

Multicentric Epithelioid Hemangioendothelioma of the Bone: Histologic and Radiographic Features

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A multicentric epithelioid hemangioendothelioma (EHE) of the bone is reported which affected the right femur, the right patella and the right fibula of a 56-year-old man. Plain radiographs demonstrated purely lytic multicentric lesions with well-defined sclerotic borders. Computed tomography (CT) scans showed multiple osteolytic lesions in a honeycomb pattern at the epiphyseal lesion of the right femur; the other lesions of the right femoral shaft, the right patella and the right fibula were punched out sharply. Magnetic resonance (MR) images showed that the tumors contained mixed signal intensities: low intensity for T1-weighted images and high intensity for T2-weighted images. Histologically, the tumor was discerned as EHE with a spectrum of endothelial tumor, with the added feature of epithelioid hemangioma in a limited area. As for the initial treatment, curettage and bone cementing were performed for the epiphyseal tumor of the right femur, followed by surgical resections of the other tumors. Neither recurrence nor metastasis was observed 3 years after resection.

Key words: bone tumor; epithelioid hemangioendothelioma; histology; multiple; radiography

Weiss and Enzinger (1982) first described epithelioid hemangioendothelioma (EHE) as a unique tumor of the soft tissue, and proposed it as a counterpart to the vascular tumor associated with epithelioid or histiocytoid cells: the tumors are well known as lesions in the liver, the lung and other organs. EHE of the bone, however, rarely occurs and reported cases have totaled up to less than 80 (Hartmen and Stewart, 1962; Otis et al., 1968; Maruyama et al., 1985; Mirra and Kameda, 1985; Tsuneyoshi et al., 1986; Lyon et al., 1992; Abati et al., 1994; Bollinger et al., 1994; Boutin et al., 1996), including 29 cases reported by Tsuneyoshi et al. (1986) and a series of 40 cases by Kleer et al. (1996).

EHE appears in a unilocular or multicentric manner histologically with features of large,

polygonal cells with intracytoplasmic vacuolization, fibromyxoid stroma and well-formed vascular channels at varying degrees (Tsuneyoshi et al., 1986; Weiss et al., 1986). EHE of the soft tissue takes various clinical courses from benign to malignant; the malignancy is defined as intermediate by WHO classification (World Health Organization, 1993). EHE should be distinguished from benign vascular tumors though difficulty exists in differentiation among epithelioid tumors (Keel and Rosenberg, 1999). Local recurrence and metastasis were reported to be 13% and 31% in incidence, respectively (Weiss et al., 1986).

We here report a case of multicentric bone tumor that fits the category of EHE, with special reference to radiographic features.

Abbreviations: CT, computed tomography; EHE, epithelioid hemangioendothelioma; MR, magnetic resonance

