the number which were borderline on radiographs. Also, in this study, MRI could still result in false negative diagnosis and was associated with increased false positive rates.

**Effect in adjacent bone**

- **Cortical thickening** - cortical thickening has been documented consistently as a reliable sign of CS.

- **Endosteal scalloping** - deep endosteal scalloping (removing more than two thirds of the cortical thickness) is not a distinctive feature of CS. It may be better considered as a correlate of large size and/or subcortical location rather than an independent imaging sign of CS, because it has been found in eccentrically located enchondromas [1].

- **Abnormal peritumoral marrow and soft tissue signal (MRI)** - these abnormalities seem to be found only in chondrosarcoma and may be a criterion of biological activity [3].
Fig. 1  
Chondrosarcoma. Lateral radiograph in a 53 year-old man, shows a well circumscribed large soft tissue mass in the distal femur with matrix, associating cortical breakthrough.

Fig. 2  
Chondrosarcoma. AP radiograph of the distal femur in a 46 year-old man. Chondroid matrix predominates in the proximal portion of the lesion (arrowhead). There is also lytic change distal to the matrix, where there is evidence of cortical destruction (arrow).

Fig. 3  
Central chondrosarcoma. Coronal T1-weighted MR image (A), shows a distal femur lesion with homogeneous hypointensity. Postcontrast T1-weighted image (B) shows peripheral confluent enhancement in favor of chondrosarcoma. Note the diffuse endosteal scalloping and focal cortical breakthrough (arrowhead).
**Fig. 4** Peripheral chondrosarcoma. AP radiograph (A) shows an ill-defined mass with a chondroid matrix arising from the right iliac wing. CT (B) depicts the typical “rings and arcs” pattern of calcification. Axial T2-weighted image (C) demonstrates the extent of the lesion which has a typical high T2 signal.

**Fig. 3**

**Fig. 5** Enchondroma. AP radiograph shows a lesion in the proximal humerus with irregular speckled calcification typical of chondroid matrix, with no signs of aggressiveness.

**Fig. 6** Enchondroma. Coronal CT reconstruction shows a well defined lesion located eccentrically in the femoral neck, without endosteal scalloping nor cortical thickening.
Fig. 7  Enchondroma, MRI appearance. A, coronal T1-weighted MRI shows a hypointense nodular pattern of a nonencapsulated lesion in the proximal humerus. B, Coronal T2-weighted, fat-saturated image of the same lesion shows hypointense calcified matrix (arrow) as well as hyperintense cartilage nodules (arrowhead).

Fig. 5

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Fig. 8  Enchondroma in hands and feet. Fifty percent of enchondromas arise in the hands or feet. Although they present most often with a lytic pattern, there is a wide variety of appearance of cartilaginous matrix. A, AP radiograph of the hand shows in the fourth digit a lytic expanded central lesion, with endosteal scalloping. B, AP radiograph of the hand shows a well defined lytic lesion in the fifth finger with endosteal scalloping. C, AP radiograph of the foot demonstrates a lytic lesion in the hallux. In these cases, the location in the hands and feet makes enchondroma the most likely diagnosis.

Fig. 6

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Conclusion

Despite the application of imaging criteria for assessing the probability of malignancy of a cartilage lesion, "borderline" lesions remain troublesome for radiologists and pathologists, and there is high interobserver variability both in applying the criteria as well as interpreting them. Low-grade chondrosarcoma often cannot be distinguished from enchondroma neither by imaging nor histology. The clinical finding of localized pain which is not attributable to other causes seems to be a more useful indicator than any imaging finding [1].

Outcomes analysis is the most definitive method to distinguish most tumors. Outcomes are easy to determine if lesions are not resected. However, it is difficult to use outcomes to confirm the diagnosis of low-grade chondrosarcoma, since they have a low rate of recurrence and metastasis.

Given the low risk of metastasis from grade 1 chondrosarcoma, serial follow-up may be reasonable instead of curettage for non-painful cartilage lesions of any size.


