Recurrent solitary sarcoidosis in bone—a case report

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In 1991, a 34-year-old Japanese woman presented with gradually increasing pain and swelling of her right middle finger over a 4-month period. Radiographics revealed an osteolytic lesion in the proximal end of the middle phalanx (Figure 1). Routine hematological and serum biochemical examinations were normal, and the patient's past medical history was unremarkable. An excisional biopsy and curettage were performed, and showed numerous non-caseating granulomas, typical of sarcoidosis. A retrospective examination of her chest radiograph, a routine procedure before a surgical intervention, showed no evidence of pulmonary involvement by sarcoidosis. However, on careful examination with computed tomography (CT), we found slight bilateral hilar lymph node enlargement suggestive of lung sarcoidosis (Figure 2).

Because of the subsequent disappearance of these CT findings, she was not given systemic corticosteroid treatment. After relief of pain for several months, the digital pain and swelling recurred, and became very severe about 1 year after the first operation. A radiograph showed that the osteolytic lesion now involved the entire middle phalanx. Bone scintigraphy and MRI could not distinguish between recurrence of the sarcoidosis lesion and an infectious reaction (Figure 3). At operation, we found that a well-demarcated nodular mass filled the cortical bone defect which had been made by the previous curettage (Figure 4). Intraoperative histopathological examination showed a typical noncaseating granuloma, compatible with recurrent sarcoidosis. Cultures for bacteria, fungi and tubeculosis yielded no growth. After intramedullary curettage, an autogenous iliac bone graft was fashioned to prevent angulation of the phalanx, because the affected cortical bone had become very thin in some areas.

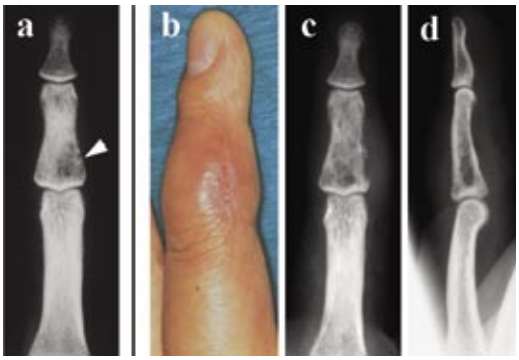

Figure 1. Osteolytic lesion and swelling of the middle finger before and after the first operation.
a. Poorly demarcated reticular lesion before the first operation (arrowhead).
b. Swelling and pain recurred 6 months after the first operation.
c, d. Reticular radiolucent lesion has spread throughout the whole phalanx.

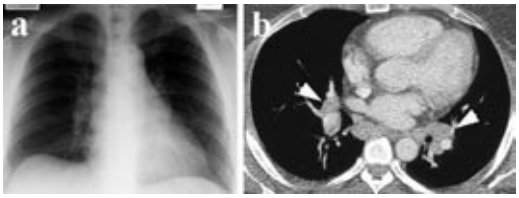

Figure 2. Minute pulmonary manifestation.
a. Routine chest radiograph does not show typical hilar lymph adenopathy, although slight enlargement may be present.
b. Chest CT shows bilateral hilar lymph adenopathy (arrowheads).

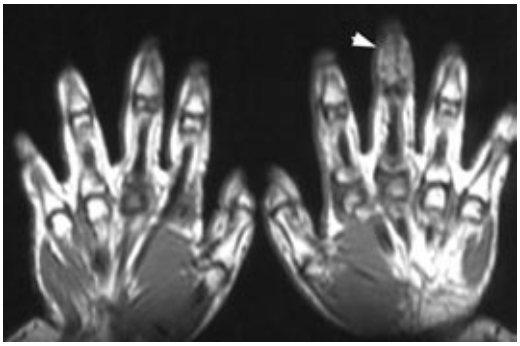

Figure 3. MRI shows a mass of intermediate intensity filling the marrow of the middle phalanx (arrowhead).

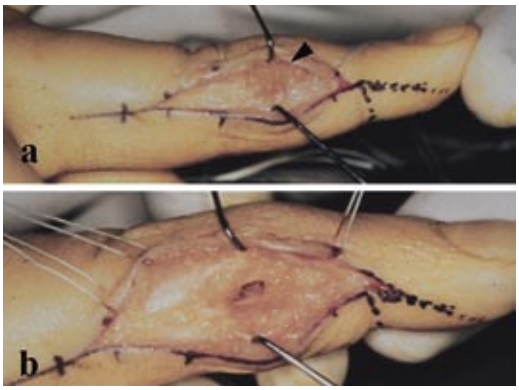

Figure 4.
a. Well-demarcated soft tissue mass can be seen immediately beneath the skin (arrowhead).
b. Removal of the sarcoid mass shows that the sarcoid lesion has completely filled the marrow of the middle phalanx.

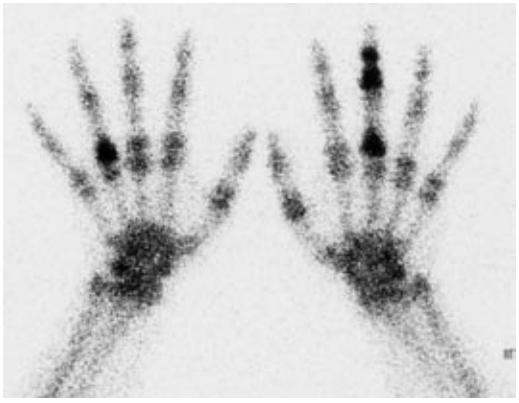

Figure 5. Bone scintigraphy showing the persistent hot spot in the middle phalanx of the right hand and a few new hot spots in both hands.

