Giant Vascular Eccrine Spiradenoma

Min Ho Kim, M.D., Eujin Cho, M.D., Jeong Deuk Lee, M.D., Ph.D., Sang Hyun Cho, M.D., Ph.D.

Department of Dermatology, Incheon St. Mary’s Hospital, College of Medicine, The Catholic University of Korea, Seoul, Korea

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.

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 pseudogl andular 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.
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      |
| Hey et al.²        | 63/F    | 3.5 x 1.5 | Right thigh         | Venous thrombosis                      |
| Senol et al.³      | 60/M    | 3 ~ 4     | Right costochondral line | Angiomatous lesion or thrombosis       |
| Ko et al.⁴        | 56/F    | 2         | Right lower back    | Angiolipoma or neuroma                  |
| Yamakoshi et al.⁷  | 76/M    | 5 x 3.4 x 2.6 | Right shoulder  | Not mentioned                          |
| Present case       | 49/F    | 2.5 x 2.5 | Right upper arm     | Vascular tumor or venous malformation   |

M: male, F: female.

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 ulceration¹,⁵,⁷. 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 rosettes³. 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 appearance⁶.

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-100⁶,¹⁰. 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 basaild cells are positive for p63, and many p63+/SMA+ myoepithelial cells are present⁶. 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 secretion¹¹.

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.

REFERENCES

1. Cotton DW, Slater DN, Rooney N, Goepel JR, Mills PM. Giant vascular eccrine spiradenomas: a report of two cases with histology, immunohistology and electron microscopy. Histopathology 1986;10:1093-1099.
2. Taylor RS, Perone JB, Kaddu S, Kerl H. Appendage tumors and hamartomas of the skin. In: Wolff K, Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, editors. Fitzpatrick’s dermatology in general medicine. 7th ed. New York: McGraw-Hill, 2008:1068-1087.
3. Ahmed TS, Priore JD, Seykora JT. Tumors of the epidermal appendages. In: Elder DE, Elenitsas R, Johnson BL, Murphy GF, Xu X, editors. Lever’s histopathology of the skin. 10th ed. Philadelphia: Lippincott Williams & Wilkins, 2009:851-909.
4. Hey A, Grouls V, Röckelein G. Vascular eccrine giant spiradenoma—a case report with histology and immunohistology of a rare variant of benign sweat gland tumors. Z Hautkr 1988;63:444-447.
5. Senol M, Ozcan A, Sasmaz S, Ozen S, Ciralik H. Giant vascular eccrine spiradenoma. Int J Dermatol 1998;37:221-223.
6. Ko JY, Lee CW, Moon SH, Song KW, Park CK. Giant vascular eccrine spiradenoma: report of a case with immunohistochemical study. J Korean Med Sci 2006;21:172-176.
7. Yamakoshi T, Makino T, Watanabe H, Furuichi M, Matsui K, Shimizu T. A case of giant vascular eccrine spiradenoma with unusual clinical features. Clin Exp Dermatol 2009;34:e250-e251.

8. Eckert F, Betke M, Schmoeckel C, Neuweiler J, Schmid U. Myoepithelial differentiation in benign sweat gland tumors. Demonstrated by a monoclonal antibody to alpha-smooth muscle actin. J Cutan Pathol 1992;19:294-301.

9. Wiley EL, Milchgrub S, Freeman RG, Kim ES. Sweat gland adenomas: immunohistochemical study with emphasis on myoepithelial differentiation. J Cutan Pathol 1993;20:337-343.

10. Meybehm M, Fischer HP. Spiradenoma and dermal cylindroma: comparative immunohistochemical analysis and histogenetic considerations. Am J Dermatopathol 1997;19:154-161.

11. McCalmont TH. A call for logic in the classification of adnexal neoplasms. Am J Dermatopathol 1996;18:103-109.