CHONDROSARCOMA VARIANTS

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KEYWORDS

• Chondrosarcoma • Dedifferentiated • Mesenchymal • Myxoid • Clear cell • Cartilage

ABSTRACT

This article presents a review of chondrosarcoma variants, with a focus on the extraordinarily rare variants of chondrosarcoma in which hyaline cartilage is not the dominant feature. Discussed are the differential diagnoses for these neoplasms, radiologic studies, gross and microscopic features, and prognosis. Summaries are provided of the key features for the major variants.

OVERVIEW

In addition to classical intramedullary localization, conventional chondrosarcoma of bone rarely may arise from other anatomic sites, including periosteum and synovium. Although criteria for separating benign from malignant chondroid lesions necessarily vary depending on their anatomic site, all share the fact that they are virtually exclusively composed of hyaline cartilage. In this article, the focus is on the extraordinarily rare variants of chondrosarcoma in which hyaline cartilage is not the dominant feature and, in fact, in most cases of myxoid chondrosarcoma (chordoid sarcoma), is absent. Although hyaline cartilage usually does not predominate in these tumors, its presence is essential to establish the diagnosis, the exception being chordoid sarcoma. Dedifferentiated chondrosarcoma, mesenchymal chondrosarcoma, clear cell chondrosarcoma, and chordoid sarcoma generally are easily recognizable (and diagnosable), provided the pathologist is familiar with and/or has encountered them previously. Small biopsy specimens, such as core or fine-needle aspiration biopsies, are especially problematic, as the hyaline cartilage may not be sampled, requiring careful attention to clinicopathologic features and judicious use of ancillary studies to establish a correct diagnosis. Not surprisingly, the differential diagnoses for these neoplasms tend to be related to the dominant cell type but not other cartilaginous neoplasms. For example, the differential diagnosis of dedifferentiated chondrosarcoma usually includes other spindle cell, noncartilaginous pleomorphic sarcomas, whereas mesenchymal chondrosarcoma is more apt to be confused with Ewing sarcoma.

DEDIFFERENTIATED CHONDROSARCOMA

OVERVIEW

Dedifferentiated chondrosarcoma represents approximately 10% of all reported cases of chondrosarcomas. Strictly defined, dedifferentiated chondrosarcoma requires the presence of a high-grade, nonchondrosarcomatous sarcoma usually sharply juxtaposed to a low-grade chondrosarcoma. Although most examples arise from intramedullary, central chondrosarcomas, it also may rarely originate from a preexisting osteochondroma or periosteal chondrosarcoma (peripheral chondrosarcoma).[1,2]

Most patients are older than 50 years at diagnosis. Common anatomic sites include the pelvic bones, femur, and humerus. The most common presenting complaint is pain, often associated with a soft tissue mass.[3,4] Radiologically, the low-grade cartilaginous component shows the typical mineralization of a cartilage tumor, whereas the dedifferentiated portion appears ill-defined,

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permeative, lytic, and associated with extraosseous extension. In many cases, the dedifferentiated component completely dwarfs the smaller, low-grade cartilaginous portion (**Fig. 1**).

GROSS FEATURES

In most circumstances, the 2 components, blue-gray cartilaginous lesion and the fleshy, hemorrhagic to necrotic component, juxtapose one another, with the latter generally dwarfing (in size) the former (**Fig. 2**). For the more common intramedullary form, the cartilaginous component is within bone (central), whereas the dedifferentiated portion extends through the bony cortex, forming a large, extraosseous mass.

MICROSCOPIC FEATURES

In en bloc or resected specimens, the diagnosis of dedifferentiated chondrosarcoma is relatively straightforward if the low-grade cartilaginous component is recognized as malignant. The histologic hallmark is the finding of an abrupt transition between a low-grade chondrosarcoma and a high-grade, often spindle cell, sarcoma, giving the appearance of 2 separate neoplasms (**Fig. 3**); however, focal areas may show a more gradual transition between the 2 components (**Fig. 4**). The

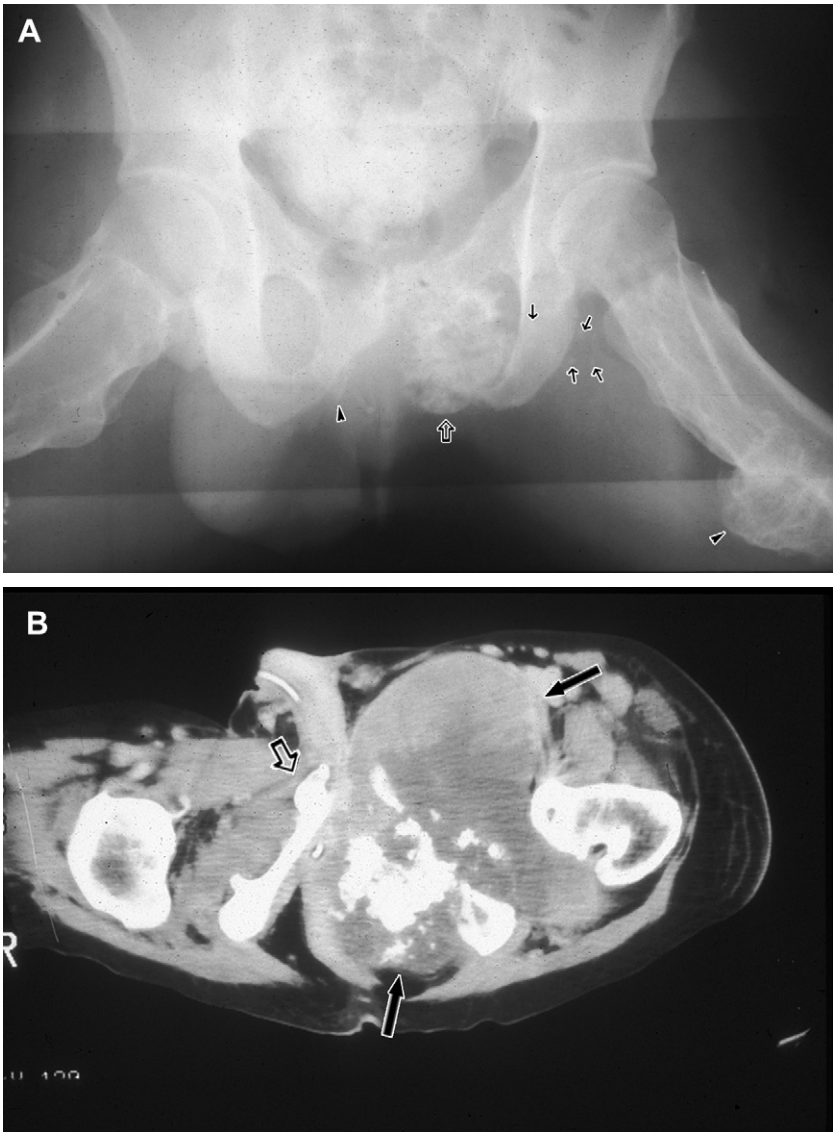

Fig. 1. Dedifferentiated chondrosarcoma arising in a patient with multiple osteochondromatosis. (*A*) An anteroposterior radiograph of the pelvis shows multiple osteochondromas (*arrowheads*). A large soft tissue mass is accompanied by multiple, irregular calcifications at the left pubic ramus (*arrows*). (*B*) An axial computed tomographic scan shows the extension of soft tissue mass anterior (*dark arrow*) and posterior to the left pubic rami. The mass is heterogeneous, probably secondary to necrosis, and calcifications are noted. An osteochondroma (*open arrow*) arises from the right pubic ramus.