Her pain subsided at first, but about 6 months after the second curettage, she again complained of pain and swelling. The second bone scintigram performed 1 year after the second curettage showed that she still had a hot spot at the same site as in the previous study, and had now developed a few other sites, which suggested spread of the sarcoidosis reaction (Figure 5). A radiograph showed apparent resorption of the grafted bone and enlargement of the osteolytic lesion. Although these findings strongly suggested a third recurrence of the sarcoidosis lesion, the patient's pain gradually subsided after its peak in 1994. Spontaneous pain relief occurred in 1997, and radiographic bone remodeling was complete in 1998 (Figure 6).

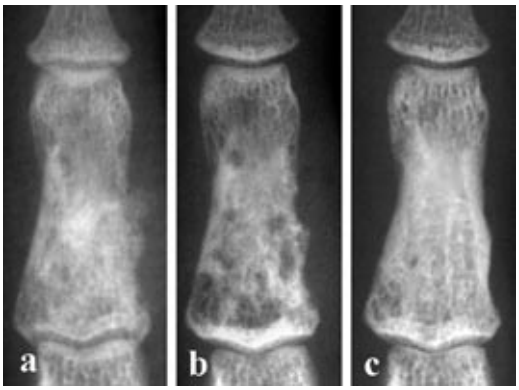

Figure 6. Serial radiographs showing recurrence and healing of bone lesion.
a. Radiograph taken immediately after the second operation. After thorough excision of the intramedullary sarcoid lesion, iliac bone is grafted into the defect.
b. Apparent resorption of the grafted bone and new small punched-out lesions can be seen.
c. After digital pain had been particularly severe in 1994, the radiographs of the hand showed gradual bone consolidation. Bone remodeling was almost complete in 1998.

No sign of a recurrence could be detected in 2002. During the whole follow-up period of 11 years, the patient had no symptoms in other organs.

Discussion

Sarcoidosis is a systemic disease of unknown etiology, characteristically involving the lung with granulomatous lesions. Involvement of the skeletal system is far less common than that of the lungs. Although radiographic evidence of bone involvement has been reported in as many as one tenth of all cases at some time during their clinical course (Greenfield 1975), a worldwide survey of 3,676 patients with sarcoidosis showed osseous lesions in only 109 (3%) (James et al. 1976). Bone involvement normally occurs in patients with generalized and advanced disease, and has been thought to indicate a chronic, persistent, and irreversible systemic disease. The reported mortality of 21%, 4 times more than that seen in other patients with sarcoidosis during the same follow-up period, attests to the serious nature of advanced sarcoidosis involving the skeletal system (James et al. 1976).

Three types of osseous sarcoidosis can be distinguished on radiographs. Minute osteolysis, which

has the appearance of small punched-out defects, with diameters less than 5 mm, is seen in 83%, and comprises the commonest type. The second commonest type is permeative or reticular changes in cortical bone, and is found in 33%. The least common type, a destructive process with larger osteolytic lesions, causes permanent destruction of the normal structure, and is seen in only 10%. The osseous involvement in sarcoidosis is usually painless, since most of the cases are minute or permeative types of lesions (Greenfield 1975, Neville et al. 1976).

Because of their insidious nature, many cases of osseous involvement are found incidentally at multifocal sites during follow-up of advanced sarcoidosis (Greenfield 1975, James et al. 1976, Neville et al. 1976). Unlike painless radiographic osteolysis, patients with destructive osseous lesions have symptoms such as pain and deformities (Pierson and Willett 1978, Adelaar 1983, Landi et al. 1983, Posner et al. 1991, Gonzales del Pino et al. 1997, Kwon et al. 1997). These destructive processes often simulate manifestations of a bone tumor, and excisional biopsy is done despite a previously established diagnosis of sarcoidosis in other systems. In all of the reported cases, histological examination of the noncaseating granuloma in an osteolytic lesion leads to the realization that the bone lesion is due to the systemic disease (Adelaar 1983, Landi et al. 1983, Posner et al. 1991, Gonzales del Pino at al. 1997, Kwon et al. 1997).

Osseous sarcoidosis as the initial and only involvement, like our case is exceedingly rare and only a single case has been reported by Pierson and Willett (1978). Because of lack of a preceding diagnosis in other organs, the diagnosis in the present case was bsed on the histological finding of a noncaseating granuloma. The transient pulmonary adenopathy, shown on the CT scan, supported the diagnosis. However, since spontaneous healing has been reported to be characteristic of osseous sarcoidosis, we first thought that the reucurrence of osteolysis after the first excision was due to an infection. However, the absence of infectious agents and the demonstration of a noncaseating granuloma on the second excision confirmed that the recurrent osteolysis had also been caused by sarcoidosis. Symptomatic osseous lesions treated by surgery, although rare, have been reported to respond well to curettage (Pierson and Willett 1978, Adelaar 1983, Landi et al. 1983, Posner et al. 1991, Gonzales del Pino at al. 1997, Kwon et al. 1997). Thus, our case is unusual even in terms of persistence of disease, which was not eliminated by the repeated curettage.

The periodic aggressiveness of osseous sarcoidosis is shown by the resorption of grafted bone in our case. Landi and his colleagues (1983) have reported a patient who had to undergo an amputation because of severely compromised finger function. Posner et al.'s (1991) case had amalangulated phalanx due to severe osteolysis. However, it should be emphasized that osseous lesions are self-limiting, as in our case. A careful follow-up is recommended for the rare aggressive subtype of osseous sarcoidosis so that surgical treatment can be done at an appropriate time so as to prevent damage due to osteolysis.

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