administered to the patient with the resulting tumor developing in the path of the radiation beam; (3) the existence of a latency period of at least 3 to 4 years between the administration of radiation therapy and sarcoma development; (4) the existence of histologic proof of a sarcoma different from the irradiated primary lesion.

The secondary neoplasm usually arises in bone, although it may also occur in the soft tissues. In a 1985 study of 66 patients with postradiation sarcomas, 59 lesions were osseous and 7 developed in the soft tissue. In a 2001 study, 52 of 63 postradiation sarcomas (83%) were believed to have arisen in bone with the remaining 17% originating in soft tissue.

**Clinical Presentation and Incidence**

The characteristic clinical presentation of a postradiation sarcoma is an abrupt onset of swelling and pain with an associated palpable mass in a previously irradiated location (Figure 1). For lesions arising in bone, imaging studies often show bone destruction, periosteal reaction, and a soft-tissue mass. For sarcomas arising in soft tissue, imaging studies always show a tissue mass and may show adjacent bony destruction; however, periosteal reaction or expansion of the bone will be absent.

Although the true incidence of postradiation sarcomas is difficult to determine, the overall incidence in adults appears to be low. However, because patients who receive radiation therapy are now living longer, the relative risk will increase with time. A population-based study was performed in Switzerland to quantify the risk of secondary soft-tissue sarcoma after treatment for breast cancer. Follow-up on 122,991 patients (74% received radiation therapy) between 1958 and 1992 showed the development of 116 sarcomas. The mean age for diagnosis of the primary breast malignancy was 58.7 years and the mean age for presentation of the secondary sarcoma was 68.4 years. The mean time between diagnoses and presentation was 10.1 years (range, 1.1 to 30.9 years); most secondary sarcomas occurred near the involved breast or on the ipsilateral arm. The mean dose of radiation given was 188 J (defined as energy imparted to the patient). Forty of the secondary malignancies were angiosarcomas, which the authors believed correlated more with lymphedema of the arm than with the radiation therapy. The remaining malignancies were fibrosarcomas, malignant fibrous histiocytomas, or other soft-tissue sarcomas; in these lesions the radiation dose positively correlated with their occurrence. Several more recent reports have also established a relationship between radiation therapy for breast carcinoma and postradiation angiosarcoma of the breast.

Children who receive radiation therapy for Ewing’s sarcoma, bilateral retinoblastoma, and soft-tissue sarcoma have the highest risk of postradiation sarcoma because of the hereditable nature of these conditions and the resulting genomic instability. Children who survive the initial disease also have an increasing latency period as they age, putting them at an additional risk for the development of a secondary neoplasm. The incidence of postradiation sarcoma in children ranges from 5.5% to 35% at 20 years after radiation therapy for Ewing’s sarcoma. Other studies of children treated with radiation therapy for Ewing’s sarcoma using doses of less than 60 Gy suggest a 3% to 7% risk for the development of sarcoma by 20 years after treatment. A significantly higher risk is likely for those treated with radiation doses of more than 60 Gy.

Other studies show a wide range of latency periods (3 years to more than 30 years) between the initial radiation treatment and the development of the secondary neoplasm. One study reported a mean latency period of 7.6 years. It appears that the latency period is inversely related to the radiation dose, although this theory is controversial. It also has been suggested that below a threshold of 10 Gy, the risk of a secondary neoplasm is
There is active growth over time, and they can be moderately tender on examination. Stage 2 bone lesions remain encapsulated and have a thin layer of reactive tissue between the lesion and surrounding structures. Radiographs show areas of trabecular bone destruction with or without cortical involvement. There may be soft-tissue extension. Despite their progressive growth, stage 2 lesions remain limited by natural barriers. The bone destruction and soft-tissue extension are limited. The integrity of the articular surfaces and the continuity of the diaphysis, metaphysis, and epiphysis are not threatened. Many benign bone entities are stage 2 active lesions. Common examples are most giant cell tumors, chondroblastomas, chondromyxoid fibromas, osteoid osteomas, and most osteoblastomas.

**Stage 3**

Stage 3 benign, aggressive bone lesions cause marked bone destruction, soft-tissue extension, or pathologic fractures. The aggressive biologic behavior of stage 3 lesions makes their removal and reconstruction very difficult. In some instances, resection of the bone and reconstruction with either an allograft or prosthesis is necessary. Patients with stage 3 bone lesions report pain and/or an enlarging mass and may have a pathologic fracture. These lesions can grow rapidly and permeate through natural barriers including cortical bone, fascial layers, articular cartilage, and joint capsules. The radiographic characteristics of an aggressive lesion may include a more permeative appearance at the interface between tumor and normal bone. Cortical bone destruction and periosteal reaction often occur. Some benign, aggressive lesions such as giant cell tumors can develop pulmonary metastasis despite their histologically benign appearance. Examples of benign aggressive tumors include some giant cell tumors, osteoblastomas, and aneurysmal bone cysts. These aggressive lesions are difficult to remove, reconstruction is challenging, and they have a 25% or higher rate of local recurrence.

