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Authors
Dong, Paul R
Seeger, Leanne L
Eckardt, Jeffrey J
et al.

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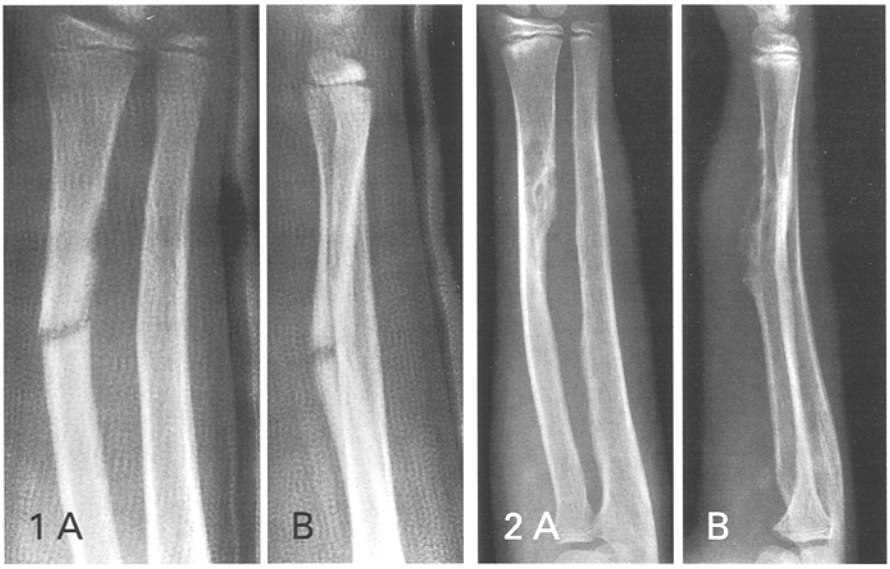
Case report 847

Paul R. Dong, M.D.[1], Leanne L. Seeger, M.D.[1], Jeffrey J. Eckardt, M.D.[2], Joseph M. Mirra, M.D.[3]

[1]Department of Radiological Sciences, UCLA School of Medicine, Los Angeles, California, USA
[2]Department of Orthopaedic Surgery, UCLA School of Medicine, Los Angeles, California, USA
[3]Hospital of the Good Samaritan, Los Angeles, California, USA



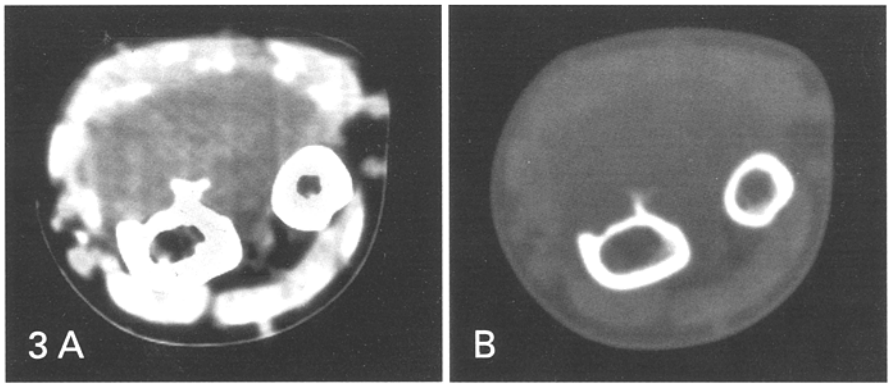


Fig. 1. A Anteroposterior and **B** lateral radiographs taken following the second fracture. Bony detail is somewhat obscured by the cast. The radius is mildly bowed in a lateral and ventral direction at the level of the fracture. A 1.5-cm-long lucency is evident along the ulnar aspect of the bone distal to the fracture

Fig. 2. A Anteroposterior and **B** lateral radiographs at the time of presentation. The fractures have healed. The radius remains bowed. There is benign-appearing saucerization of the mid-radial shaft associated with mature periosteal new bone

Fig. 3A,B. Computed tomographic scan without intravenous contrast: soft tissue (**A**) and bone (**B**) windows. The lesion is of lower density than muscle, and is clearly shown to originate from the surface of the radius. The interosseous membrane is displaced

