

Primary Cutaneous Epithelioid Angiosarcoma

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Sir,

Epithelioid angiosarcoma is a rare subtype of angiosarcoma that is histologically characterized by oval or polygonal cells with abundant, faintly eosinophilic cytoplasm (1). It commonly occurs in deep soft tissues, but can rarely occur on the skin. We report here a case of cutaneous epithelioid angiosarcoma located on the buttock and review the previously published cases.

CASE REPORT

A 75-year-old woman noticed a nodule on her left buttock one week prior to admission. On physical examination, an elastic hard intradermal nodule, 6 × 10 cm in diameter, was noted (Fig. 1a). Inguinal lymph nodes were not palpable. Histological examination showed a tumour mass in the dermis extending into the upper subcutaneous tissues, which did not invade the fascia. The tumour cells consisted of oval or polygonal epithelioid cells with eosinophilic cytoplasm and large eosinophilic nuclei (Fig. 1b). A number of mitotic figures were noted. Although well-formed vascular walls consisting of malignant cells were not found, there were prominent slit-like structures. Immunohistochemical studies revealed that these slit-like structures were positive for CD31 (Fig. 1c). CD31 corresponds to platelet endothelial cell adhesion molecule-1 (PECAM-1). The tumour cells were also positive for vimentin, CD34, and factor VIII-related antigen, but not for S100 protein, AE1/AE3, or HMB-45. Thus, the histological diagnosis of epithelioid angiosarcoma was made. The tumour was resected with a 7-cm margin from the periphery, together with a part of the gluteus maximus muscle. Postoperatively, the patient received docetaxel hydrate chemotherapy. Eighteen months after excision, the patient has remained healthy without clinical evidence of tumour recurrence.

DISCUSSION

Previously reported cases of cutaneous epithelioid angiosarcoma since 1986 are presented in Table I. The male/female ratio was 11/3. Of the 14 cases, 7 were located on the scalp or forehead, and 3 on the extremities. None of the cases had lesions on the trunk, except for cases 4 (breast) and 11 (shoulder). Thus, the buttock is involved extremely rarely. Only 2 cases had pre-existing diseases (case 6 had variceal surgery 12 years earlier, and case 8 had lymphoedema due to

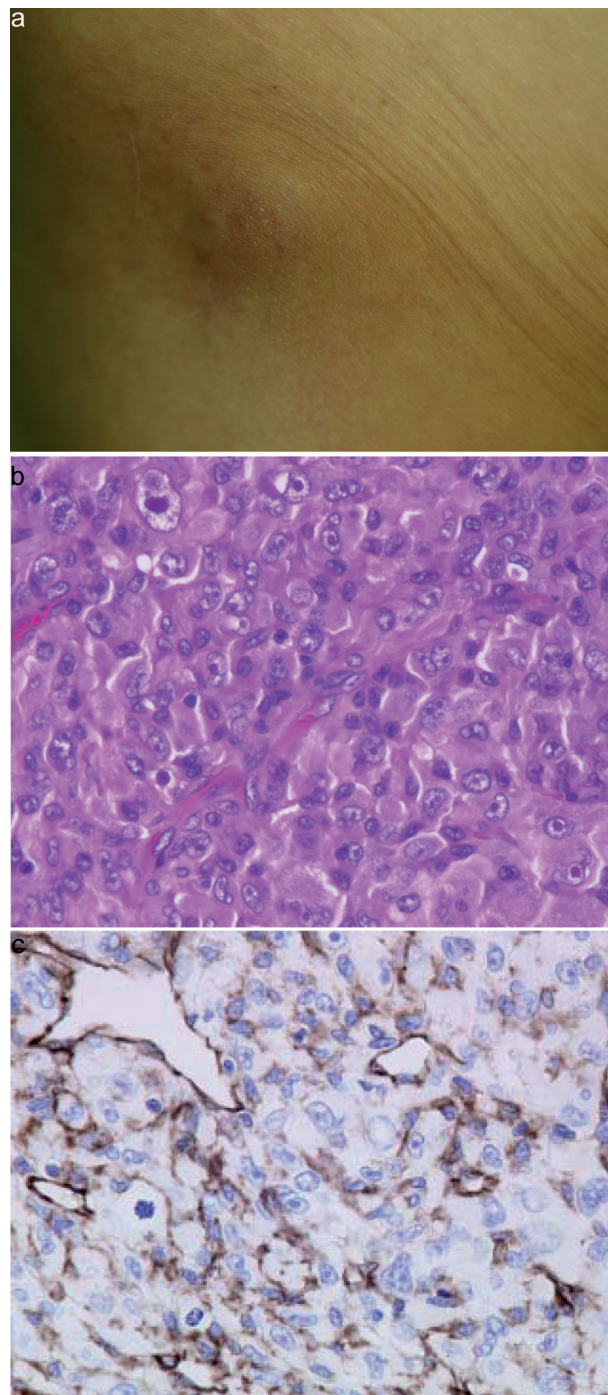


Fig. 1. (a) A large dermal elastic-hard mass is noted on the buttock. (b) Tumour cells are composed of large cells with faintly eosinophilic cytoplasm, forming slit-like structures (haematoxylin and eosin (H&E)). (c) The luminal surfaces of the slit-like structures are positive for CD31.

Table I. Clinical findings of reported cases of primary cutaneous epithelioid angiosarcoma

Case	Age (years)/gender	Site	Disease duration	Therapy	Prognosis	Reference
1	41/M	Shoulder	3 years	S and R	67 months	2
2	53/M	Nose	2 months	S and R	53 months	2
3	69/M	Scalp	9 months	S	18 months	3
4	64/M	Scalp	NA	NA	NA	5
5	83/F	Forehead	NA	R	6 months	5
6	45/F	Breast	NA	S and R	5 years	5
7	74/M	Forehead	NA	S and R	4 years	5
8	90/M	Scalp	NA	NA	<30 months	6
9	71/M	Thigh	NA	NA	<30 months	6
10	81/M	Scalp	NA	NA	<5 years	6
11	52/M	Upper leg	NA	NA	<30 months	6
12	65/M	Scalp	1 year	NA	<5 years	7
13	39/M	Forearm	NA	NA	64 months	8
14	21/M	Thigh	1 year	S, R and C	16 months	9
15	75/F	Buttock	1 week	S and C	18 months (disease-free)	Present study

M: male; F: female; R: radiation therapy; S: surgical excision; C: chemotherapy; Prognosis: Duration of survival excluding (cases 1–3) or including (cases 5–15) disease duration; NA: data not available from the literature.

lymphadenectomy for malignant melanoma 18 years earlier); both of these pre-existing conditions may have triggered angiosarcoma.

A standard therapeutic protocol for cutaneous epithelioid angiosarcoma has not been established. Of the reported cases, 4 received combination therapy involving surgical excision and radiation therapy, and 2 were treated with either surgery or radiation therapy alone. In the other cases the therapies were not documented. Our case was treated with surgical excision and post-operative docetaxel hydrate chemotherapy.

The difference in the prognosis between epithelioid angiosarcoma and ordinary angiosarcoma is still controversial. Some reports suggest that epithelioid angiosarcoma is a lower grade malignancy than ordinary angiosarcoma (1, 2), while other reports suggest that angiosarcoma has a high risk of recurrence and metastasis (3, 4). Further study is required to determine the malignancy grade and to standardize the therapeutic protocol for patients with cutaneous epithelioid angiosarcoma.

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