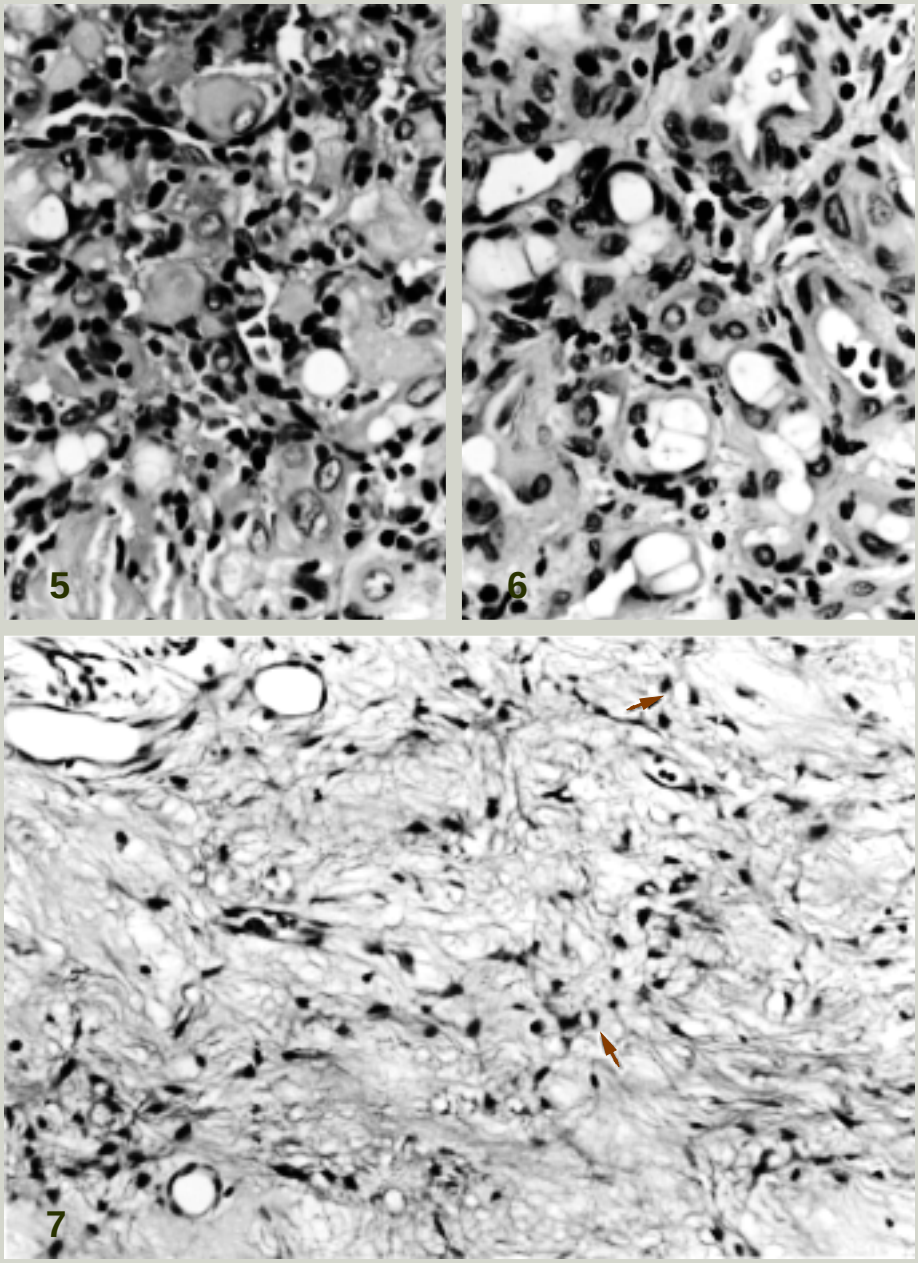

Fig. 5. The tumor consists of an accumulation of epithelioid cells forming nests in the growth pattern with capillaries.

Fig. 6. Epithelioid cells showing abundant, eosinophilic cytoplasms with intracytoplasmic vacuoles in various stages. The vascular channel is lined by large, plump tumor cells with an aggregation of epithelioid cells outside.

