Skeletal Angiomatosis Limited to the Hand: Radiographic and Scintigraphic Correlation

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Three-phase bone scintigraphy elegantly demonstrates the vascular nature of the expansile, lytic lesions of skeletal angiomatosis and the induced bony changes in the involved areas. Unusual features of the case include its confinement to the hand and rapid progression as shown by serial radiographs.

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Of unknown etiology, angiomatosis is likely part of a spectrum of primary cystic disease arising from uncontrolled angiomatous and/or lymphangiomatous proliferation in multiple bones, viscera and/or soft tissues (*1-5*). The current case of skeletal angiomatosis presents two unique features: (1) involvement limited to a single distal extremity and (2) an aggressive nature as shown by rapid progression on serial radiographs. The three-phase bone scintigraphic findings exquisitely reflect the underlying histopathology and correlate with radiographic distribution of lesions.

CASE REPORT

A previously healthy and otherwise asymptomatic 15-yr-old male was evaluated for a painful left wrist following minimal trauma. Routine laboratory studies were normal. Initial radiographs revealed expansile lytic lesions involving the distal radial metaphysis and epiphysis with sparing of the growth plate, the scaphoid, trapezium, capitate, hamate, second metacarpal and fifth metacarpal without definite evidence of fracture (Fig. 1A). A follow-up radiograph 13 mo later (Fig. 1B) showed marked progression and expansion of the previously noted lesions and new involvement of the third and fourth metacarpals, the trapezoid and thumb phalanges.

Three-phase bone scintigraphy of the hands and whole-body delayed imaging using ^{99m}Tc-oxidronate were performed several weeks prior to the more recent, follow-up plain film. Blood flow (Fig. 2A) was dramatically increased to those lesions seen on the corresponding plain film (Fig. 1B). Well-circumscribed foci of intense "blood-pool" activity were present in a similar distribution (Fig. 2B). A striking discordance of tracer activity was present between the "blood-pool" and delayed hand images. The areas of early increased vascularity were relatively photopenic and surrounded by rims of slightly increased activity on the delayed image, with the most pronounced findings in the metacarpals (Fig. 2C). The remainder of the skeleton was normal.

An open biopsy of the radius revealed endothelial-lined vascular channels filled with blood, without lymphatic vessels, consistent with angiomatous proliferation. Clinically, the patient has experienced progressive pain, deformity and marked loss of function of the left hand.

DISCUSSION

Skeletal angiomatosis is a disease characterized by multiple cystic lesions of blood vessel, lymphatic or mixed origin, confined to bone. This particular entity is thought to fall within a spectrum of disease due to a basic angiomatous/lymphangiomatous proliferative process (*1,2*). Clinical conditions range from a mild form localized to bone or soft tissue to diffuse skeletal angiomatosis to the most severe form with widespread involvement of bone, viscera and soft tissues. The etiology is unknown but may represent a developmental abnormality, a so-called vascular hematoma, based on the similar histologic characteristics of all the various lesions, or a true neoplasm as reflected by its disseminated appearance (*1,2*).

The blood flow and blood-pool portions of the three-phase bone scan demonstrate the striking vascular nature of the skeletal lesions. The delayed image, which shows markedly decreased activity in several of these lesions surrounded by rims of increased activity, confirms absence of normal bony architecture. The rims of increased activity likely reflect ongoing bony metabolism in response to continued cortical expansion.

Despite different underlying pathologies, a similar scintigraphic pattern of disparate increased early and decreased delayed uptake surrounded by a rim of increased activity, likened to a "doughnut" or "ring", have been described for other bone lesions such as giant-cell tumor (*7,8*) and metastatic disease (*9*). Based on the above examples, this disparate scintigraphic pattern might be found at a site of skeletal disease possessing both vascular proliferation and loss of normal bone matrix, the combination of which stimulates continued cortical bone remodeling. Whole-body imaging is a useful means of identifying other sites of skeletal involvement.

