A 15-year-old adolescent boy presented with a swelling of the anterior and inner aspect of the proximal right tibia that was growing slowly during the 3-month period prior to presentation. The swelling became painful after a minimal injury caused by a fall. A firm, palpable mass was present on physical examination. The remainder of the findings from the examination were unremarkable. Radiography showed a juxtacortical heterogeneous mass measuring 8 cm in length on the proximal tibial diaphysis (Figure 1). Magnetic resonance imaging demonstrated a well-defined periosteal lesion with a cortical lysis but without involvement of the medullary cavity. On magnetic resonance imaging, the lesion had a hypo signal intensity on the T1-weighted image, a high signal intensity on the T2-weighted image, and an intense enhancement after gadolinium injection. A biopsy initially showed hypercellular and ill-defined lobules of moderately to well-differentiated neoplastic cartilage. The tumor cells showed moderated nuclear pleomorphism, enlargement, and hyperchromasia. There were scattered areas of necrosis and endochondral ossification on the surface of some lobules but no evident areas of malignant osteoid or bone formation. The tumor infiltrated soft tissues. The initial diagnosis was low-grade juxtacortical chondrosarcoma. Later, a wide and partial resection of the tibia was performed. The specimen measured 13 cm in length and 6 cm in diameter. A longitudinal section of the specimen showed an extraosseous, fusiform, and well-demarcated tumor of 6 × 2 cm surrounding the diaphyseal bone and elevating the periosteum. The cut surface was somewhat pearly and chondroid (Figure 2). The underlying cortex seemed intact. Microscopically, the tumor developed from the periosteum, eroding the underlying cortex without involving the medullary cavity. It was composed of large lobules of a chondromyxoid matrix that contained neoplastic cells with enlarged and hyperchromatic nuclei, often binucleated. In rare areas, the tumor presented hypercellular spindled or more rounded cells contained in areas with a fine lacelike or more pronounced osteoid formation between tumor cells at the periphery of the cartilaginous lobules (Figure 3). The margins of the specimens were free of tumor cells.

What is your diagnosis?
Pathologic Quiz Case—Aubert et al

Pathologic Diagnosis: Periosteal Osteosarcoma

Periosteal osteosarcoma was first described by Unni et al. It is a rare variant of osteosarcoma arising from the cortical surface with a predilection for the tibia and the femur. Less than 2% of all osteosarcomas are periosteal osteosarcomas, with a 1:1.7 male-female ratio. The peak incidence is during the second decade of life, and most patients present after a few weeks or months of pain, swelling, tenderness, or a mass. In our case, mainly on the basis of biopsy specimens, the main differential diagnosis was juxtacortical chondrosarcoma. Historically, there was confusion between these 2 entities. It is now widely accepted that periosteal osteosarcoma is a chondrogenic bone-forming tumor arising on the surface of long bones and that juxtacortical chondrosarcoma is an exclusive chondrogenic surface tumor. Periosteal osteosarcoma has a high local recurrence after local excision and a metastatic rate of 15%, whereas juxtacortical chondrosarcoma rarely recurs and exceptionally metastasizes. Therefore, these 2 distinct entities must be differentiated. Juxtacortical chondrosarcoma tends to occur chiefly in the third or fourth decade of life with a male predominance. Periosteal osteosarcoma more frequently involves a diaphyseal location, whereas juxtacortical chondrosarcoma is usually metaphyseal in location. Radiologically, periosteal osteosarcoma is a radiolucent and fusiform lesion that presents brushlike spicules of bone extending perpendicularly from a relatively normal underlying cortex. Juxtacortical chondrosarcoma is more bulky and round with popcorn-shaped opacities and shows a saucerclike depression with a thickening and sclerosis of the underlying cortex. Histologically, juxtacortical chondrosarcoma is composed of coalescing lobules of malignant cartilage with foci of myxoid change, calcification, and surface endochondral ossification. This ossification is solely reactive with no bone-forming malignant osteoblastic cells in contrast to periosteal osteosarcoma. Malignant osteoid or bone formation can be very focal in periosteal osteosarcoma. Therefore, multiple sections of biopsy specimens and extensive sampling of the resected specimen must be examined to demonstrate direct tumor ossification. Often, larger cartilaginous benign tumors such as periosteal chondroma may suggest a diagnosis of periosteal osteosarcoma, which is usually ruled out by microscopic examination. Other surface osteosarcomas must be distinguished from periosteal osteosarcoma. Periosteal osteosarcoma has an intermediate prognosis between low-grade parosteal osteosarcoma and high-grade surface osteosarcoma. Parosteal osteosarcoma is more common in the third to fifth decades of life, more frequently affects women than men, and is considered a low-grade osteosarcoma, with infrequent metastases and a low incidence of relapse. The single most common site of parosteal osteosarcoma is the posterior aspect of the distal femur. Radio logically, parosteal osteosarcoma differs from periosteal osteosarcoma by its relatively well-defined, bosed, and radio-opaque appearance; by its relatively broad attachment to the underlying cortex giving a typical radiolucent line between tumor and cortex; and by its lack of elevated peristeum and periosteal new bone formation. Microscopically, parosteal osteosarcoma presents only foci of cartilage at the periphery of the tumor on a background of relatively innocuous spindle cells that produce well-formed bony trabeculae. In contrast, periosteal osteosarcoma presents a typical thin ramification or lace-like osteoid cascading outward from the periphery of the cartilage lobules. High-grade surface osteosarcoma may consist predominantly of chondroblastic elements, such as a chondroblastic variant of conventional osteosarcoma, which can be misdiagnosed for a periosteal osteosarcoma. But the chondroblastic elements are more likely anaplastic in high-grade surface osteosarcoma, and any high-grade area with pleomorphic fibroblastic or osteoblastic cells will preclude a diagnosis of periosteal osteosarcoma. A diagnosis of conventional osteosarcoma is ruled out since it shows medullary involvement by either radiology or histology. Indeed, medullary involvement is by definition absent in periosteal osteosarcoma. Periosteal osteosarcoma is a rare surface chondroblastic variant of conventional osteosarcoma, usually treated by wide excision. Its prognosis is better than that of high-grade intramedullary or high-grade surface osteosarcomas but worse than that of parosteal osteosarcomas or juxtacortical chondrosarcomas.

References