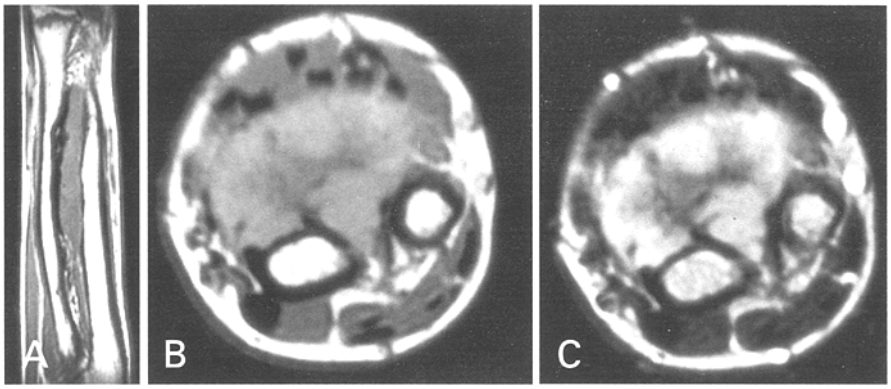


Fig. 4A–C. Magnetic resonance scan. **A** Coronal T1-weighted image (SE 600/11). The tumor is interposed between the radius and ulna and is isointense to muscle. The marrow signal of both bones is normal. **B** Axial spin density image (SE 2200/20). The lesion is lobulated and inhomogeneous. **C** Axial T2-weighted image (SE 2200/80). The tumor is inhomogenous, with ill-defined margins indicating either tumor infiltration or peritumoral edema

Clinical information

The patient is a 14-year-old white male who presented with an enlarging mass on the volar aspect of his left forearm. Two years earlier he had noted a small mass after a fracture of the mid-shaft of the radius. Four months after the initial injury, he sustained a second fracture in an identical location. Radiographs at the time of the second fracture (Fig. 1) revealed the radius to be bowed at the level of the fracture, and an elongated lucency could be seen scalloping the more distal radial cortex.

At the time of presentation, physical examination was significant only for the large, fusiformmass on the left forearm, which was firm and tender to palpation. Past medical history and review of systems were negative with the exception of the two fractures and subsequent forearm mass. Laboratory examination revealed an elevated alkaline phosphatase level (320 U/l, normal 30–105 U/l), but was otherwise normal. Plain radiographs (Fig. 2) again displayed mild bowing of the radius. The fractures had healed, but the mid-radial shaft now appeared saucerized in association with mature periosteal new bone. Computed tomography (CT; Fig. 3) indicated that the mass was closely associated with the surface of the radius, and vascular structures and tendons were smoothly displaced by the mass. Magnetic resonance imaging (MRI; Fig. 4) revealed the mass to be lobulated with slightly ill-defined margins. The marrow signal intensity of the radius and ulna was normal. The patient was taken to the operating room for open biopsy. Total excision was planned if the lesion proved to be benign.

Correspondence to: L.L. Seeger, Department of Radiological Sciences, UCLA School of Medicine, 200 Medical Plaza, Suite 165-59, Los Angeles, CA 90024-6952, USA

Diagnosis: Juxtacortical aggressive fibromatosis (desmoplastic fibroma) of the forearm

At surgery, the mass appeared to have arisen in the region of the interosseous membrane or the periosteum of the radius. There was saucerization of the radius but no apparent invasion of bone. The mass neither invaded nor was adherent to the soft tissue of the forearm. Rather, the soft tissues were displaced.

Frozen section revealed aggressive fibromatosis (desmoplastic fibroma) without evidence of malignancy. The mass was excised, and surgical margins were judged to be free of tumor.

