Periosteal Osteosarcoma

14

Carrie Y. Inwards and Doris Wenger

Abstract

Periosteal osteosarcoma is an intermediate-grade chondroblastic osteosarcoma arising on the surface of the bone. It has a slight female predominance. Majority of patients are in the second and third decades of life. The median age is 18 years and average age 22 years. The most common location is the femur followed by the tibia. Less common sites include the humerus, fibula, ulna, and pelvis. Majority involve the diaphyseal region of the bone. Radiologically, the image shows a broad-based attachment of mass to the cortex in the diaphyseal region of the long bone. Histologically, it predominately contains lobules of malignant hyaline cartilage with obvious cytologic atypia. Peripheral condensation and spindling of the tumor cells are commonly seen. Lesser amount of malignant osteoid, often in the form of spicules arrayed perpendicular to the cortical surface, is usually found in the center of the cartilage lobules. Treatment is surgical resection with a wide margin. The role of neoadjuvant and adjuvant chemotherapy is unclear.

Definition

- An intermediate-grade chondroblastic osteosarcoma arising on the surface of the bone

Synonyms

- Juxtacortical chondroblastic osteosarcoma

Etiology

- Unknown

C.Y. Inwards, MD (✉)
Division of Anatomic Pathology, Mayo Clinic, Rochester, MN, USA

College of Medicine, Mayo Clinic, Rochester, MN, USA
e-mail: inwards.carrie@mayo.edu

D. Wenger, MD
Department of Diagnostic Radiology, Mayo Clinic, Rochester, MN, USA

Clinical Features

Epidemiology

- One to two percent of all osteosarcomas

Sex

- Slight female predominance

Age

- Majority of patients are in the second and third decades of life.
- Median age: 18 years; average age: 22 years.

Sites of Involvement

- The most common location is the femur followed by the tibia.
- Less common sites include the humerus, fibula, ulna, and pelvis.
- Majority involve the diaphyseal region of the bone.

E. Santini-Araujo et al. (eds.), *Tumors and Tumor-Like Lesions of Bone: For Surgical Pathologists, Orthopedic Surgeons and Radiologists*, DOI 10.1007/978-1-4471-6578-1_14,

Clinical Signs and Symptoms

- Swelling or mass and pain

Image Diagnosis

Radiographic Features

- Broad-based attachment of mass to the cortex in the diaphyseal region of the long bone
- Cortical thickening and scalloping of the underlying bone
- Periosteal reaction, aggressive and nonaggressive, frequently perpendicular to the osseous long axis (the so-called "hair on end" appearance) extending into surrounding soft tissue

CT Features

- Heterogeneous mass intimately associated with the periosteal surface of the bone with mineralized and nonmineralized components
- Cortical thickening and periosteal reaction at the site of attachment
- Variable degree of mineralization of the mass, with mineralization typically more marked near the base
- Nonmineralized component may be of low attenuation reflecting myxoid change within the chondroid tissue
- Well-defined margins

MRI Features

- Heterogeneous solid mass with mixed signal characteristics on T1 and T2. Soft tissue component often has signal intensity which is isointense or hypointense relative to muscle on T1-weighted images, heterogeneous but predominately hyperintense signal intensity on T2-weighted images, and prominent regions with minimal to no enhancement with gadolinium reflecting the myxoid component of the chondroid tissue.
- Well-defined margins.
- Cortical thickening and scalloping.
- Areas of poorly marginated hazy abnormal signal intensity (low signal on T1-weighted images and high signal on T2-weighted images) in the soft tissues and marrow about the mass compatible with reactive change.

