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Lymphangiectatic Variant of Low-Grade Malignant Eccrine Spiradenoma

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Low-grade malignant eccrine spiradenoma (spiradenocarcinoma) is a rare sweat gland tumor, which usually arises from a pre- existing benign eccrine spiradenoma. This paper presents the case of a 55-year-old male who had a lesion in his right elbow for 10 years. The microscopic examination revealed a well-demarcated, multilobulated tumor in the dermis and subcutis, which presented with many blood-filled vessels and extensive hemorrhage. The tumor was composed of hyperchromatic, round to oval cells with nucleolar prominence, mild to moderate atypia, and increased mitotic index. Additionally, lymphangiectatic appearance was observed in areas with prominent stromal lymphedema. P53 and Ki-67 had high positivity. Surgical excision of the lesion was performed with adequate surgical margins, and the dissected lymph nodes in the axilla were tumor-negative. After 15 months of follow-up, there was no recurrence or distant metastasis.

Keywords: Lymphangiectatic variant, Malign eccrine spiradenoma, Spiradenocarcinoma

INTRODUCTION

In this report, we present the case of low-grade malignant eccrine spiradenoma (spiradenocarcinoma) (LGMES) which can easily be confused with vascular lesions.

CASE REPORT

A 55-year-old male patient was admitted to our hospital with a lesion that had been present on his right elbow for 10 years. The patient had noticed enlargement, ulceration, intermittent bleeding, and pain for the past one year. The skin lesion and the axillary mass were surgically removed. The macroscopic examination revealed an ulcerated, hemorrhagic, bluish-reddish tumor of 4.5 cm in size, 2 cm raised from the surface (Fig. 1). The microscopic examination revealed a well-demarcated, ulcerated, multilobulated tumor in the dermis and subcutis which presented with many blood-filled vessels and extensive hemorrhage (Fig. 2A). The benign eccrine spiradenoma (ES) was present at the edge of the tumor, as a thin compressed rim (Fig. 2B). Additionally, lymphangiectatic appearance was observed in areas with prominent stromal lymphedema (Fig. 2C~E). Hyperchromatic tumor cells showed mild to moderate atypia, increased mitotic index (15 mitotic figures/10 high-power fields), nucleolar prominence, loss of polarity, and dual cell population (Fig. 3A). Ki-67 expression was 20%

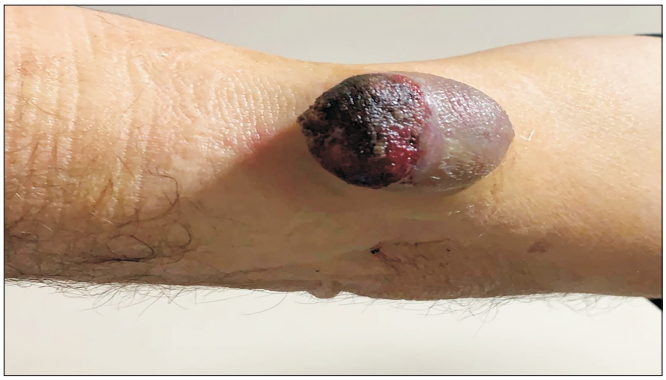

Fig. 1. An ulcerated cutaneous nodule with red-blue-brown discoloration. We received the patient's consent form about publishing all photographic materials.