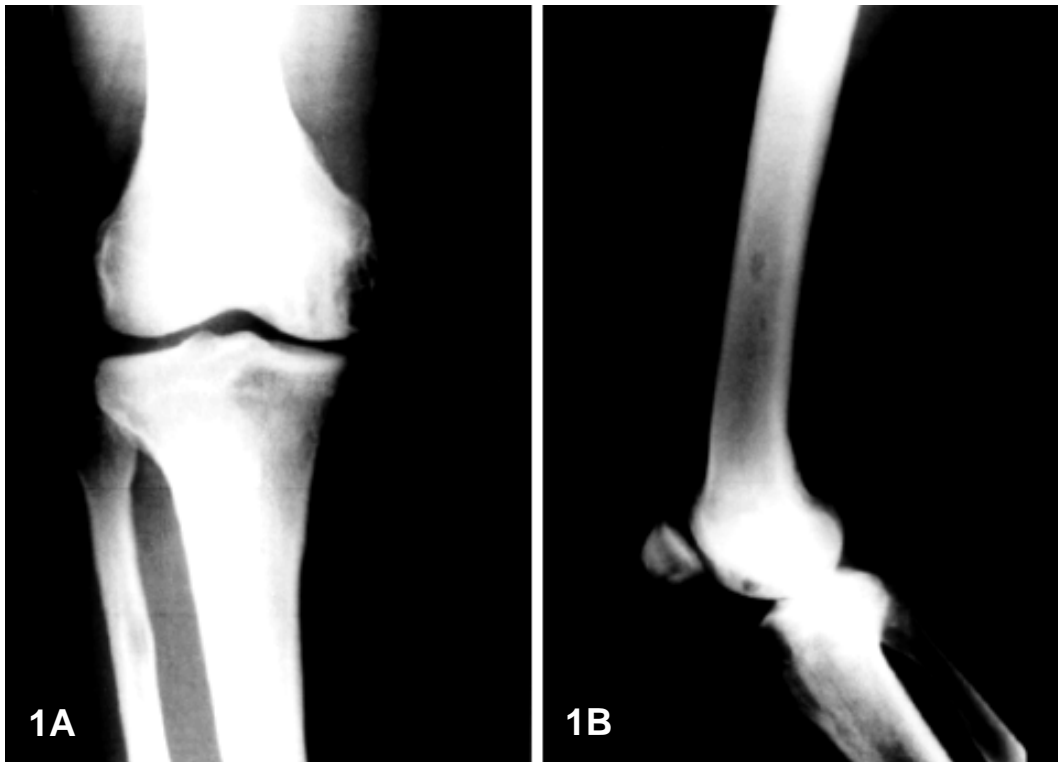


Fig. 1. Plain radiographs. **A:** Lesions in a honeycomb pattern in the epiphysis of the right femur and in the right fibula. **B:** In the lateral view, lesions of the right femoral shaft, the right epiphysis and the right patella are punched out.

Patient Report

A 56-year-old man visited Tottori Red Cross Hospital with a 1-month history of a spontaneous pain in his right knee joint in December 1996. No history of trauma to the thigh was noted. Physical examination on admission revealed tenderness to palpation without any mass. In plain radiographs, anteroposterior and lateral views demonstrated multiple radiolucent lesions in a honeycomb pattern in the distal epiphysis of the right femur, the right fibula and the right femoral shaft (Figs. 1A and B). Computed tomography (CT) scans showed an osteolytic area with marginal sclerosis in the epiphyseal lesion of the right femur; the other lesions of the right femoral shaft, the right patella and the right fibula were all sharply punched out (Figs. 2A and B). The tumors contained mixed

signal intensities in magnetic resonance (MR) images: the intensity was low in T1-weighted images, and high in T2-weighted images (Figs. 3A and B). Technetium bone scan revealed increased uptakes of isotopes at the lesions of the right femur, the right patella and the right fibula (Fig. 4). Histologic examination was carried out for biopsy specimens tentatively suspected of having an EHE. For the initial treatment, curettage and bone cementing were employed for the epiphyseal tumor of the right femur, and then the other lesions of the patella and the fibula were surgically resected.

The surgically obtained specimens were examined and histologically defined as multicentric bone tumor which satisfies the category of EHE. The tumors forming nests in the growth pattern consisted of accumulated epithelioid cells, with various-sized vasculatures of the capillary type (Fig. 5). Epithelioid cells