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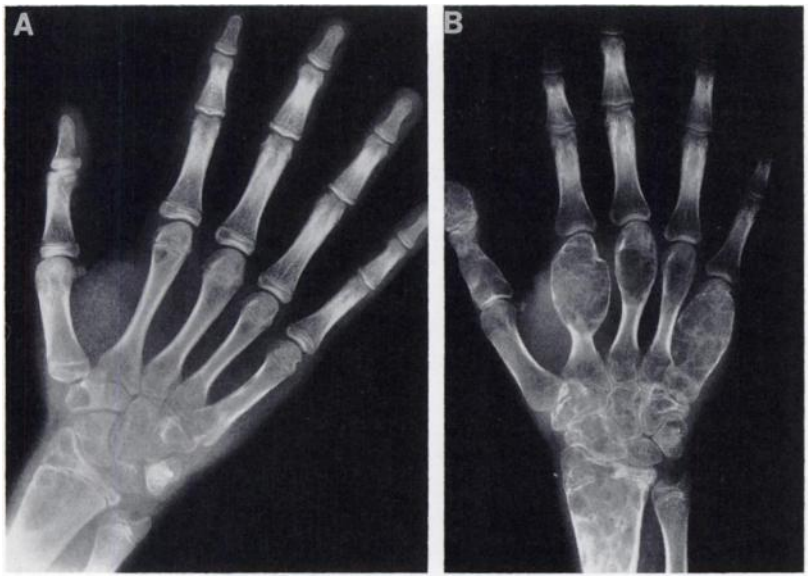

FIGURE 1. Radiographs of the left hand at presentation (A) and 13 mo later (B) demonstrating dramatic progression of expansile, lytic lesions.

The current case of skeletal angiomatosis is interesting for its confinement to a single distal extremity and its dramatic progression over a relatively short, 13-mo, period of time. These lesions usually exhibit slow growth. Such rapid progression suggests that angiomatous proliferation may have begun only recently. Vascular hamartomas have exhibited the phenomenon of delayed appearance (*1*). Together, these factors imply that the etiology in this patient was likely a delayed manifestation of a primary endothelial developmental defect confined to a single limb bud. To our knowledge, only one other case of skeletal angiomatosis in the hand has been reported (*5*).

Another entity which is probably part of the spectrum of angiomatous/lymphangiomatous proliferation is cystic angiomatosis, a disease with multiple cystic lesions scattered throughout the skeleton (*6*). Such skeletal lesions, which tend to be distributed more centrally than peripherally, are frequently associated with similar angiomatous changes in viscera (notably, the spleen) and soft tissues. Lesions limited to bone, as in the current case, are rare; only 9 of 26 patients in a review by Wallis et al. (*4*) were found to be free of visceral involvement. Whatever the name or location, the basic histology common to all lesions within this disease spectrum appears to be an endothelial-lined cyst representing dilated vascular channels of angiomatous or lymphatic origin as indicated by their contents (*1,2*)

Historically, angiomatosis has been refractory to chemotherapy or radiation. Isolated lesions may be amenable to surgery (*1,2*). Angiomatosis has shown no tendency toward malignant degeneration and the major complication of skeletal lesions is pathologic fracture with pain. At present, only the bones in the left hand of this patient appear involved. There is no clinical or biochemical evidence of skin or visceral involvement. Prognosis for isolated skeletal disease generally is good, with only local morbidity. However, half of the patients with cystic angiomatosis present around the age of puberty (*1*) and delayed visceral involvement, which would carry a much graver prognosis, is still a possibility in this patient. Continued skeletal, visceral and soft-tissue surveillance will be required.

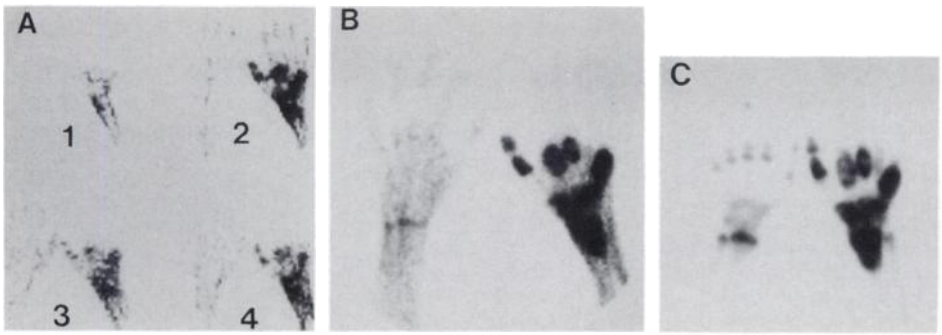

FIGURE 2. Summed 5 sec/image (2 frames at 2.5 sec/frame) palmar blood flow (A) and static "blood-pool" (B) images demonstrate the marked vascularity of the lesions in the left hand. Three-hour delayed palmar image (C) shows several prominent central photopenic areas consistent with angiomatous proliferation surrounded by rims of increased activity reflecting increased cortical metabolism. Compare these sites with the spared areas in the affected wrist and hand and normal right hand.