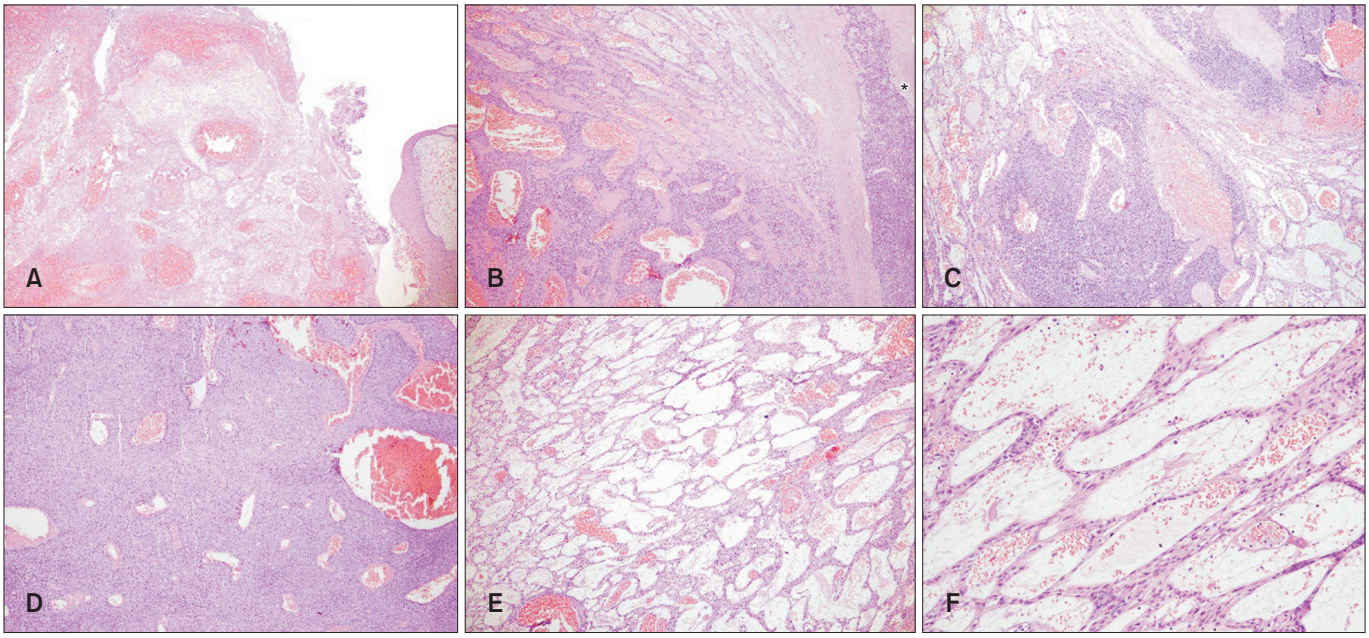

Fig. 2. (A) An ulcerated, hemorrhagic tumor in the epidermis-dermis. (B) The benign eccrine spiradenoma (asterisk) was present at the edge of the tumor. (C~E) A multilobulated tumor with wide congestive vein structures embedded in microcystic spaces. (F) Lymphangiectasia-like microcystic areas consisting of spindle cells (A~E: H&E, ×40; F: H&E, ×100).

