Ann Dermatol Vol. 27, No. 4, 2015 http://dx.doi.org/10.5021/ad.2015.27.4.435

CASE REPORT

Multiple Segmental Eccrine Spiradenoma with a Zosteriform Pattern: A Case Report and Literature Review

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Eccrine spiradenoma usually occurs as solitary tender nodules. However, it rarely occurs as multifocal localized tumors or has zosteriform distribution. We report the case of a 32-year-old woman with a 20-year history of itchy lesions on the left side of the back and forearm with a zosteriform distribution. Before disease onset, almost no patients have inducing factors, but our patient received an injection beforehand; however, whether this was coincidental or causative remains unknown. The lesions became very itchy after perspiration or eating spicy food, which has never been reported. A literature search revealed 22 cases of multiple segmental eccrine spiradenoma; we summarized the clinical characteristic in order to aid diagnosis and treatment selection. **(Ann Dermatol 27(4) 435～438, 2015)**

-Keywords-
Eccrine spiradenoma, Itchy, Multiple, Zosterifrom

INTRODUCTION

Eccrine spiradenoma (ES) is a rare benign dermal tumor of apocrine differentiation originating from cutaneous sweat glands that may be sporadic or familial; it was first described in 1956 by Kersting and Helwing. It mainly affects adults, with the highest incidence in the fourth decade of life[1]. Yoshida et al.[2] divided ES into solitary and multiple ES on the basis of its clinical appearance. Most presentations of ES are solitary and show no predilection for any sex. However, multiple ES predominantly affects women, with a male/female ratio of 1:3. Multiple ES can be described as multifocal or localized with a linear zosteriform distributionand nevoid or blaschkoid pattern (Table 1); however, there is no definitive classification[3].

CASE REPORT

A 32-year-old Chinese woman presented to our hospital complaining of slow-growing painless itchy lesions on the left side of the back and forearm starting approximately 20 years ago with onset about 12-year-old previously as multiple nodules on the arm that gradually increased in size and number. When she was 12 years old, she received a hepatitis B vaccine injection on the left forearm. She stated her lesions were itchy but not painful, and that the itchiness increased after perspiration or eating spicy food. She also felt formication on occasion. She was otherwise well with no other significant medical or family history. Physical examination revealed multiple asymptomatic well-circumscribed blue-to-violet round-to-oval subcutaneous nodules; they exhibited a zosteriform distribution from the left upper back to the forearm (Fig. 1). On palpation, the lesions were rubbery, firm, and fixed to the overlying skin, ranging between 0.5 and 1.5 cm in size.

Histological analysis revealed 2 kinds of tumor cells: small darkly staining basaloid cells, and large pale cells. The nodules comprised basaloid cells arranged in a trabecular pattern surrounded by eosinophilic fibrous strands (Fig. 2). Small darkly staining basaloid cells and larger cells with pale nuclei in the center of the clusters and numerous scattered lymphocytes were observed, consistent with a diagnosis of ES. There was no evidence of malignant transformation.

Received September 18, 2014, Revised December 10, 2014, Accepted for publication December 15, 2014

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Table 1. Clinical features of reports cases of segmental eccrine spiradenoma

