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CASE REPORT

Giant Vascular Eccrine Spiradenoma

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Giant vascular eccrine spiradenomas (GVESs) are a rare variant of the eccrine spiradenoma that develops from the sweat gland. It is different from the eccrine spiradenoma in its larger size and greater degree of vascularity. Bleeding and/or ulceration are common clinical features of this tumor, and are the reason why it is often clinically confused with a vascular or malignant tumor. Here, a rare case of GVES without bleeding or ulceration is reported. **(Ann Dermatol 23(S2) S197～S200, 2011)**

-Keywords-
Eccrine spiradenoma, Giant vascular eccrine spiradenoma, Sweat gland neoplasms

INTRODUCTION

Giant vascular eccrine spiradenoma (GVES) is a variant of an eccrine spiradenoma. GVES is very rare and differs from the eccrine spiradenoma in its larger size clinically and marked vascularity histopathologically. GVES is usually accompanied by bleeding and/or ulceration, and often misdiagnosed as a vascular or malignant tumor clinically[1].

Here, we report a case of GVES. The lesion presented as a slightly bluish mass and the overlying skin of the lesion was intact without erosion, ulceration, or hemorrhage. The diagnosis of GVES was made via histopathological examination.

CASE REPORT

A 49-year-old woman presented with a solitary deep mass of 10-year duration on the right upper arm. The lesion was asymptomatic, firm, and 2.5×2.5 cm in size, with a bluish color in the overlying intact skin (Fig. 1). On physical examination, there were no abnormal findings other than the cutaneous lesion. An excisional biopsy was performed and the specimen was grossly hemorrhagic. The histopathological findings of the lesion showed a large, well-circumscribed, encapsulated, multilobulated tumor in the dermis. There was extensive hemorrhage and numerous dilated vascular spaces containing red blood cells or pale pinkish lymph fluid in the tumor (Fig. 2A). The lobules were composed of two different types of cells forming cords, bands, or pseudoglandular rosettes: cells with small, dark nuclei at the periphery and cells with large pale nuclei in the center of the lobules (Fig. 2B). Immunohistochemically, the tumor cells were positive for cytokeratin (CK), epithelial membrane antigen (EMA), S-100, smooth muscle actin (SMA), and p63 (Fig. 3). They were negative for carcinoembryonic antigen (CEA), CD15,

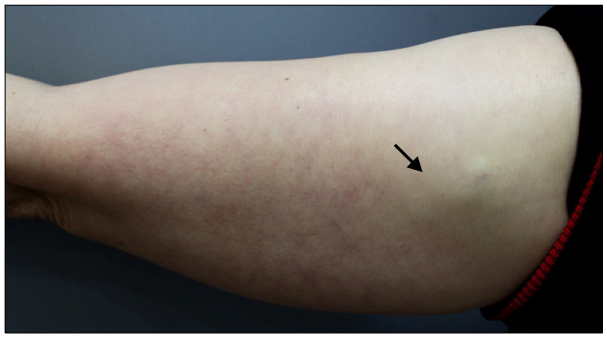

Fig. 1. A solitary, bluish, 2.5×2.5 cm, firm deep mass (arrow) on the right upper arm.

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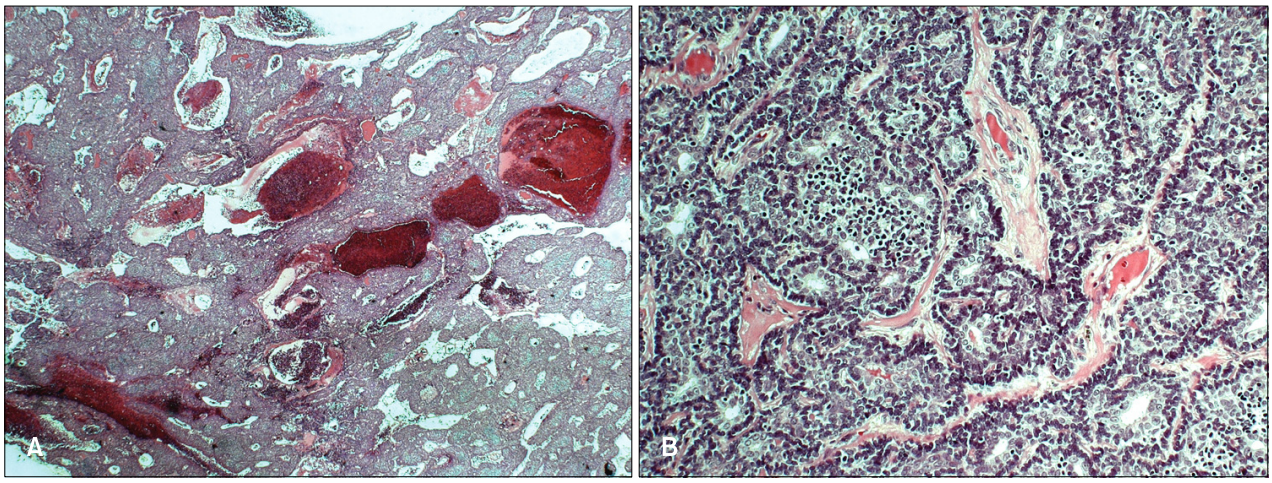