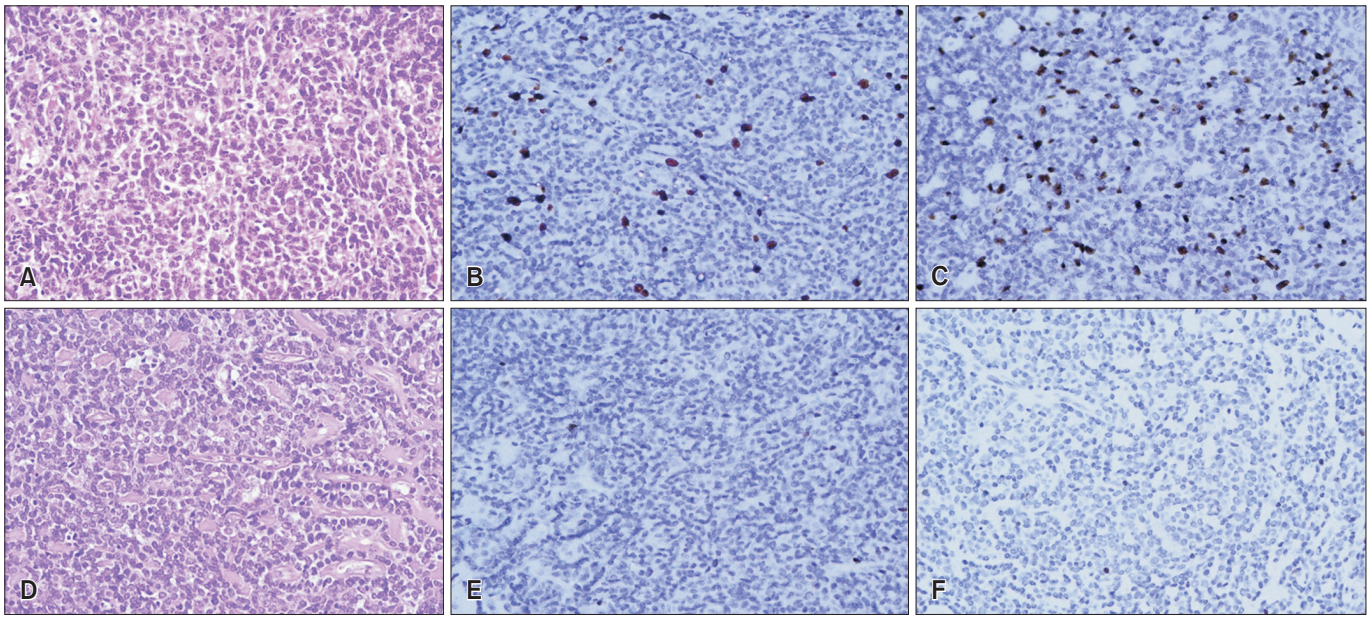

Fig. 3. (A) Hyperchromatic cells with mild to moderate atypia, increased mitotic index, nucleolar prominence, loss of polarity, and dual cell population in low-grade malignant eccrine spiradenomas (LGMES). (B, C) Ki-67 and p53 positive cells of LGMES (20% and 80%, respectively). (D) Eccrine spiradenoma (ES) showed dual cell population composed of large cells with pale nuclei arranged in the center and small cells with hyperchromatic nuclei at the periphery. (E, F) Very low Ki-67 and p53 expression in the ES (A~F: H&E, ×400).

(Fig. 3B), and p53 expression was 80% (Fig. 3C). However, ES showed dual cell population composed of large cells with pale nuclei arranged in the center and small cells with hyperchromatic nuclei at the periphery (Fig. 3D) and very low Ki-67 and p53 expressions (Fig. 3E, F). Immunohistochemistry spindle cells in lymphangiectatic areas were positive