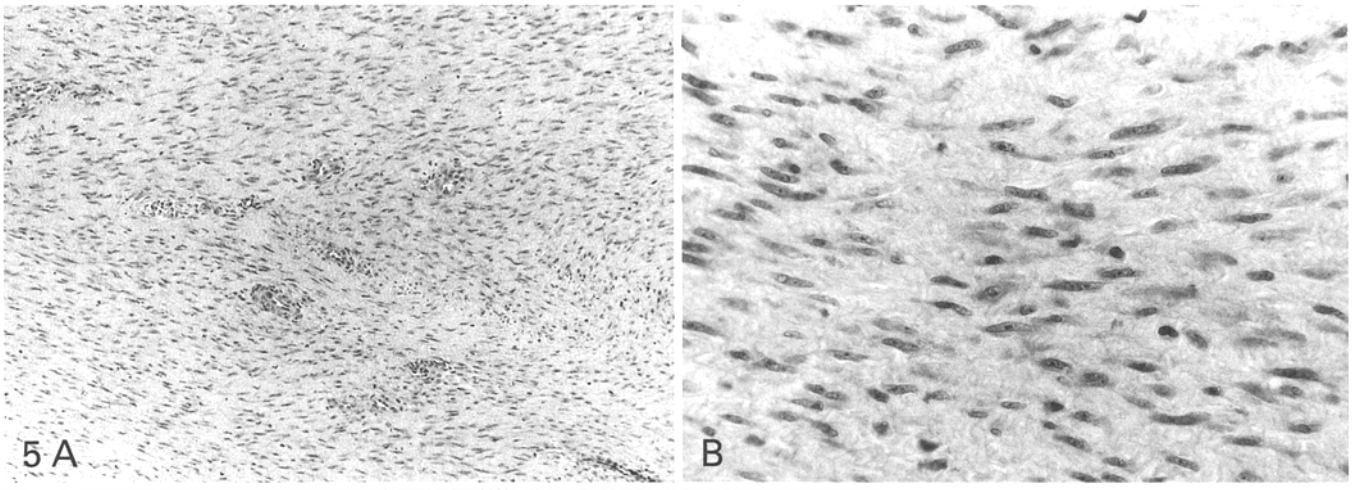
The specimen consisted of the tumor mass, marrow and periosteum of the radius, and overlying fascia and skeletal muscle. A portion of the distal interosseous membrane and bone fragments from radial cortex debridement were also submitted.

Grossly, the tumor consisted of an elongated, firm, light brown mass which was covered by a tan, glistening fascia. On cut section, the tumor was gray-white in color. Histologically, the tumor was fibroblastic and monotonous throughout. The cells were oriented parallel to each other and contained elongated, plump nuclei with fine stippled chromatin (Fig. 5). Typical mitoses were scant, about one per ten high-power fields. No anaplasia was noted. The collagen was finely fibrillar and wavy. There was no osteoid, bone, or cartilage matrix production. The tumor was shown to involve radial periosteum, and fascia and skeletal muscle. The closest margins were at the proximal and lateral (radial) margins, where tumor was present to within 1–2 mm from the inked margin. Marrow space involvement was present on one section of tissue taken from the radial excision.

The patient received no additional postoperative treatment and was followed carefully clinically. Eight months following initial resection, radiographs revealed erosion of the radius and constriction of the ulna, suggestive of recurrence (Fig. 6A). MRI and CT showed an extensive soft tissue mass involving the forearm distal to the previous area of resection, which was associated with invasion of the radius (Fig. 6B,C). The patient was again taken to surgery, and a wide resection was carried out which included excision of the distal ulna and a hemiresection of the distal radius. He is presently undergoing postoperative radiation therapy.

Discussion

Juxtacortical aggressive fibromatosis is an extremely rare low-grade neoplasm which has histologic features indistinguishable from intramedullary and soft tissue desmoid tumors. There have been several reports of "juxtacortical" and "periosteal" desmoids in the radiology literature; however, these reports refer to benign periosteal new bone formation most commonly found along the posteromedial aspect of the distal femoral metaphysis [9, 11, 14, 15]. These are appropriately referred to in the pathology lexicon as "periostitis ossificans" or "cortical irregularity syn-



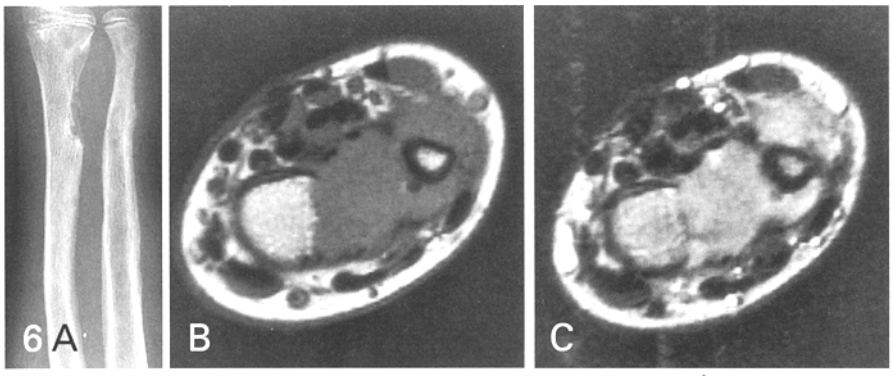


Fig. 5A,B. Spindly fibroblasts with monotonous nuclei associated with wavy, fibrillar collagen production characteristic of aggressive fibromatosis were seen in both the biopsy and the resection specimen. **A** H & E, × 125 **B** H & E, × 400

