Skeletal Radiol (1988) 17:378–381

Skeletal Radiology

Case report 494

Lawrence Yao, M.D., and Joong K. Lee, M.D.
Department of Radiology, Albany Medical Center Hospital, Albany, New York, USA

Radiological studies

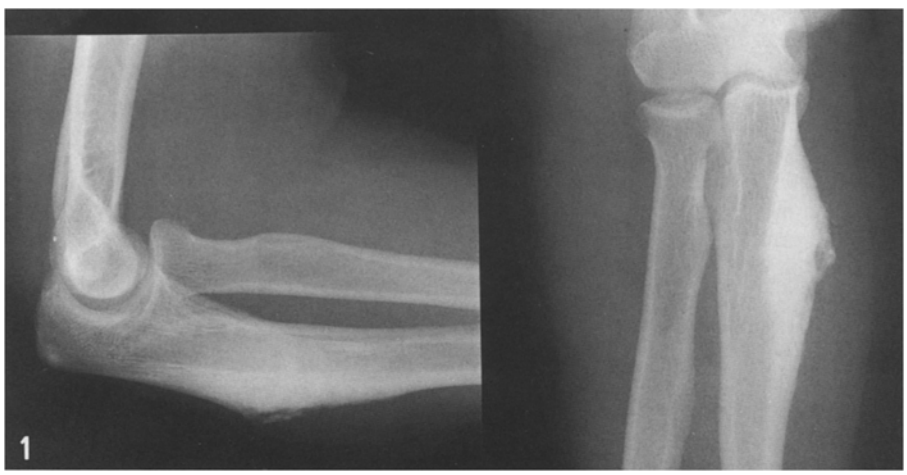

Fig. 1. Roentgenograms of the proximal half of the right forearm prior to surgery show eccentric cortical thickening along the proximal aspect of the ulna, and a peripherally situated radiolucent focus within the region of the sclerosis

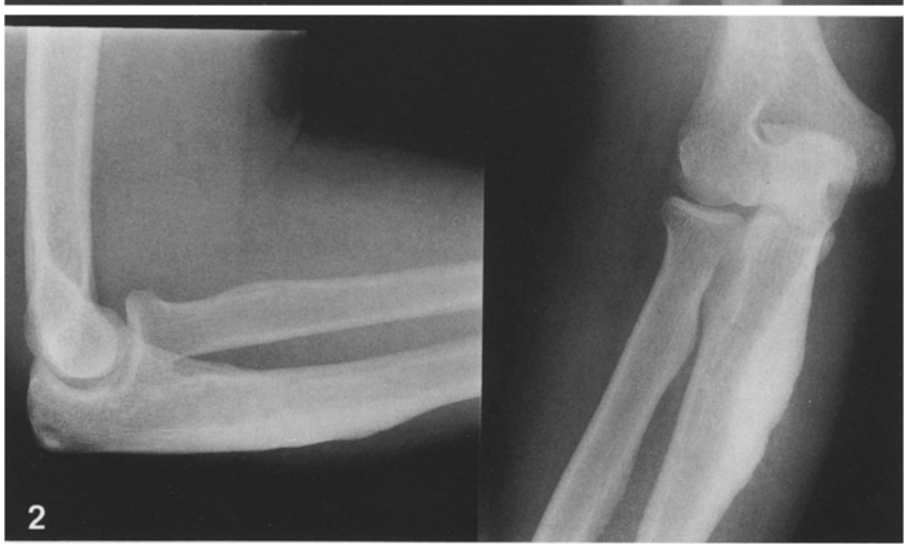

Fig. 2. Follow-up studies one year after surgery again show eccentric cortical thickening, but no evidence of a radiolucent focus

Address reprint requests to: Joong K. Lee, M.D., Department of Radiology, Albany Medical Center Hospital, New Scotland Ave, Albany NY 12208, USA