An overlap in assigning the stage to a benign bone tumor can occur. For example, it may be arbitrary whether to classify a giant cell tumor of the distal femur with cortical destruction and a moderate soft-tissue mass (3 to 5 cm) and preservation of the articular cartilage as a stage 2 active or stage 3 aggressive lesion. Enneking originally estimated that 15% of giant cell tumors of bone were stage 1 lesions, 70% stage 2, and 15% stage 3 lesions.

**Evaluation and Assignment of Stage**

Orthopaedic surgeons and oncologists often evaluate patients with bone lesions to determine the need for biopsy and eventual treatment. Although approximately 3,000 malignant bone tumors occur each year in the United States, many thousands of benign bone neoplasms and reactive and developmental conditions also occur. The orthopaedic surgeon must use a systematic approach to ensure that those lesions are classified according to the correct staging system (benign versus malignant). After a careful patient history, physical examination, and review of imaging studies, a determination is made on whether a stage can be assigned to the lesion without a biopsy. The initial treatment decision often involves the choice of observation or incisional, needle, or excisional biopsy.
mor. In recent years, methylmethacrylate has been used more routinely than bone graft in this patient population. Methylmethacrylate is composed of a monomer and polymer combined in a 2:1 ratio of powder to liquid. Its use allows immediate support of the bone and decreases the amount of time needed before full weight bearing in patients with lower extremity lesions. Some authors believe that there is an additional thermal adjuvant effect with methylmethacrylate that independently decreases local tumor recurrence. In addition, recurrence can be easily recognized as a lytic area adjacent to the radiopaque methylmethacrylate. It should be emphasized that the importance of performing an adequate curettage far outweighs the type of reconstruction in preventing local recurrence.

Tumor ablation may be performed without using open surgical techniques. Closed techniques may be used for some less aggressive lesions such as unicameral bone cysts or osteoid osteomas. Closed ablation techniques include steroid injection, percutaneous bone grafting, and radiofrequency ablation. A technique of percutaneous injection has been used in the treatment of intact active unicameral bone cysts. This procedure involves injecting methylprednisolone into the cyst under fluoroscopic control after aspirating the cyst to confirm straw-colored fluid. Radiopaque dye is injected to confirm entry into the cyst. If grossly bloody fluid is encountered, a formal biopsy is advised to ascertain whether the lesion is an aneurysmal bone cyst or a unicameral bone cyst with a fracture. The cyst is then flushed with saline, and methylprednisolone (80 to 160 mg depending on the size of the cyst) is injected. Steroid injection is the current treatment of choice for unicameral bone cysts; however, this procedure has not

Figure 2  AP (A) and lateral (B) radiographs of the wrist of a 43-year-old man with a giant cell tumor. There is no cortical destruction or soft-tissue mass. AP (C) and lateral (D) radiographs after curettage and cementation of the lesion. The wrist joint was maintained and the patient was free of local recurrence 2 years after surgery.
Introduction
Soft-tissue sarcomas represent a comprehensive group of connective tissue malignancies that are capable of invasive, locally destructive growth and have a tendency to recur and to metastasize. Sarcomas, however, have varying characteristics. Some sarcomas, such as dermatofibrosarcoma protuberans, rarely metastasize. In contrast, undifferentiated high-grade pleomorphic sarcoma often metastasizes to the lungs. Myxoid liposarcoma may metastasize to retroperitoneal and other non-pulmonary sites. This chapter will discuss the more common soft-tissue sarcomas.

Supposed Fibrohistiocytic Tumors
Until recently, malignant fibrous histiocytoma (MFH) has been the most common soft-tissue sarcoma that occurs in adults; however, the origins of the cell type(s) remain unclear. Current debate involves whether MFH is a distinct entity or a diverse group of sarcomas with a histologically similar appearance. The most current sarcoma classification from the World Health Organization (WHO) no longer includes MFH as a distinct entity. The current nomenclature for most MFHs is undifferentiated pleomorphic sarcoma (Figure 1). Historically, five different histologic subtypes of MFH have been described in the literature. Only three subtypes, storiform-pleomorphic, giant cell, and inflammatory histiocytoma, are now recognized as components of MFH. Myxoid MFH has been reclassified as myxofibrosarcoma (Figure 2). In 1994, the angiomatoid subtype was reclassified by the WHO as an intermediate fibrohistiocytic tumor. It is now called angiomatoid fibrous histiocytoma.

The previously accepted clinical and histologic characteristics of the MFH subtypes are still in clinical use by some pathologists and are outlined in Table 1.

Liposarcomas
Liposarcoma is the second most common soft-tissue sarcoma after undifferentiated high-grade pleomorphic sarcoma, often occurs in older patients, and can be large