Fig. 6. A Anteroposterior radiograph 8 months following resection. There is destruction of the medial cortex of the radius and constriction of the distal shaft of the ulna. **B** Axial T1-weighted magnetic resonance image (SE 500/21). **C** Axial T2-weighted magnetic resonance image (SE 2500/80). Soft tissue tumor can be seen invading the radius and surrounding the ulna

drome." Cortical irregularity syndrome is due to avulsion of muscular or ligamentous attachments from bone with subsequent formation of fibro-osseous tissue [2, 13]. This condition is not neoplastic.

Jaffe has referred to interosseous desmoids as "desmoplastic fibroma," and soft tissue pathologists often use interchangeably the terms "aggressive fibromatosis" and "desmoid." In order to avoid confusion in terminology, it is probable best to refer to the juxtacortical neoplastic process presented in this paper as "aggressive fibromatosis" or "desmoplastic fibroma" rather than "desmoid."

There have been few reported cases of aggressive fibromatosis arising in the periosteum. Mirra has found only one [10]. Crim et al. [4] reported an additional periosteal tumor, and felt that in the cases presented by Jaffe [7] and Young et al. [17] independently, reported as intramedullary desmoplastic fibromas, the radiographic appearance was more consistent with a periosteal origin. In the case we report here, it is impossible to determine whether the tumor arose in the region of the interosseous membrane or within the periosteum of the radius.

Aggressive fibromatosis can extend into the soft tissues, and can invade bone. Radiographically, periosteal fibromatosis typically appears as a lytic bulge with focal cortical erosion and saucerization of the underlying bone. Fibromatosis arising in soft tissues immediately adjacent to bone may also cause erosion and saucerization. For both interosseous and periosteal primary desmoid tumor, periosteal new bone is generally associated with a fracture [4].

The CT appearance of fibrious tumors is non-specific [3, 6]. The margins of the lesion may be sharp or ill-defined, and the tumors show variable attenuation relative to surrounding muscle both before and after the administration of intravenous contrast. The CT scan of the lesion presented here demonstrated a well-defined lobulated mass with a saucer-shaped scalloped cortical margin indicating pressure erosion. The tumor itself was inhomogeneous and of lower attenuation than the surrounding muscle.

The reported MRI appearance of tumors of fibrous origin is confusing and often contradictory. Early MRI studies suggested that fibrous tumors possessed a distinctive pattern of low signal intensity on all pulse sequences [1]; however, subsequent studies show them to have a variable MRI appearance [5, 12]. The predominate cellular components of the tumor seem to dictate the MRI appearance [9, 12, 16]. In T2-weighted images, tissues with a high content of fibroblasts and vascular endothelial tissue have high signal intensity, and tissues with abundant collagen show low signal intensity [9]. The lesion in our study demonstrated low to isointensity on the T1-weighted images and progressively increasing signal intensity with T2-weighting. These characteristics are non-specific, and offered no assistance in tissue characterization. There was a rim of low signal intensity surrounded the lesion, suggesting encapsulation, but the margin was not sharp. Histologically, the mass was well circumscribed but unencapsulated.

At surgery, the low-grade nature of juxtacortical aggressive fibromatosis should be recognized and radical resection or amputation should be avoided. A wide surgical margin should be obtained whenever possible. Although benign, this tumor is locally aggressive and when surgical margins are close or positive for the presence of tumor cells, postoperative radiation is probably indicated. Close clinical follow-up should look for local recurrence which should be managed by conservative surgical resection.

In *summary,* a case has been presented of a 14-year-old male patient who developed a fusiform mass on the volar aspect of his left forearm following two fractures. Microscopic features and plain radiography, CT, and MRI appearance of juxtacortical aggressive fibromatosis are discussed.

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