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SELF-STUDY TEST
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ANSWERS

ITEMS 1–5: Prostatic Carcinoma

ANSWERS: 1, F; 2, F; 3, F; 4, F; 5, T

Patients with prostatic carcinoma metastatic to bone may present with bone pain; however, in at least one series, 43% of patients presented with no pain. Patients may complain of mild arthralgias or other discomforts without accurate localizing signs. The absence of bone pain does not exclude metastatic disease nor does elevation of serum acid phosphatase necessarily imply metastatic disease to bone. Patients with elevated serum acid phosphatase levels and normal bone scintigrams were shown by Pollen et al. to have developed no evidence of bony metastases on mean follow-up of 17 mo. The authors concluded that the elevated serum acid phosphatase levels were caused by extension of the carcinoma through the prostatic capsule in these patients. Patients in whom serial scintigrams show progression of metastatic disease have a significantly shorter mean survival time than those who show improvement or stable scintigrams. Levenson et al. found that radiography and scintigraphy were equally sensitive for documenting progression of disease, but the radiographic abnormalities rarely resolved when the metastatic disease improved or healed. Scintigraphy provided evidence of progression of disease in advance of detectable changes in serum acid phosphatase levels in 74% of patients in one series.

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ITEMS 6–10: Osteosarcoma

ANSWERS: 6, T; 7, F; 8, F; 9, T; 10, F

The most common malignant primary bone tumor is multiple myeloma. Osteosarcoma is the second most common malignant primary bone tumor. Although osteosarcoma can occur anytime between the first decade and the eighth decade of life, the peak incidence is in the second decade.

When sarcomas occur in bones that have been subjected to irradiation, the average latency period is 15 yr, with a range of 2.8–55.0 yr. It is less than 5 yr in only 8% of patients and is more than 20 yr in 30% of patients. Although osteosarcomas do occur with increased frequency in foci of Paget's disease and at sites exposed to radiation therapy, only 19% of the "older" patients with osteosarcoma had preexisting conditions in Dahlin's series.

In patients with osteosarcoma, pulmonary metastases predominate; however, bone metastases can occur alone or can develop before pulmonary metastases occur. Recent studies suggest that the pattern of metastatic disease in patients who are not cured by primary therapy is changing, with a greater frequency of extrapulmonary disease.

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ITEMS 11–15: Ewing's Sarcoma

ANSWERS: 11, F; 12, T; 13, T; 14, F; 15, F

Ewing's sarcoma, on average, occurs in a younger age population than any other primary malignant tumor of bone. Although Ewing's tumors have been seen in patients in the seventh decade of life, the majority of cases occur before the end of the second decade. The most common sites are the long bones of the extremities (femur 22%; tibia 11%; humerus 10%; fibula 9%); however, all bones of the body can be involved with Ewing's sarcoma. The lower extremities and pelvic girdle accounted for approximately 60% of cases in Dahlin's series. Ribs are involved in about 8% of cases.

The most common presenting symptoms of Ewing's tumor are pain and swelling, with pain as the primary symptom in more than 50% of the patients. Swelling in the region of the tumor is common by the time the patients come to medical attention, although swelling alone is rarely a first symptom.

The typical roentgenographic appearance of Ewing's sarcoma is that of a lesion involving a long bone characterized by permeative lytic destruction and periosteal elevation with an "onionskin" appearance. However, these features can be seen in a variety of other primary osseous malignancies, including malignant lymphoma, eosinophilic granuloma, and osteosarcoma, and in acute and chronic osteomyelitis. Pathologic evaluation is always required to establish the diagnosis.

In a recent article, unsuspected sites of skeletal metastasis were demonstrated on bone scintigraphy in approximately 39% of patients. In 53 patients presenting with Ewing's sarcoma, 25 had metastatic disease, and in 20 this was not suspected clinically. Other series have reported a frequency of osseous metastatic disease varying from 11% to 21% at the time of initial presentation.

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ITEMS 16–21: Paget's Disease

ANSWERS: 16, T; 17, F; 18, T; 19, F; 20, T; 21, F

The incidence and prevalence of Paget's disease throughout the world is quite variable. It is common in the United States, Central Europe, England, Australia, and New Zealand, but rare in the Middle East, Asia, and Africa. Although this may be partially racial in origin, the prevalence in the black population in United States cities seems to be similar to that in the white population.

The lumbar spine and pelvis are involved in a majority of patients, followed by the femur and skull. Tibial involvement is less common but often shows the characteristic "blade of grass" radiographic appearance. All

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