© 1988 International Skeletal Society

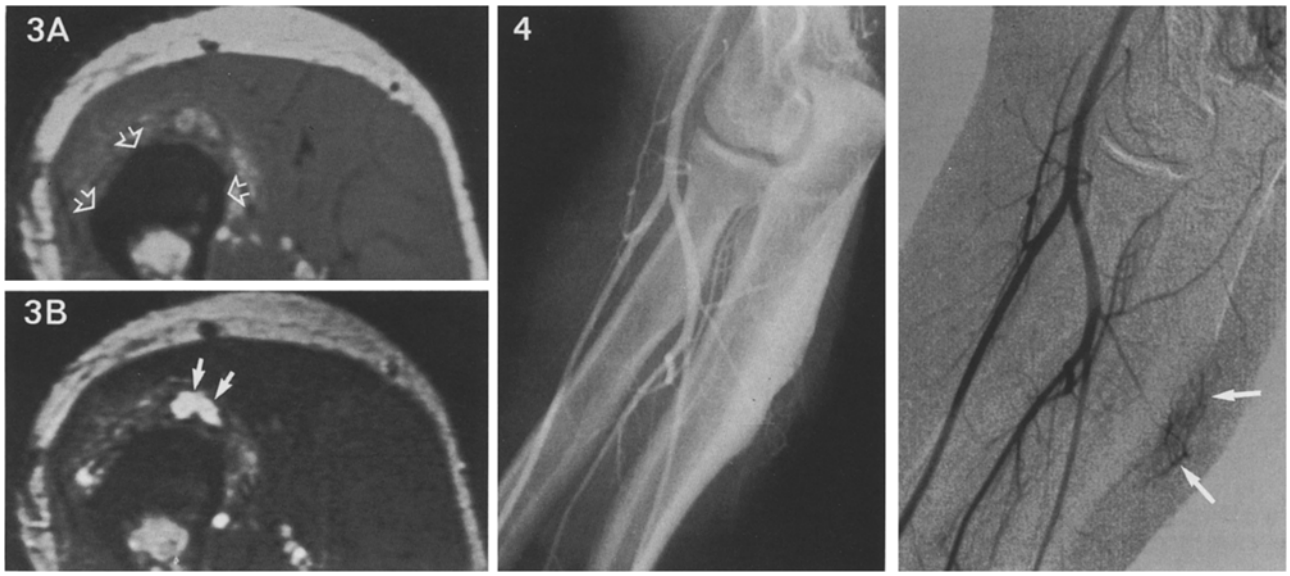

Fig. 3. A An early echo MR image (TR-1500 ms, TE 20 ms) demonstrates eccentric cortical thickening (*open arrows*) and an irregular, juxtacortical area of inhomogeneous signal. **B** A delayed echo MR image (TR-1500 ms, TE 70 ms) demonstrates irregular areas of high signal (*white arrows*) over the posterior aspect of the thickened cortex

Fig. 4. Standard and digitally subtracted images from the arterial phase of an axillary angiogram show a vascular blush, fed by branches of the interosseous and ulnar arteries, in and around the area of cortical thickening

Clinical information

This 20-year-old man presented with a palpable mass in his right forearm, first noted 3 years ago, at which time he refused a diagnostic work-up. A year ago the patient complained of pain in the region of the mass, and an excisional biopsy was performed. The patient recently presented with recurrent and increasing pain in the forearm, unrelieved by aspirin.

On physical examination, a hard, tender, nonpulsatile mass was noted, extending along the postero-medial aspect of the upper segment of the right forearm for a length of approximately 8 cm, without associated warmth or erythema. Strength and sensation were normal. Routine hemogram, erythrocyte sedimentation rate, electrolytes, BUN, and creatinine were normal. Serum calcium, phosphorus, and alkaline phosphatase also were normal. The creatine-phosphokinase was elevated at 485 IU/1 (normal <140).

The radiographs of the right forearm prior to surgery 1 year earlier showed an area of marked cortical thickening along the postero-medial surface of the upper portion of the right ulna and a well-defined radiolucent area within the region of sclerosis (Fig. 1). The radiolucent focus was no longer evident on the post-operative study (Fig. 2).

Swelling was noted in the overlying soft tissues, without evidence of dystrophic calcification or discrete mass.

MR and angiography were performed upon recurrence of symptoms after initial excision of the tumor growth.

On the transaxial MR images (TR 1500 ms, TE 20 and 70 ms), cortical thickening was again evident over an area approximatedly 10 cm in length (Fig. 3A, b). On the more distal delayed echo images, an irregular area of high signal was noted over the posterior surface of the ulnar cortex.

On the right axillary arteriogram, an arterial blush was observed in the soft tissues over the area of bony change, fed by the interosseous artery and branches of the ulnar artery (Fig. 4). A persistent stain was noted, without evidence of an early draining vein.

