Epithelioid Sarcoma-like Haemangioendothelioma: A Case Report

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Sir,

Epithelioid sarcoma is a soft tissue tumour of uncertain origin. Epithelioid cells with eosinophilic cytoplasm are present together with spindle cells. On the other hand, epithelioid sarcoma-like haemangioendothelioma (ES-H) is a rare, low-grade, potentially malignant vascular neoplasm first described by Billings et al. in 2003 (1). Until now only 8 cases have been reported (1, 2). Because of their clinical and histological similarities, it may be misdiagnosed initially as epithelioid sarcoma. However, ES-H is generally indolent compared with epithelioid sarcoma. We describe here the clinical and histopathological features of a case of ES-H that developed in a middle-aged Japanese man.

CASE REPORT

A 56-year-old Japanese man presented with a 1-year history of tender masses on the right index finger and right forearm. Physical examination revealed 2 firm, immobile, poorly demarcated masses on the right index finger (Fig. 1a, b) and 2 similar masses on the flexor aspect of the right forearm (Fig. 1c). Magnetic resonance imaging revealed hyper-intense areas in the subcutaneous layer (Fig. 1d). Biopsied specimens obtained from tip of the index finger and the forearm showed multiple ill-defined nodules composed of rounded epithelioid cells and spindle cells. The epithelioid cells contained a prominent eosinophilic cytoplasm with an indistinct cytoplasmic border (Fig. 2a). There were transitional zones in which the cells acquired a spindle shape and lost their cohesion (Fig. 2b). In the spindle cell areas the composing cells were arranged in a fascicular or haphazard fashion (Fig. 2c). Some of them contained intracytoplasmic lumens, and there were a few multicellular vascular channels. Hyaline matrix was identified in some nodules (Fig. 2d). Immunohistochemically, cytokeratin and CD31 tested positive, whereas other stains including CD34, S-100, HMB-45, smooth muscle actin and desmin were negative (Fig. 3). Furthermore, 5% of the tumour cells were Ki67-positive. Based on these histopathological and immunohistochemical features, we diagnosed the subcutaneous tumours as ES-H. Although all the tumours were surgically removed, a new lesion developed in the right index finger 6 months later. The patient refused our suggestion of amputating the right arm, but accepted amputation of the right index finger at the base.

DISCUSSION

ES-H is a vascular tumour of low-grade malignancy that closely mimics epithelioid sarcoma. Clinically, it is a subcutaneous tumour chiefly developing in the extremities, in a fashion similar to epithelioid sarcoma. Histopathologically, it is composed of ill-defined nodules and fascicles containing eosinophilic epithelioid

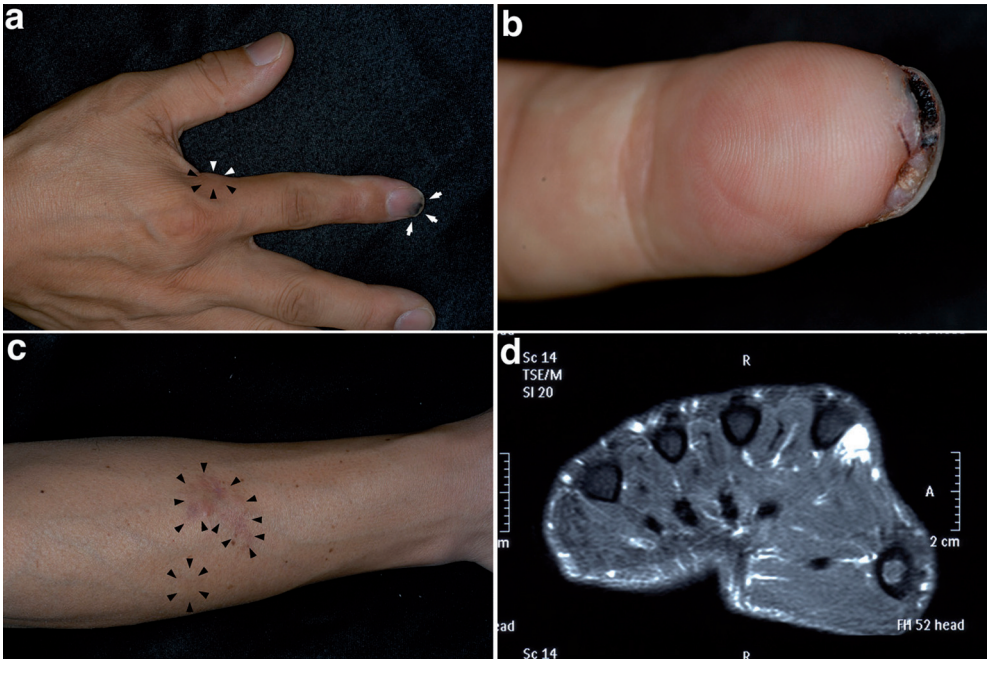

Fig. 1. Clinical features of the subcutaneous tumours (a, b) on the right index finger and (c) on the right forearm. (d) Magnetic resonance imaging of the right hand: hyperintense nodular shadow at the base of the right index finger.