Bone Scan

- Marked uptake of radionuclide, eccentrically positioned on the bone

Imaging Differential Diagnosis

Parosteal Osteosarcoma

- Involves the metaphyseal region of the underlying bone
- More heavily mineralized with mineralization throughout the entire lesion, including the surface of the mass

Parosteal Chondrosarcoma

- Involves the metaphyseal region of the underlying bone
- Lacks perpendicular periosteal reaction

High-Grade Surface Osteosarcoma

- Usually surrounds a greater amount of the bone circumference
- Lacks features of myxoid change suggestive of cartilaginous component (low attenuation at CT, hyperintense signal intensity on T2-weighted and minimal enhancement on gadolinium-enhanced MR images)

Pathology

Gross Features

- Firm, lobulated gray-white mass resembling cartilage.
- Broad-based attachment to the underlying bone.
- Tumor wraps around the underlying bone with occasional full circumferential envelopment.
- Well-defined outer pushing margin.
- Focal cortical erosion by the tumor with minimal medullary invasion only rarely present.
- Average size: 10 cm.

Histological Features

- Predominately contains lobules of malignant hyaline cartilage with obvious cytologic atypia. Peripheral condensation and spindling of the tumor cells are commonly seen.
- Cartilage matrix may contain areas with myxoid change.
- Lesser amount of malignant osteoid, often in the form of spicules arrayed perpendicular to the cortical surface, is usually found in the center of the cartilage lobules.

- Occasionally, areas composed of malignant spindle cells, with or without osteoid formation, blend with the hyaline cartilage portion of the tumor.
- The degree of cytologic atypia places these tumors into an intermediate-grade osteosarcoma.

Pathologic Differential Diagnosis

Parosteal Osteosarcoma

- Predominately composed of low-grade fibro-osseous tissue. Low-grade cartilage a minor component

Parosteal Chondrosarcoma

- Entire tumor composed of malignant hyaline cartilage with or without focal areas of bland metaplastic bone formation

High-Grade Surface Osteosarcoma

- Composed of high-grade tumor cells producing abundant malignant osteoid

Bizarre Parosteal Osteochondromatous Proliferation

- Contains a cytologically benign cartilage cap that undergoes enchondral ossification to trabeculae of the bone surrounded by a bland fibrovascular stroma

Osteochondroma

- Benign cartilage cap undergoing enchondral ossification into benign underlying trabeculae of the bone surrounded by fatty marrow

Genetics

- No consistent cytogenetic abnormalities

Prognosis

- Better prognosis when compared to intramedullary conventional osteosarcoma
- Overall 5-year and 10-year survival of 89 % and 84 %, respectively
- Local recurrence uncommon, but associated with a higher incidence of metastases and death

Treatment

- Surgical resection with a wide margin
- Amputation only if an adequate surgical margin cannot be achieved by en bloc resection
- Role of neoadjuvant and adjuvant chemotherapy unclear

Images

See Figs. 14.1, 14.2, 14.3, 14.4, 14.5, 14.6, 14.7, and 14.8 for illustrations of periosteal osteosarcoma.

