The case:

An 11-year-old girl presented with a painful lump on the medial aspect of her left knee and progressive pain and fullness of the lateral aspect of the distal left leg. The patient had a medical history of stage 4 right adrenal neuroblastoma diagnosed at age 3, treated with excision, chemotherapy, and abdominal irradiation.

![Anteroposterior (A) and lateral (B) radiographs of the left leg.](image)

**Figure:** Anteroposterior (A) and lateral (B) radiographs of the left leg.

Your diagnosis?

*For answer see page 886*
Diagnosis:
Chondroblastic Osteosarcoma and Osteochondromas After Prior Total Body Irradiation

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Answer to Radiologic Case Study
Case facts appear on page 820

A 11-year-old girl presented with a painful lump on the medial aspect of her left knee and progressive pain and fullness of the lateral aspect of the distal left leg. The patient reported no trauma, weight loss, fever, or fatigue. Physical examination revealed an antalgic gait, increased circumference of the left leg (32 vs 29 cm at midshaft), and tenderness to palpation at the lateral aspect of the left fibula. Of note, the patient’s medical history revealed a stage 4 right adrenal neuroblastoma at age 3 years, treated with excision, chemotherapy, and 1000 cGy of abdominal irradiation. The patient had also received whole-body radiation as part of a conditioning regimen for peripheral stem cell transplant, although the exact dose was unknown. No family history of hereditary multiple exostoses was reported.

Imaging
Radiography

Radiographs of the left leg revealed sessile lateral distal femoral and pedunculated medial proximal tibial metaphyseal osteochondromas (exostoses), both of which demonstrated a classic imaging appearance for osteochondroma (Figure 1).

Figure 1: Anteroposterior (A) and lateral (B) radiographs demonstrating sessile distal femoral and pedunculated proximal medial tibial osteochondromas (arrowheads) without complicating features. In addition, a destructive mineralized mass involves the mid and distal fibular diaphysis (arrows), containing both osteoid- and chondroid-type matrix production.

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The authors have no relevant financial relationships to disclose.

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doi: 10.3928/01477447-20131021-01

aggressive periosteal reaction, cortical destruction, and suggestion of circumferential
Further imaging with computed tomography (CT) and magnetic resonance imaging (MRI) was performed to assess the extent of intraosseous lesion involvement and degree of soft tissue extension into the adjacent muscular compartments.

Computed Tomography
Computed tomography can confirm the cortical and medullary continuity of an osteochondroma with the parent bone and can detect nondisplaced fractures that may be occult on radiographs. However, CT is inferior to MRI in its capability to assess a thickened cartilaginous cap and differentiate the overlying cartilaginous cap with adjacent bursitis from malignant transformation.\(^1\) Computed tomography is excellent in detecting mineralized tumor and cortical destruction in the setting of osteosarcoma (Figure 2) but can be limited in its differentiation of an unmineralized tumor component from the adjacent muscle or bursa.\(^1\)

Magnetic Resonance Imaging
Magnetic resonance imaging provides optimal visualization and characterization of the osseous and/or soft tissue component of benign and
Magnetic resonance imaging defined the anatomic extent of the intramedullary and extraosseous components of this tumor and played a critical role in surgical planning.5

**Diagnosis**

Ultrasound-guided needle biopsy of the fibular lesion yielded malignant osteoid-producing spindle cells with admixed foci of chondroblastic matrix (Figure 5). A diagnosis of grade 3 chondroblastic osteosarcoma was made and confirmed with intraoperative histology at the time of definitive resection. Chondroblastic osteosarcoma is characterized histologically by nodules of myxoid matrix and neoplastic chondrocytes surrounded by osteoid, and ossification may occur at the periphery of the cartilaginous mass or within the spindle component of the lesion.6,7

A presumptive diagnosis was made of distal femoral and proximal tibial metaphyseal osteochondromas based on their classic imaging features. Although the patient’s diagnosis of radiation-induced osteosarcoma and chondroblastic osteosarcoma (combined with a history of adrenal neuroblastoma) raises the question of a genetic predisposition to neoplasm development, to the authors’ knowledge genetic testing has not been performed to date.

**Incidence of Radiation-Induced Osteosarcoma and Osteochondroma**

Ionizing radiation is associated with various skeletal complications, including osteonecrosis, scoliosis, growth retardation, bowing, osteochondromas, and osteosarcoma.8-11 Concurrent radiation-induced osteosarcoma and osteochondroma has been reported in the literature, but the incidence is rare.12 Radiation-induced osteochondroma is a benign osseous excrescence with a cartilaginous cap that occurs in up to 24% of patients irradiated prior to age 5 years.6,9 Osteochondromas form by endochondral ossification, with the hyaline cartilage cap functioning as the epiphyseal plate.1,9 Lesions typically remain stable after skeletal maturity; thus, pain or enlargement after epiphyseal closure may indicate malignant degeneration.14 Osteochondromas are 10 times more common in radiation-exposed patients, occurring in up to 28% of children irradiated prior to age 3 years.11 Lesions are typically diagnosed 5 to 8 years after exposure and are indistinguishable from primary lesions by imaging and histology.3,8,9

Osteosarcoma is an extremely rare complication of radiation therapy, with an incidence of 0.01% to 0.20% among all cases of osteosarcoma, but they account for more than 90% of radiation-induced skeletal malignancies.5,8 Whereas exostoses have been associated with as little as 900 cGy, osteosarcoma is nearly exclusive to those receiving 10,000 cGy or more.5 Radiation-induced osteosarcoma has an average latent period of 13 years, with lesions reported up to 33 years after treatment.5,10

**Treatment**

Treatment of radiation-induced osteosarcoma is chemotherapy and either limb-sparing wide-margin resection or amputation. Regardless of the surgical method used, the ultimate goal is complete tumor eradication with a wide surgical margin of at least 2 to 3 cm.6

The current patient received induction chemotherapy with adriamycin, cisplatin, and methotrexate, followed by wide-margin, en-bloc, mid-to-distal fibular resection of the chondroblastic osteosarcoma (Figure 6). No treatment was required of the patient’s osteochondromas at the time of sarcoma resection. Of note, symptomatic osteochondromas may occasionally require surgical excision if they fracture or if mechanical pressure of the exostosis on the adjacent structures results in neurovascular compression, adventitial bursitis, or pseudoaneurysm formation.8

Finally, follow-up is crucial for proper management of patients diagnosed with radiation-induced malignancy. Osteosarcoma recurrence is most common in the first 2 years after resection, and it is recommended patients undergo chest CT and radiographic and MRI surveillance of the operative site every 3 months and bone scintigraphy every year during this period. Thereafter, yearly radiographs should be obtained indefinitely, as osteosarcoma can recur decades after treatment.5,13

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**Figure 6:** Postoperative anteroposterior radiograph of the left leg demonstrating change after wide resection of the majority of the fibular diaphysis (arrows). Note the stable proximal medial tibial pedunculated osteochondroma.

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REFERENCES