Diagnosis: Hemangioma of surface of ulna with prominent sclerosis

The operative report from the initial excision a year earlier described curetting of a lesion which was confined by and was deep to the periosteum. The periosteum had been reflected and preserved. Histological examination of the specimen showed numerous vessels of various sizes, some cavernous, intervening between areas of cortical bone (Fig. 5).

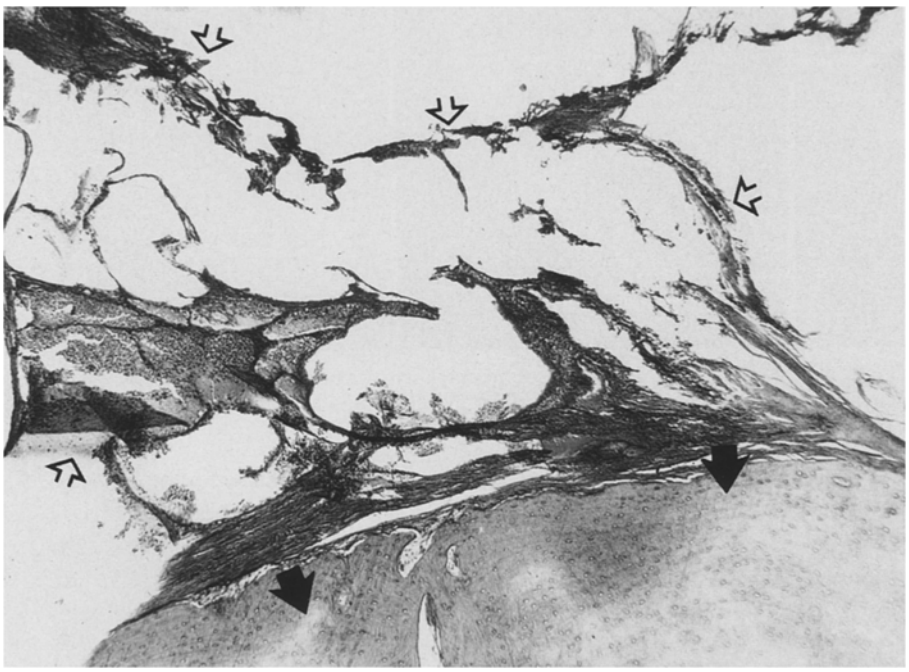

Fig. 5. Histological section after osteotomy (original magnification 4×) shows blood vessels of various sizes (*open arrows*), some cavernous, intervening between areas of thickened cortical bone (*black arrows*)

Discussion

On the basis of the plain radiographic findings and the history of pain, the diagnosis of osteoid osteoma was suggested. Osteoid osteomas are known to recur if incompletely removed. The radiographic findings would also be consistent with a chronic or sclerosing osteomyelitis, but the laboratory findings and clinical history were not supportive of an infectious etiology. Similar cortical changes in a young patient might be seen with periosteal lesions such as lipoma, fibroma, or chondroma, although recurrence of these lesions would be unexpected after resection.

MR confirmed the presence of cortical thickening and an associated juxtacortical mass, that showed areas of bright signal on T2-weighted images, possibly related to intravascular blood, hemorrhage, or edema. The elevated CK suggested the presence of adjacent necrosis of muscle or inflammation. The angiographic study confirmed the presence of a hypervascular, extraosseous component of the tumor, consistent with a hemangioma of the surface of the bone with a soft tissue component. The histological diagnosis of hemangioma would be unexpected without prior knowledge of the angiographic findings. Furthermore, without reference to the original presentation, it would be unclear whether the primary lesion was an intracortical or periosteal hemangioma. As an additional consideration, primary soft tissue hemangiomas may also produce bony changes.

It is important to note that hemangiomas of the appendicular skeleton are rare; 75% of hemangiomas of bone occur in the skull or spine [8], and of the remainder, the vast majority occur in flat bones. A hemangioma in a long bone may lack the characteristic appearance of a hemangioma found in the skull, spine, or flat bones and may thus present diagnostic difficulty, but will usually show lytic or expansile characteristics [1]. Intracortical or periosteal hemangiomas of long bones are said to be exceedingly rare; they present with the atypical radiographic finding of cortical thickening. The true incidence of these lesions may be greatly underestimated. Many cases may either be

entirely asymptomatic and go undetected, while others that are incidentally encountered may be dismissed as unproven cases of benign periosteal growths.