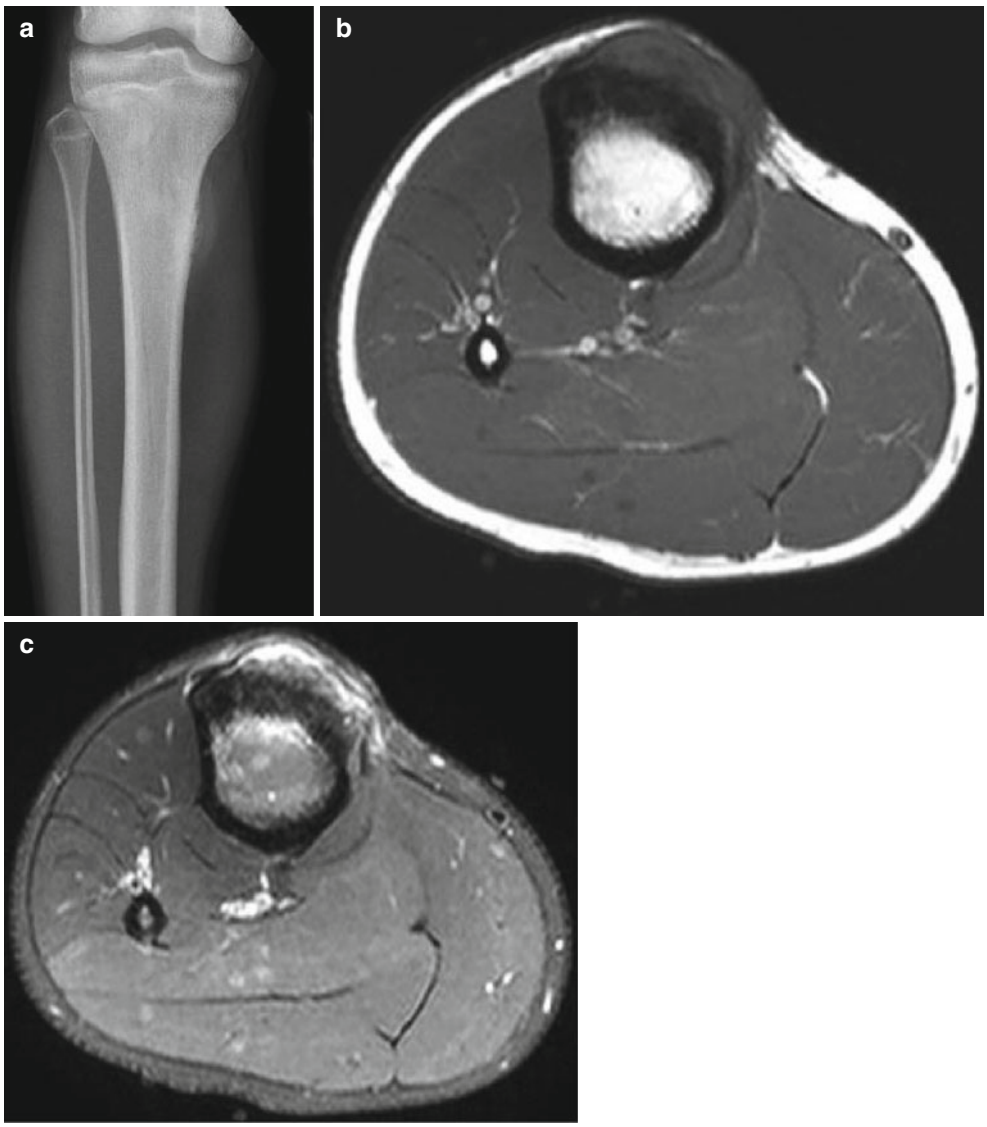
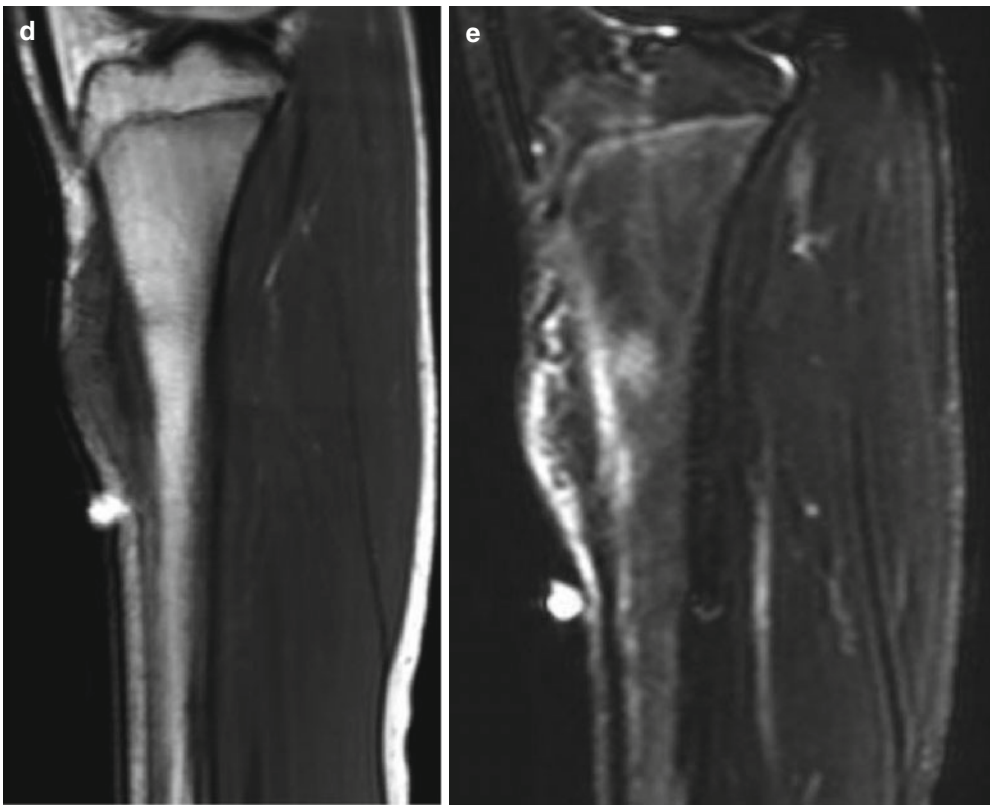


