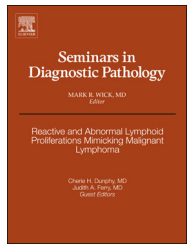
Seminars in Diagnostic Pathology 31 (2014) 10–20

Available online at www.sciencedirect.com

ScienceDirect

www.elsevier.com/locate/semdp



Cartilage-forming tumors

Shadi A. Qasem, MD, Barry R. DeYoung, MD*

Department of Pathology, Wake Forest School of Medicine, Medical Center Blvd, Winston Salem, North Carolina 27157

ARTICLE INFO

Keywords:
Chondroma
Osteochondroma
Chondroblastoma
Chondromyxoid fibroma
Chest wall hamartoma
Chondrosarcoma

ABSTRACT

Cartilage-forming tumors as a group are the most common primary bone tumors; this is largely due to the common occurrence of asymptomatic benign lesions such as osteochondroma and enchondroma. The common feature of these tumors is the presence of chondrocytic cells and the formation of cartilaginous tumor matrix. Some of these tumors are true neoplasms while others are hamartomas or developmental abnormalities. The morphologic heterogeneity of these tumors may be explained by a common multipotent mesenchymal cell differentiating along the lines of fetal–adult cartilage maturation. Recently mutations in IDH1 and IDH2 have been detected in a variety of benign and malignant cartilaginous tumors.[1–4]

Osteochondroma

It is a benign outgrowth of bone and cartilage and is one of the most common bone tumors. Often referred to as exostosis, there is debate whether they constitute a true neoplasm or bony outgrowths of the metaphyseal plate. They may develop following surgery or fracture, and they are the most common radiation-induced benign tumors. Loss of genetic material from the long arm of chromosome 8 has been detected in some of these tumors as well as mutation in exostosin genes. Exostosin genes (EXT) are involved in the biosynthesis of heparan sulfate chains, which are ubiquitous and important in the morphogenesis of cartilage.[1,5–9]

The tumors can be sporadic or multiple and they affect all age groups; however, patients usually present in their first or second decades of life. They are often asymptomatic, but they can cause pain due to pressure on adjacent structures, and sometimes they present as a palpable mass. The most common sites include the long bones (femur, tibia, and humerus), pelvis, and spine.

Radiographically, they manifest as pedunculated or sessile masses on the surface of the bone, adjacent to the metaphyseal plate and pointing away from the joint. The cortex and medulla show continuity with the underlying bone. The cartilaginous cap may not be visible on plain radiograph and is better assessed using CT or MRI (Fig. 1A). Grossly, the lesions take the form of a semicircle or a mushroom-like mass composed of a core of trabecular bone covered by a fairly smooth cartilaginous cap that can vary in thickness. Unfortunately, the specimen is often received fragmented in the surgical pathology laboratory.[1,6]

Microscopically the cartilage cap is composed of mature hyaline cartilage with clusters of chondrocytes in lacunae surrounded by abundant chondroid matrix. The underlying bone shows normal bone trabeculae with intervening bone marrow elements and/or adipose tissue. The transition zone may have blue/purple cartilage and linear arrangement of chondrocytes similar to the growth plate (Fig. 1B).

The differential diagnosis includes other forms of exostoses such as subungual exostosis, osteophytes, and bizarre parosteal osteocartilaginous proliferation (BPOP or Nora's lesion). The majority of these differentials involve the small bones (rare for osteochondroma), are usually small, and do not show continuity with the underlying bone.

* *Corresponding author.*
E-mail address: sqasem@wakehealth.edu. (S.A. Qasem)