Two intracortical hemangiomas in long bones similar in radiographic appearance to osteoid osteoma, and to the lesion in this case, have been reported [3, 7]. The lesions showed marked cortical thickening and a well-defined, radiolucent, intracortical area similar to the initial radiographs of the case presented here. In the present case, the radiolucent focus was no longer evident upon second presentation, presumably due to surgical intervention which also may have contributed to extracortical extensions of the tumor.

Periosteal hemangiomas have been reported in the tibia, femur, and ulna [2,5]. All cases exhibited changes of cortical thickening and local sclerosis without a radiolucent focus. The osteolytic characteristics typical of hemangioma of long bones also were absent. Presumably, the cortical response is secondary either to stretching or irritation of the periosteum, or to local hyperemia caused by the hypervascular lesion.

The angiographic findings in this case, considered alone, might suggest the presence of a primary hemangioma of soft tissues, since such hemangiomas may produce changes in adjacent bone. In such cases of bone involvement, the radiographic features are reported to be manifested by: (1) signs of external pressure erosion; and (2) coarsening of trabeculae with widening of the trabecular interspaces [8]. However, in the reported cases of hemangiomas of soft tissues with bony changes soft tissue calcifications or phleboliths are said to be characteristic, and the lesions typically have been multifocal [4].

The recurrence of symptoms and growth of the lesion in this case is not surprising. In general, the complete surgical extirpation of congenital arteriovenous malformations of extremities is felt to be impossible [6]. In the case of intracortical or surface hemangiomas, complete excision of all reactive sclerotic bone has been suggested for cure [3]. Experience with these rare lesions is too limited to comment conclusively on prognosis or optimum management. Radiotherapy, believed to be effective in the management of surgically inaccessible hemangiomas of the spine, may also serve an adjunctive role in the treatment of such cases of symptomatic (surface) or intracortical hemangiomas of long bones.

In *summary*, the case is presented of a 20-year-old man with symptoms referable to the right forearm near the elbow for 3 years. The patient refused a diagnostic work-up until 1 year prior to presentation when he complained of more severe pain and an excisional biopsy was performed. The patient presented himself at this time with recurrent and increasing pain in the forearm, unrelieved by aspirin.

Physical examination demonstrated a hard, tender, non-pulsatile mass just below the elbow over the area of the ulna. Laboratory studies were normal except for elevation of the creatinine-phosphokinase. Imaging studies, including MR, were obtained. Angiography also was performed. The operative report from the initial excision a year earlier described curetting of the lesion confined by and deep to the periosteum which had been reflected and preserved. Histological studies of the specimen showed numerous blood vessels of various sizes, indicating the diagnosis of hemangioma.

The issue of the site of origin of the hemangioma was considered and it was finally decided that the most appropriate statement in terms of the diagnosis should be hemangioma of the surface of the ulna with prominent sclerosis. The major criteria available indicating that the lesion was intracortical at its outset, were the original imaging studies. A photomicrograph, on the other hand, as well as the angiogram, suggested that the hemangioma was on the outer surface of the ulna, with a soft tissue component. The different types of hemangiomas with regard to location were considered, with particular emphasis placed on intra cortical or periosteal hemangioma of long bones as well as hemangioma of soft parts invading bone. Treatment also was discussed briefly.

References

1. Wilner D (1982) Radiology of bone tumors and allied disorders. WB Saunders, Philadelphia
2. Dorfman HD, Steiner GC, Jaffe HL (1971) Vascular tumors of bone. Human Pathol 2:349
3. Schajowicz F, Rebecchini AC, Bosch-Mayol G (1979) Intracortical haemangioma simulating osteoid osteoma. J Bone and Joint Surg [Am] 61:94
4. Willinsky RA, Rubenstein JD, Cruickshank B (1982) Case Report 216 (Intracortical hemangioma of tibia). Skeletal Radiol 9:137
5. Loxley SS, Thiemeyer JS, Ellsasser JC (1972) Periosteal Hemangioma; a report of two cases. Clin Orthop 85:151–154
6. Sugiura I (1975) Tibial periosteal hemangioma. Clin Orthop 106:242
7. Sherman RS, Wilner D (1961) The roentgen diagnosis of hemangioma of bone. AJR 86:1146–1159
8. Szilagyi DE, Smith RF, Elliott JP, Hageman JH (1979) Congenital arteriovenous anomalies of the limbs. Arch Surg 111:423