Fig. 14.1 AP radiograph (**a**) of a 15-year-old male shows a region of cortical thickening, aggressive periosteal reaction, and subtle matrix production along the medial aspect of the proximal tibial diaphysis with the suggestion of an associated unmineralized soft tissue mass. The axial and sagittal T1 (**b**, **d**) and axial and sagittal fat-suppressed T2 weighted (**c**, **e**) MR images show that the mass involves the surface of the bone without involvement of the medullary canal. The mass is heterogeneous with a region of predominantly low T2 signal intensity near the attachment to the cortex that correlates with the periosteal reaction and matrix production on the radiograph. In addition, there is an unmineralized component to the soft tissue mass with nonspecific intermediate T1 and hyperintense T2 signal

Fig. 14.1 (continued)



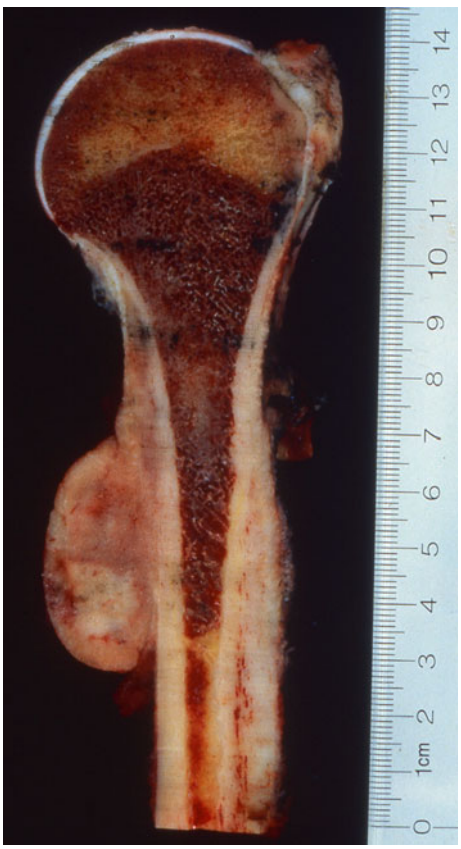


Fig. 14.2 Periosteal osteosarcoma forming a surface-based mass involving the diaphyseal portion of the underlying bone. The tumor contains zones of *gray-white* tissue reflecting areas of cartilaginous differentiation

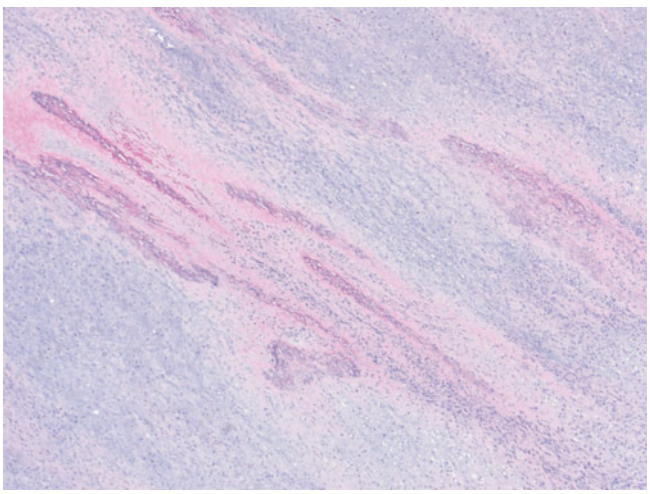


Fig. 14.3 Periosteal osteosarcoma showing histological features of a chondroblastic osteosarcoma. The malignant osteoid forms spicules of bone arranged perpendicular to the surface of the underlying cortical bone