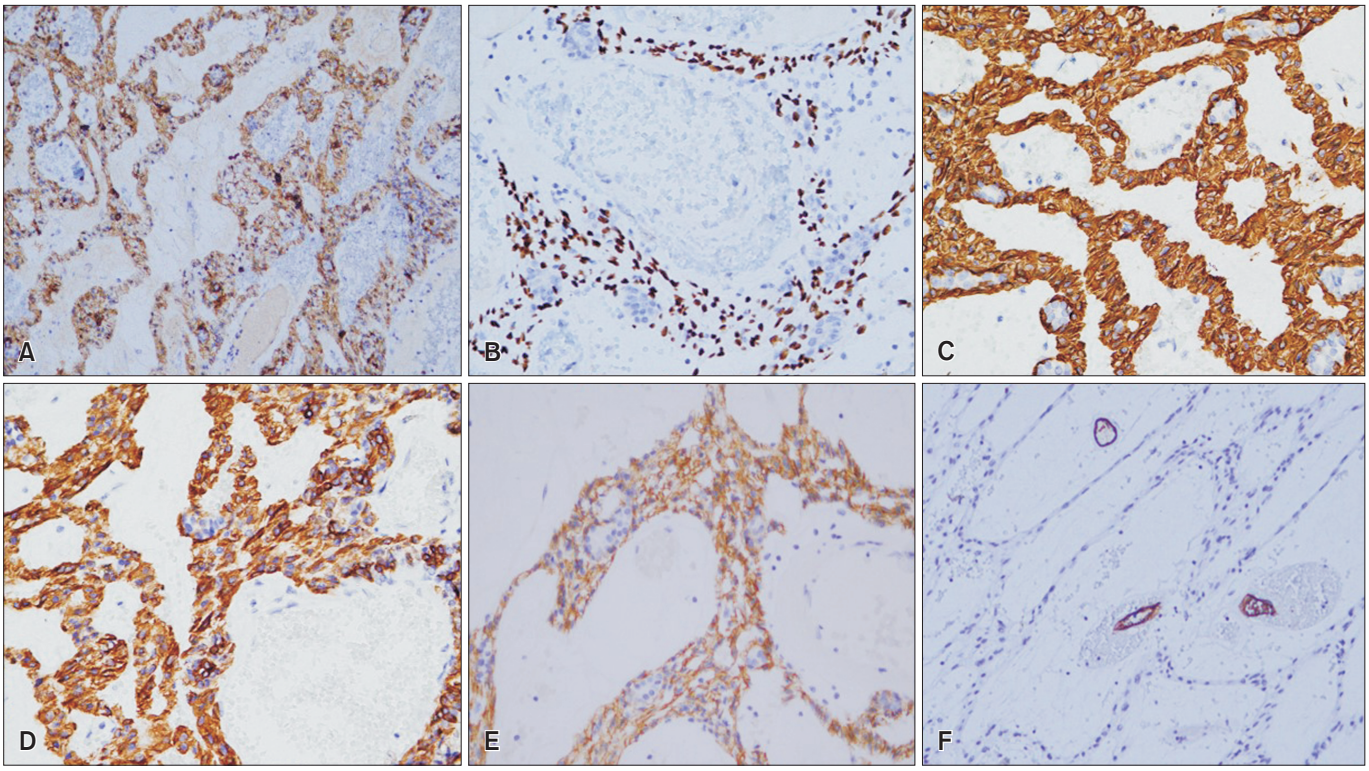

Fig. 4. (A) S100, (B) p63, (C) CK5/6, (D) smooth muscle actin (SMA), and (E) D2-40 indicating the myoepithelial cell origin of spindle cells. (F) CD34 negativity in spindle cells (A~F: H&E, ×400).

for S100, p63, smooth muscle actin (SMA), CK5/6, and D2-40, indicating a myoepithelial cell origin (Fig. 4A~E). CD34 was negative in spindle cells (Fig. 4F). A 4-cm right axillary mass was compatible with reactive follicular hyperplasia. After 15 months of follow-up, there was no recurrence or distant metastasis.

DISCUSSION

LGMES, first described by Leonard et al.,[1] are rare tumors, mostly located in the head and neck. LGMESs are generally found among elderly patients admitted to clinics with long-standing lesions which show rapid growth[1-6].

The histopathological criteria for distinguishing LGMES from ES are the loss of a periodic acid-Schiff-positive basement membrane, necrosis, ulceration, nuclear pleomorphism and hyperchromasia, nucleolar prominence, loss of polarity, dual cell population, an increased mitotic index, and invasion of surrounding tissues. Our patient exhibited all these malignancy criteria. In addition to high p53 and Ki-67 indexes, the loss of MYB expression has been reported as an important finding in malignancy[2,3]. Our patient had 80% P53 positivity and 20% Ki-67 positivity.

LGMESs have a very good clinical course, total excision and follow-up are sufficient as treatment, and there is no need for a sentinel lymph node biopsy and extended surgical margin[2-6].

In our patient, surgical excision with a negative and adequate surgical margin was performed. Clinically palpable axillary lymph nodes were dissected, and were negative for tumor. After 15 months of follow-up, there was no recurrence or distant metastasis.

In the literature, the lymphangiectatic variant was first defined in 1983 by Mambo[7]. To the best of our knowledge, there are very few cases of lymphangiectatic variants of ES in the English literature[8,9]. Lymphangiectatic appearance caused by the pronounced lymphedematous stroma containing mononuclear cells and small vascular channels is reported to simulate lymphangioma[8,9]. Here, we presented the lymphangiectatic variant of LGMES. In this lesion, the predominant cell type was spindle cells showing positive staining with myoepithelial markers. This variant only shows histopathological appearance due to edema, and in the very few

cases that have been reported, no relationship was observed with prognosis[8,9].

The current case represented a diagnostic challenge, and our aim in presenting it was to alert pathologists and clinicians to the possibility of this unusual variant of malignant eccrine spiradenoma, which can easily be confused with vascular lesions.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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