Fig. 2. (A) Well-demarcated, encapsulated, multilobulated tumor in the dermis with many blood-filled vessels and extensive hemorrhage (H&E, ×25). (B) The tumor was composed of two types of cells: cells with small, dark nuclei at the periphery of the lobules and cells with large, pale nuclei in the center of the lobules (H&E, ×200).

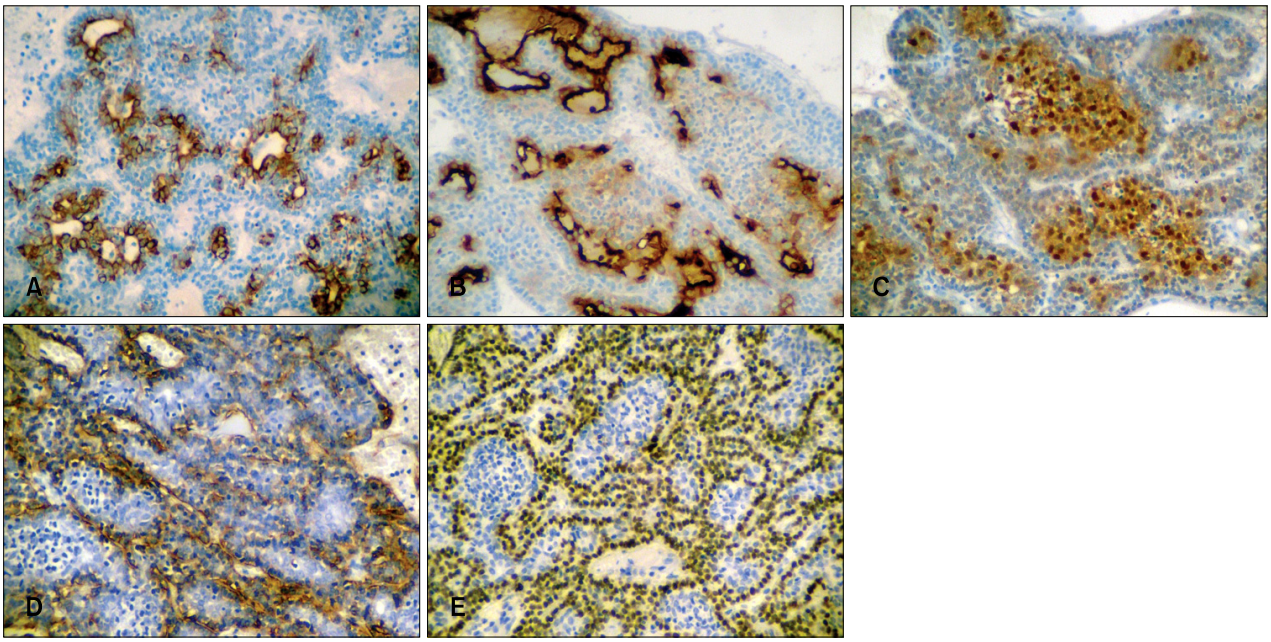

Fig. 3. Immunohistochemical staining for CK (A: ×100), EMA (B: ×100), S-100 (C: ×100), SMA (D: ×100), and p63 (E: ×100). CK: cytokeratin, EMA: epithelial membrane antigen, SMA: smooth muscle actin.

gross cystic disease fluid protein-15, and lysozyme. The lesion was removed completely with an excisional biopsy and no recurrence has occurred for six months following the procedure.