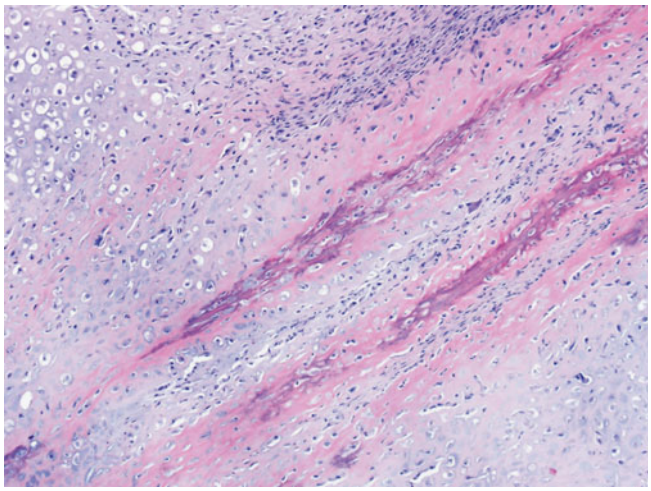


Fig. 14.4 Periosteal osteosarcoma composed of an intermediate-grade chondroblastic osteosarcoma

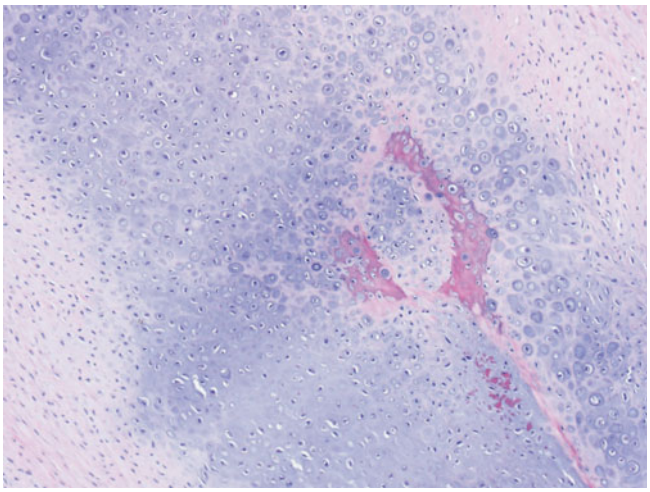


Fig. 14.7 Spicules of malignant osteoid within the central portion of a cartilaginous nodule in periosteal osteosarcoma

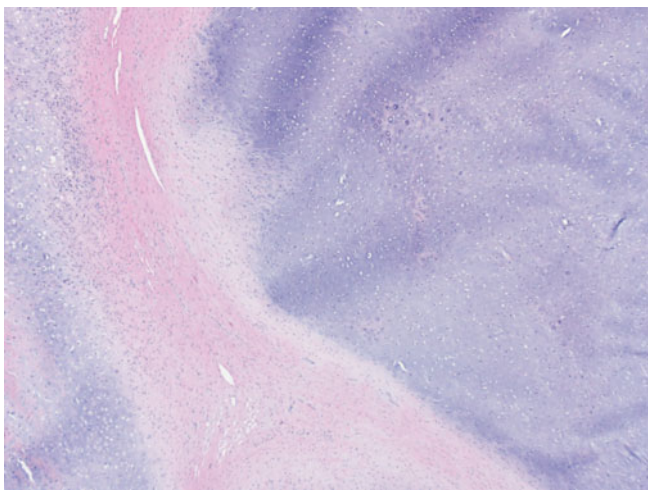


Fig. 14.5 The predominant tissue type in periosteal osteosarcoma is malignant hyaline cartilage