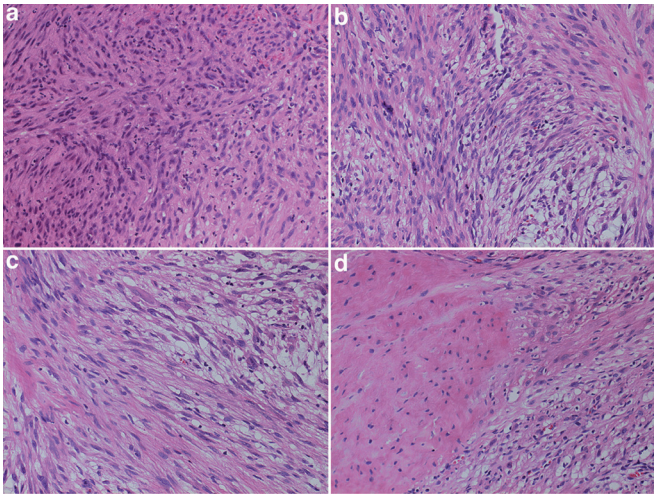

Fig. 2. Histopathological features. (a) Epithelioid cells with abundant eosinophilic cytoplasm. (b) Transition between epithelioid cells and spindle cells. (c) Spindle cells within collagen bundles. (d) Hyaline matrix within the nodules Haematoxylin-eosin ×200.

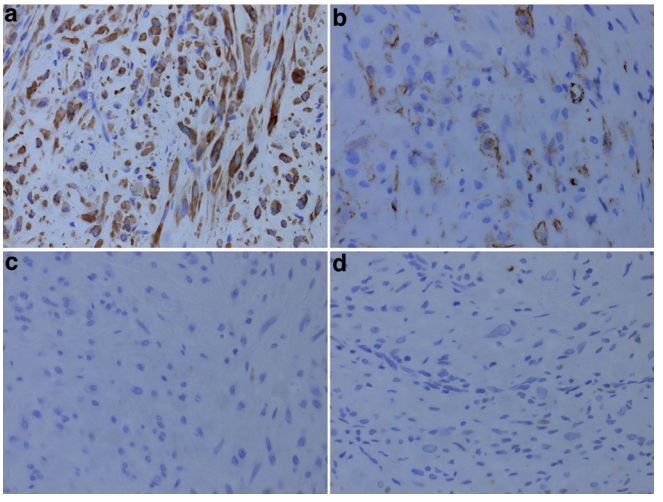

Fig. 3. Immunohistochemical features. (a) Cytokeratin (AE1/AE3) staining. (b) CD31 staining. (c) CD34 staining. (d) S-100 staining. All images haematoxylin-eosin ×400.

cells. Cytoplasmic vacuoles, which are suggestive of its vascular differentiation, are present, but multicellular channel formation is rarely observed. In addition to these features, its intense cytokeratin expression leads us to consider other diagnoses, such as an epithelioid sarcoma or undifferentiated neoplasms.

Immunohistochemical examination is informative for a correct diagnosis. In contrast to epithelioid sarcoma, which is CD31-negative (3), ES-H usually expresses CD31 in a linear membranous pattern, which is highly specific for endothelial cells (1). Furthermore, ES-H seems to be negative for CD34 even though it originates from vascular cells. Since CD34 is expressed in most of the vascular tumours (3, 4), it can be used for the differentiation from epithelioid haemangioendothelioma and epithelioid angiosarcoma, both of which are CD34-positive.

Epithelioid sarcoma has a high risk for local recurrence and metastasis and requires long-term follow-up, since its recurrence or metastasis may occur many years after the initial diagnosis (5). Despite similar clinical and histopathological features, ES-H has been considered to exhibit indolent behaviour because of the absence of metastases to internal organs or beyond the regional lymph nodes (1). However, ES-H should be considered as a tumour of intermediate malignancy because of the possibility of multiple tumours, local recurrence and regional soft tissue metastasis similar to other haemangioendotheliomas. Although ES-H seems to be generally indolent, radical amputation was more pertinent, and close follow-up is necessary in the present case. It is important to accumulate follow-up information for many more cases in order to reach a firm conclusion concerning the biological behaviour of this condition. ES-H must be kept in mind as an entity belonging to the haemangioendothelioma family.

The authors declare no conflict of interest.

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