DISCUSSION

GVES is a rare variant of eccrine spiradenoma and was first described by Cotton et al.[1] in 1986. Eccrine spiradenoma usually presents as a solitary, reddish-brown, sometimes painful, deep nodule measuring 1 to 2 cm in diameter[2,3]. GVES is different from eccrine spiradenoma in that it is larger than the eccrine spiradenoma clinically, and has a marked degree of vascularity histopathologically. In the English literature, there are only six cases of GVES (Table 1)[1,4-7]. All previously reported cases of GVES, including this case, were in adults or the elderly, and had a large lesion of more than 2 cm in size. The locations were variable in each case and included the abdomen, back of scalp, costochondral line, lower back, and

Table 1. Clinical features of patients with giant vascular eccrine spiradenoma in the English literature

Reference	Age/Sex	Size (cm)	Location	Clinical diagnosis
Cotton et al.[1]	74/M	5	Abdomen	Angiosarcoma or malignant melanoma
	84/F	2	Back of scalp	Sebaceous cyst
Hey et al.[4]	63/F	3.5×1.5	Right thigh	Venous thrombosis
Senol et al.[5]	60/M	3~4	Right costochondral line	Angiomatous lesion or thrombosis
Ko et al.[6]	56/F	2	Right lower back	Angiolipoma or neuroma
Yamakoshi et al.[7]	76/M	5×3.4×2.6	Right shoulder	Not mentioned
Present case	49/F	2.5×2.5	Right upper arm	Vascular tumor or venous malformation

M: male, F: female.

shoulder.

Bleeding and ulceration are common clinical features of GVES. Therefore, GVES may result in misdiagnosis of angiomatous lesion or malignant neoplasm. Among previously reported cases of GVES, the lesions in four cases showed bleeding or ulceration[1,5,7]. The lesions had been misdiagnosed clinically as vascular lesion or malignancy with diagnoses such as angiosarcoma, malignant melanoma, and thrombosis. Compared with other cases, this case had a distinctive clinical appearance of a slightly bluish color with no bleeding or ulceration.

The histopathological features of GVES are similar to those of eccrine spiradenoma. Eccrine spiradenoma consists of several lobules located in the dermis without connections to the epidermis. The lobules are sharply demarcated and encapsulated with fibrous tissue. Two types of cells are present in the tumor. One cell type has small, dark nuclei, and is located at the periphery of the lobules. The other cell type has large, pale nuclei, and is located in the center of the lobules. These cells are arranged in anastomosing cords and bands or pseudoglandular rosettes[3]. In addition, GVES has characteristic histopathological features. It has a large number of wide vessels filled with red blood cells or lymph fluid in the stroma and an extensive hemorrhagic appearance[6].

Eccrine spiradenoma is thought to originate from eccrine sweat glands. By immunohistochemistry, the tumor cells of eccrine spiradenoma express CK, CEA, EMA, and myoepithelial differentiation and is identified by positive staining for SMA and S-100[8-10]. Immunohistochemical study of the GVES shows a large lumen and pale epithelial cells positive for CK, CK7, Cam5.2, and EMA. In addition, the outer layer of the small basaloid cells are positive for p63, and many p63+/SMA+ myoepithelial cells are present[6]. Therefore, eccrine spiradenoma has been regarded as a tumor differentiating mainly to the secretory portion of the secretory coil of eccrine sweat glands. In the present case, the CK, EMA, and S-100 were positive in the tumor cells and the CEA was weakly positive only in the luminal secretions which is consistent with prior reports. However, there are several lines of evidence that suggest that eccrine spiradenoma is a neoplasm of apocrine lineage rather than eccrine origin. First, eccrine spiradenoma can be accompanied by cylindroma or trichoblastoma which are tumors of apocrine or folliculosebaceous-apocrine origin. Second, eccrine spiradenoma usually occurs where apocrine elements are rich, such as breast, axilla, genitalia, and ear. Third, eccrine spiradenoma occasionally presents the histopathological features of decapitation secretion[11].

GVES is rare and easily mistaken for vascular or malignant tumors because of its hemorrhagic and/or ulcerative tendencies. Here, we describe a case of GVES with a bluish deep mass and no hemorrhage or ulceration.

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