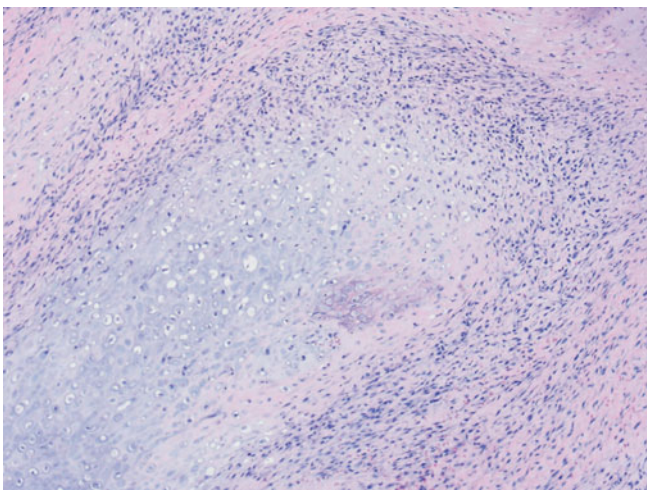


Fig. 14.8 Peripheral condensation of tumor cells at the periphery of the cartilaginous nodules is frequently seen in periosteal osteosarcoma

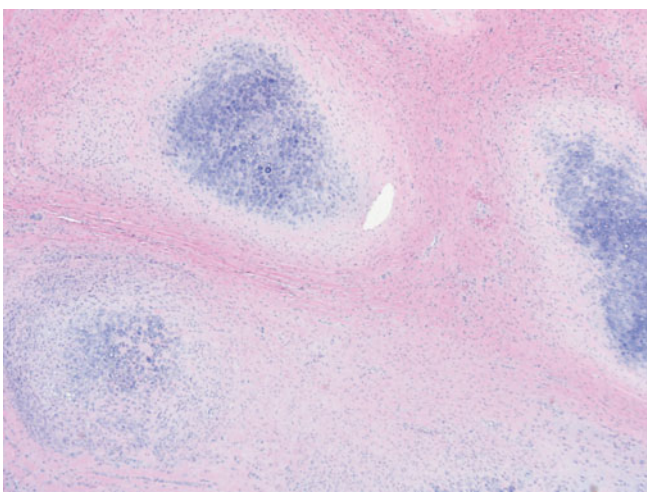


Fig. 14.6 Periosteal osteosarcoma containing nodules of malignant hyaline cartilage permeating peripheral portions of fibrous tissue

Recommended Reading

Cesari M, Alberghini M, Vanel D, Palmerini E, Staals EL, Longhi A, et al. Periosteal osteosarcoma: a single-institution experience. Cancer. 2011;117(8):1731–5. doi:10.1002/cncr.25718. Epub 2010 Nov 8.

Grimer RJ, Bielack S, Flege S, Cannon SR, Foleras G, Andreeff I, European Musculo Skeletal Oncology Society, et al. Periosteal osteosarcoma–a European review of outcome. Eur J Cancer. 2005;41(18):2806–11. Epub 2005 Nov 14.

Murphey MD, Jelinek JS, Temple HT, Flemming DJ, Gannon FH. Imaging of periosteal osteosarcoma: radiologic-pathologic comparison. Radiology. 2004;233(1):129–38. Epub 2004 Aug 27.

Revell MP, Deshmukh N, Grimer RJ, Carter SR, Tillman RM. Periosteal osteosarcoma: a review of 17 cases with mean follow-up of 52 months. Sarcoma. 2002;6(4):123–30.

Rose PS, Dickey ID, Wenger DE, Unni KK, Sim FH. Periosteal osteosarcoma: long-term outcome and risk of late recurrence. Clin Orthop Relat Res. 2006;453:314–7.

Unni KK, Dahlin DC, Beabout JW. Periosteal osteogenic sarcoma. Cancer. 1976;37(5):2476–85.