http://dx.doi.org/10.1053/j.semdp.2014.01.006
0740-2570

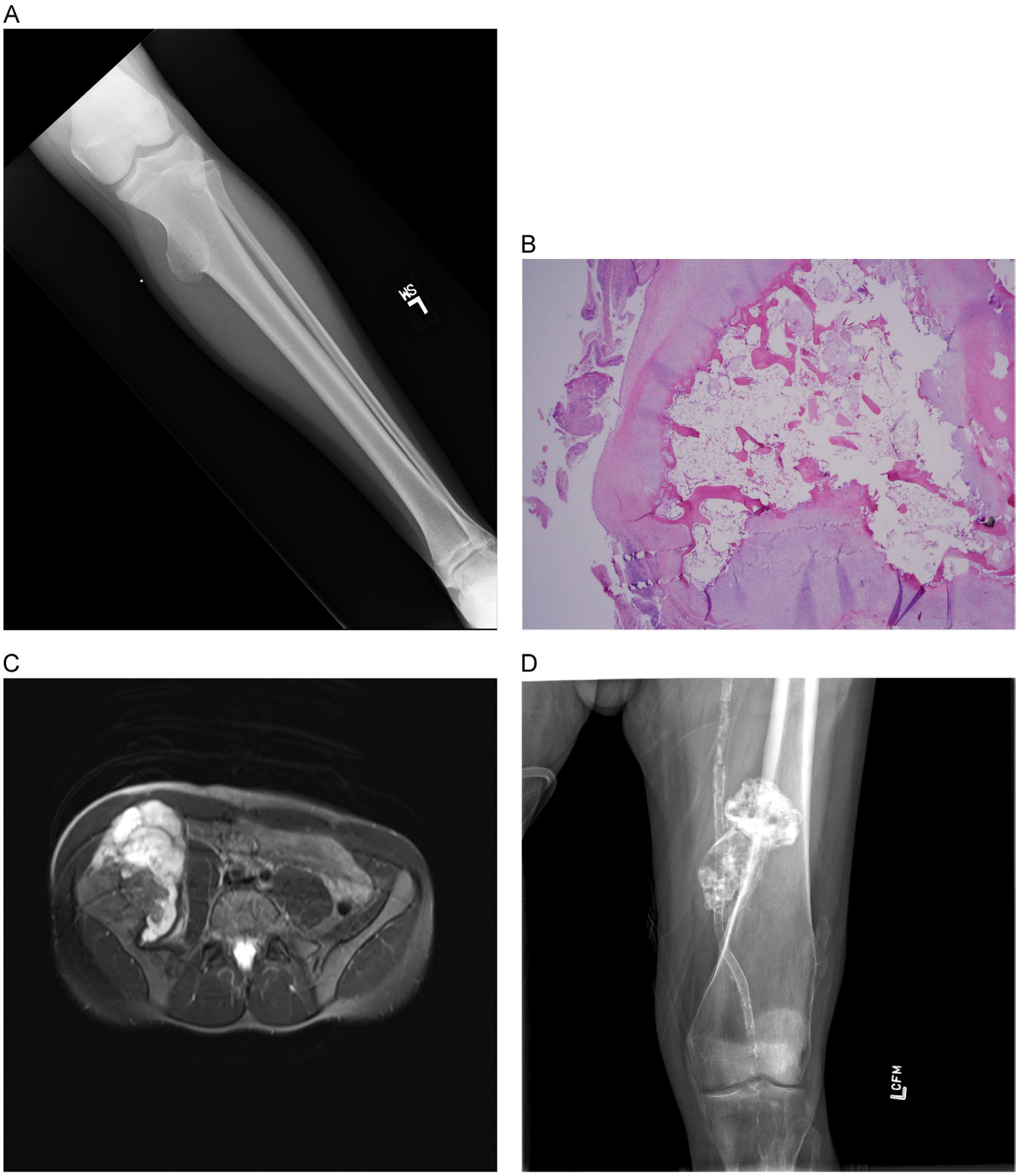


Fig. 1 – (A) An X-ray of leg showing osteochondroma involving the proximal tibial metaphysis and pointing away from the knee joint. (B) Histology of osteochondroma showing thin cartilaginous cap and fatty replaced bone marrow (H&E, 2 ×). (C) MRI of osteochondroma with secondary chondrosarcoma in the ilium. Notice the thick lobulated cartilaginous cap. (D) A patient with multiple osteochondromas of the tibia (there is a vascular bypass graft in the background).

Surgical excision is indicated for symptomatic lesions or those showing atypical features (e.g., rapid growth and thick cap). Recurrence may occur if incompletely excised. Malignant transformation to chondrosarcoma is a small potential risk estimated to affect 1% of patients with solitary lesions and 3–5% of those with multiple ones. A soft tissue mass, thick cartilaginous cap (>2 cm), and/or irregular contours are suggestive features (Fig. 1C). Microscopically, there is