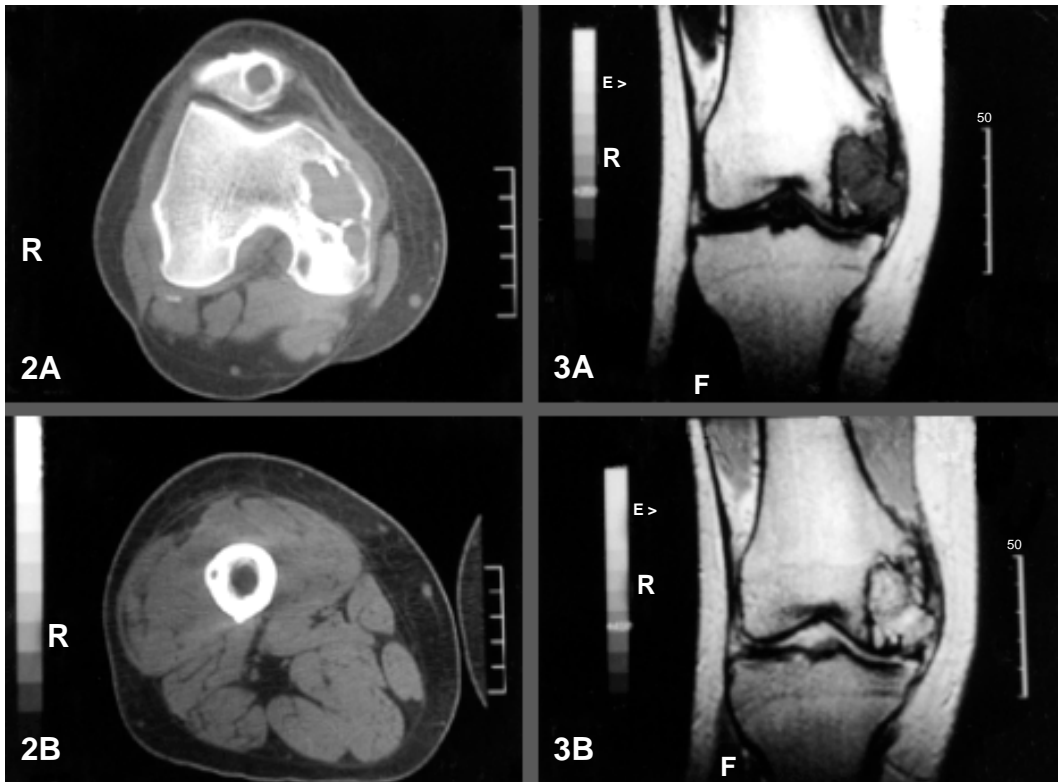


Fig. 2. Computed tomography (CT) scans. **A:** Well-defined, osteolytic lesions with a thin sclerosing margin in the right femur and the patella. **B:** Intracortical lesion of the right fibula.

Fig. 3. Magnetic resonance (MR) images. **A:** T1-weighted image shows low signal intensity in the right femoral tumor, which is close to the skeletal muscle. **B:** In T2-weighted image, the intensity of the right femoral tumor is high.

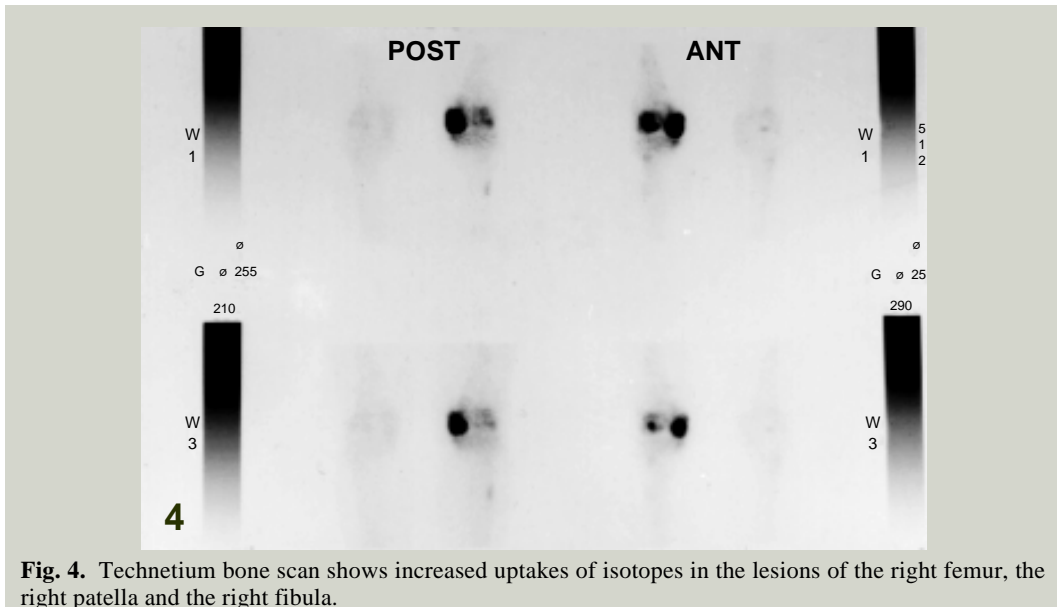


Fig. 4. Technetium bone scan shows increased uptakes of isotopes in the lesions of the right femur, the right patella and the right fibula.