Author (year)	Age (yr)/sex	Age at onset	Location	Size (cm)	Distribution pattern	Accompanying symptom
Hashimoto et al. (1966)	15/female	9	Forearm	-	Segmental	Asymptomatic
Shelly et al. (1980)	41/female	0	Neck, submandibular	1.0～2.5	Zosteriform	Asymptomatic
Tsur et al. (1981)	35/male	9	Arm	1.0～5.0	Linea	Painful
Blanchard et al. (1981)	24/female	12	Leg	0.2～1.0	Linear	Asymptomatic
Ikeya et al. (1987)	37/female	6	Face, trunk, arm	0.1～0.8	Linear	-
Eckert et al. (1990)	16/male	-	Trunk	-	Linear and arched Blaschko's lines	Painful
Bourrat et al. (1992)	16/female	12	Arm, shoulder	-	Blaschko's lines	-
Noto et al. (1994)	16/female	0	Face, neck, trunk, leg	0.5～5	Nevoid	Painful
Criton et al. (1996)	11/male	9	Chest, extremities	0.4～1.0	Zosteriform	-
Bedlow et al.[15] (1999)	19/female	4	Chest, extremities	-	Nevoid	Asymptomatic
Guptas et al. (2000)	9/male	6	Face	0.1～1.4	Zosteriform	Asymptomatic
Guptas et al. (2001)	23/female	18	Face	0.3～1.5	Linear	Asymptomatic
Ohtsuka et al. (2002)	47/female	17	Arm	0.7～2.0	Localized	-
Braun Falco et al. (2003)	-/female	15	Leg	-	Linear	-
Altinyazar et al. (2003)	32/female	30	Leg	-	Blaschko's lines	Asymptomatic
Yoshida et al. (2004)	30/female	15	Face, neck	0.3～1.5	Zosteriform	Painful
Yoshida et al. (2004)	57/female	50	Scalp	1.0～2.0	Localized	Painless
Han et al. (2007)	24/female	12	Face, neck, chest, extremities	0.3～2.0	Zosteriform	Painless
Alfonso-Trujillo et al. (2009)	17/female	10	Leg	0.3～0.5	Zosteriform	-
Nath et al. (2009)	14/male	2	Leg	0.5～1	Linear	Painful
Yoshida et al.[2] (2010)	61/female	13	Face, neck	1.5～2.5	Linear	Painful
Englander et al.[8] (2011)	55/male	35	Chest, arm	0.5～6	Dermatomal	Painful
Gordon et al.[17] (2013)	15/female	8	Face, neck	-	Segmental	Painful

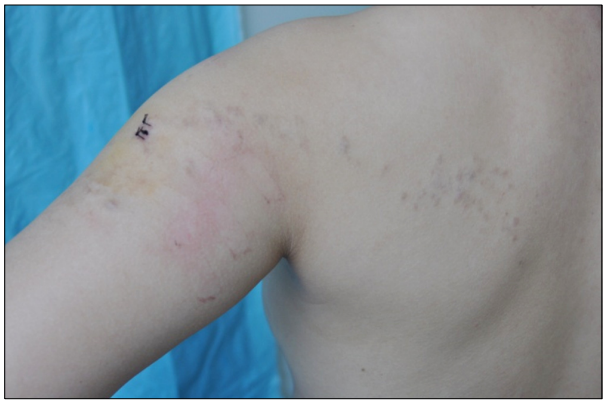

Fig. 1. Zosteriform dermal nodules on the back and forearm.

DISCUSSION

Eccrine sweat glands are simple tubular glands that open directly to the surface; they are found all over the body, especially in the palms, soles, and axillae[4]. No cases of ES have been reported on glabrous skin or strong occurrence on the trunk, and head and neck regions. ES presents as a dermal or subcutaneous papulonodules of varying size up to 6 cm.It may mimic other painful skin tumors including leiomyoma, angiolipomas, neuroma granular cell tumors, etc.[4,5]. As such, computed tomography and magnetic resonance imaging can aid the diagnosis of this kind of tumor, especially for tumors in the deep dermis or subcutaneous tissues[6,7]. However, the definitive diagnosis of ES requires skin biopsy.

The etiology of ES remains unknown. Although autosomal dominant transmission has been suggested, this has not been confirmed[2]. ES has traditionally been designated a tumor of eccrine lineage thought to be an undifferentiated or poorly differentiated benign adnexal neoplasm[8]. The proposed mechanism of multifocal ES is that an abnormal clone of multipotent stem cells of the folliculosebaceous-apocrine unit arises during embryogenesis, subsequently producing a proliferation of abnormal cells and resulting in nodule formation and possible malignant transformation[9,10]. Trauma is proposed to be aprecipitating factor[4,11], but the mechanism remains unsubstantiated. Although the present patient had a history of an injection, it is uncertain if this is coincidental or causative. Most cases of ES are solitary nodules, and multiple ES is rare: only 22 cases of multifocal segmental ES have been reported. ES exhibits no specific