Fig. 7. A fibromyxomatous stroma provides the background. The tumor cells (arrow) with vacuoles are arranged in a cord-like pattern.

showed an abundant, eosinophilic cytoplasm often with various-sized intracytoplasmic vacuoles. The vascular channel was lined by large, plump tumor cells with an aggregation of epithelioid cells outside (Fig. 6). Mitotic figures were scarcely observed in all 4 lesions. A fibromyxomatous stroma provided as the background in a large area taken from the tumors of the patella and the fibula; in the fibromyxomatous stroma, the tumor cells with cytoplasmic vacuoles were arranged in a cord-like pattern (Fig. 7). Lymphocytic aggregation was scarce in the stroma in a limited area. We performed immunohistochemical examination for Factor VIII-related antigen and CD34 with monoclonal antibodies, and detected strongly positive reactions from the tumor cells including the vacuolating cells.

Discussion

The diagnostic term EHE was introduced for unusual bone and soft-tissue tumors as a counterpart to vascular tumors, similar to those originating in the visceral organs with a 1% incidence among all vascular tumors (Weiss and Enzinger, 1982; Costa et al., 1996; Keel and Rosenberg, 1999). EHE is histologically characterized by clusters of epithelioid cells with various-sized vasculatures in the background of myxoid or fibrous stroma (Weiss and Enzinger, 1982; Maruyama et al., 1985; Tsuneyoshi et al., 1986; Bollinger et al., 1994). The epithelioid cells frequently show various-sized intracytoplasmic vacuoles, which mimicked a primitive vascular channel: positive reactions for Factor VIII-related antigen and CD34 and the presence of Weibel-Palade granules proved that the epithelioid cells have a distinctive nature among endothelial cells (Tsuneyoshi et al., 1986). In the present patient, the tumors were initially observed to consist of clusters of epithelioid cells which had vacuoles compatible to the primitive blood vessels. The cord-like arrangement of CD34-positive, vacuolated cells was also recognized in the fibromyxoid matrix, which clearly indicated EHE as mentioned by Weiss and Enzinger (1982). Epithelioid cells were accumulated in a small area around the well-developed vascular channel, which mimicked epithelioid hemangioma. Initially, EHE was proposed as one type of vascular tumor with epithelioid cells including epithelioid hemangioma and epithelioid angiosarcoma (Weiss et al., 1986). Epithelioid hemangioma takes a benign clinical course, and is often mistaken for EHE (O'Connell et al., 1993; Costa et al., 1996).

On plain radiographs, EHE appears solitarily or multicentrically, being purely lytic and expansive with well-defined sclerotic borders, as in benign vascular tumors (Tsuneyoshi et al., 1986). A coarse trabecular or honeycomb pattern is also common, but is rarely seen with a destruction pattern in the elderly (Bollinger et al., 1994), as Murphy et al. (1995) suggested. In pulse sequences emphasizing a T1 contrast, bone tumors of a vascular origin show higher intensities than skeletal muscles but lower than fat; while in pulse sequences emphasizing a T2 contrast, signal intensities of bone tumors of a vascular origin are significantly higher than muscles and fat (Boutin et al., 1996). In the present patient, signal intensity of the epiphyseal tumor of the right femur was close to that of muscles in T1-weighted images, whereas in T2-weighted images, the intensity of the bone tumor was higher than that of muscles and with well-defined margins.

EHE of the bone is generally categorized as a tumor of intermediate malignancy, slowly growing with initial symptoms of pain and local swelling. The clinical course of EHE of the bone depends on the degree of tumor malignancy, between an epithelioid hemangioma and an epithelioid angiosarcoma. It is most likely that cancellous bone and cortical bone of the lower extremities are involved in EHE (Boutin et al., 1996). In EHE, lesions appear in a multicentric manner with an incidence of over 50% and below 62%, particulary with a predilection for bones of the lower extremities in one anatomic region (Bollinger et al., 1994). Bones of the upper extremities, ribs, vertebrae and scapulae might be also involved in EHE, though the incidence would be lower (Tsuneyoshi et al., 1986). The multicentric type of EHE seems to

take a benign course in affected regions, but is more indolent than the solitary type (Kleer et al., 1996; Weiss et al., 1986). EHE associated with cutaneous and visceral organs gives a serious influence on the clinical course (Kleer et al., 1994). Therefore, CT scanning and MR imaging should be employed in predicting the prognosis of vascular tumors for a thorough examination of a patient.

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