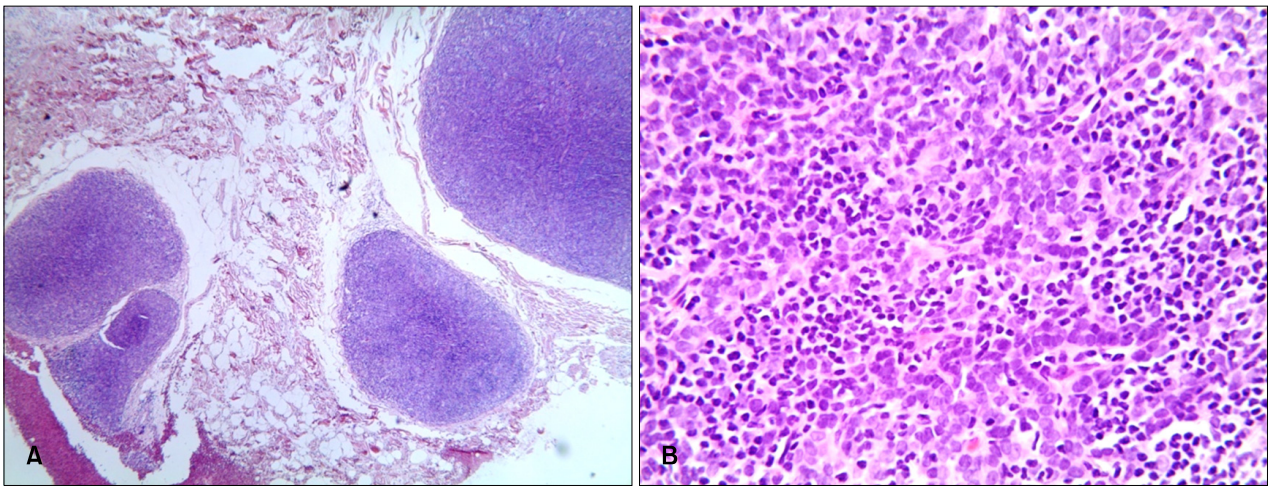

Fig. 2. (A) Multiple well-circumscribed dermal nodules comprising basophilic cells in the deep dermis. The basaloid cells are arranged in a trabecular pattern with eosinophilic fibrous strands (H&E, ×40). (B) High-powered view demonstrating small dark basaloid cells with hyperchromatic nuclei, larger cells with pale nuclei in the center of the clusters, and numerous lymphocytes scattered throughout (H&E, ×400).

characteristic location, but the literature shows that most tumors are distributed on the front of the body and above the waistline (Table 1). The tumors 0.1～6.0 cm, and the age of onset ranges from 0～61 years. The most significant clinical characteristic of the nodules is pain or tenderness in 91% of patients, which occurred intermittently and was never continuous[12]; however, Mambo[13] reports that pain and tenderness are less common. The cause of pain in ES is unclear. Moreover, the reasons for the various presentations of ES are unclear. The present patient exhibited multiple itchy ES nodules arranged in a zosteriform pattern; however, she had no family history of the disease.

Although most reported cases of ES are benign, long-standing benign ES can transform into malignant ES. Malignant transformation of ES is associated with aggressive behavior, a high recurrence rate, and the subsequent development of fatal metastases[14]. Although cases of benign ES with epidermal ulceration have been reported[13], epidermal ulceration is still a clinical characteristic associated with carcinoma. *De novo* malignant ES has also been reported. Malignant transformation occurs more often in multifocal ES than solitary ES[1].

Surgical excision is the predominant choice of treatment for tumor clearance; surgery is unnecessary if the lesions are not disfiguring, increasing in size and number, or painful. There are no clear recommendations for margins for standard excision[1]. However, if the tumors are multifocal or extensive, surgery is impractical; in such cases, radiotherapy or CO_2 laser should be considered as alternative treatments[15]. Ter Poorten et al.[16] treated cases with small discrete lesions by surgical resection combined with CO_2 laser ablation; the treatment was effective with symptomatic relief. Trials using intralesional botulinum toxin A and intralesional triamcinolone have been used to treat multiple ES[17]. Nevertheless, additional long-term follow-up is required to determine the optimal management strategy.

ES is a benign tumor of unclear etiology originating from the eccrine sweat glands. It may present as multifocal tumors arranged in zosteriform pattern. Pain is usually the most striking symptom, but our patient only reported itching, especially after perspiration or eating spicy food; this is probably because ES originates from eccrine sweat glands, which would be most active in ES during perspiration[16]. Surgical excision is the mainstay treatment. Nonetheless, better methods are requiredto treat this disease.

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