Syringoid Eccrine Carcinoma of the Thigh

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Syringoid eccrine carcinoma (SEC) is a rare cutaneous malignant tumor thought to be derived from eccrine sweat apparatus. It is usually present in the head, neck and trunk region, and often occurs in the fourth to seventh decades of life. A 94-year-old male patient visited our department with an 80-year history of a lesion showing a 2×2 cm sized well-demarcated round shaped erythematous to pinkish colored nodule with ulcer on his left thigh. Histological findings revealed a tumor consisted mainly of numerous small cords and nests forming luminal or tubular structures and tumor cells showing variable atypia. Some ductal structures showed tadpole appearance. On immunohistochemical staining, epithelial membrane antigen, S-100, cytokeratin 7 and carcinoembryonic antigen were reactive and Ki-67 showed less than 10% positivity. Based on these findings, the final diagnosis was made as SEC. The patient was treated with local wide excision and didn’t show any recurrence during the follow-up period of 12 months. Herein, we report a very rare case of SEC which occurred on the left thigh and discuss 10 cases of SEC presented on the extremities, including our case. (Ann Dermatol 29(6) 786 ~ 789, 2017)

-Keywords-
Sweat gland neoplasms, Syringoid eccrine carcinoma

INTRODUCTION

Syringoid eccrine carcinoma (SEC) is a very rarely diagnosed tumor. It is locally very invasive, destructive and often shows recurrences. Metastasis in the regional lymph nodes is extremely uncommon as are disseminated metastasis. It was first described as eccrine epithelioma (basal cell carcinoma with eccrine differentiation) by Freeman and Winkelmann in 1969 and SEC has been called diverse synonyms including eccrine epithelioma, malignant syringoma, sweat gland carcinoma with syringomatous features, squamous eccrine ductal carcinoma, syringomatous carcinoma, eccrine carcinoma and sclerosing sweat duct carcinoma. SEC most often present in the fourth to seventh decade of life and present as a solitary, firm nodule or plaque on the head, neck and less commonly trunk. To the best of our knowledge, while only 9 cases of SEC were located on the extremities.
CASE REPORT

A 94-year-old male patient visited our clinic with an 80-year history of a lesion on his left thigh. Physical examination revealed a 2×2 cm sized well-demarcated round shaped erythematous to pinkish nodule with an ulcerative cutaneous lesion on the left thigh (Fig. 1). Personal and family medical history was unremarkable. Routine laboratory tests were either normal or negative except elevated eosinophil level (15%) and erythrocyte sedimentation...

Fig. 2. (A) Many atypical infiltrating glands from superficial to deep dermis (H&E, ×40). (B) Some ductal structure showed tadpole appearance and were consisted of anaplastic cell with deep basophilic nucleus (H&E, ×100).

Fig. 3. Carcinoembryonic antigen (CEA) is very slightly reactive only limited to upper dermis portion. cytokeratin (CK)7, S-100 and epithelial membrane antigen (EMA) are strongly positive. (A) CEA, ×100, (B) CK7, ×100, (C) S-100, ×100, (D) EMA, ×100.
rate (36 mm/h). Histological findings from the first punch biopsy of skin lesion on the left thigh revealed many atypical infiltrating glands from superficial to deep dermis. Some ductal structures showed tadpole appearance and were consisted of anaplastic cell with deep basophilic nucleus (Fig. 2). Positive S-100, epithelial membrane antigen (EMA), cytokeratin (CK)7, focal positive carinoembryonic antigen (CEA), Ki-67 level of less than 10% and negative reactivity to the transcription factor-1, prostate-specific antigen, CK20 were noted immunohistochemically (Fig. 3). The histological diagnosis was SEC. Computed tomography scan indicated a 1.3 cm × 2 cm × 3 cm sized triangular shaped focal soft tissue lesion in subcutaneous fat layer and dermis layer of the lesion and didn’t show any metastasis to other organs. After diagnosis and evaluation of the tumor, the patient was treated with local wide excision. The histopathological results of excised specimen showed the same result of biopsy specimens. The patient didn’t show any recurrence during the follow-up period of 12 months.

**DISCUSSION**

SEC is an extremely rare malignant adnexal tumor of eccrine origin, first described by Freeman and Winkelmann in 1969 and has been described under diverse synonyms: eccrine epithelioma, malignant syringoma, sweat gland carcinoma with syringomatous features, squamoid eccrine ductal carcinoma, syringomatous carcinoma, eccrine carcinoma and sclerosing sweat duct carcinoma. Primary eccrine carcinomas are rare tumors and make up less than 0.01% of all skin cancers. Lesions of SEC often present similarly to morphoform basal cell carcinoma, with a solitary, firm nodule or plaque with fine telangiectasis on the head, neck and less commonly trunk, usually in the fourth to seventh decade of life. The current case also exhibited a coin sized solitary pinkish nodule with fine telangiectasis. To the best of our knowledge, only 10 cases of SEC presented on the extremities, including our case, have been reported (Table 1). In this series of 10 cases, there were 7 women and 3 men, ranging age at diagnosis of SEC from 42 to 94 years with an average age of 56 years. Although onset age of 3 cases is not precisely informed, the onset age ranged from 14 to 68 years. Five lesions occurred on the hand and arm, five lesions on the leg and toe.

SEC often shows locally very destructive lesions with deep invasion and perineural invasion; local recurrences are common, but metastasis are rare. Our case was also localized and didn’t show any metastasis and recurrences.

**Histologically, SEC have a variety of histological feature and show dilated tubules within dense fibrocollagenous**

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**Table 1. Cases of SEC presented on the extremities**

| Case | Site           | Age (yr) | Sex | Treatment | Immunohistochemical stains | Duration (yr) | Recurrence history |
|------|----------------|----------|-----|------------|----------------------------|----------------|-------------------|
| Evans et al. | Left hand     | 55       | F   | Excision  | CEA(+), EMA(+), S-100(±)   | 11             | 1 local recurrence & 1 metastatic recurrence |
| Malmusi and Collina | Left leg | 70       | F   | Excision  | CEA(+), S-100(+), CK AE1(+)/AE3(±), vimentin(+), GCDFP-15(–) | 2              | None               |
| Serrano et al. | Left forearm  | 53       | F   | Excision (4 times) | PAS(+), alcian blue(+) | 27             | 4 local recurrence |
| Sequeira et al. | Right forearm | 42       | F   | Excision  | PAS(+)                       | 3              | None               |
| Grady et al. | Left great toe | 22       | F   | Excision  | CEA(+), CK AE1(+)/AE3(±), S-100(+), EMA(–) | 1              | None               |
| Mehregan et al. | Palm of hand | 50       | F   | Excision  | PAS(+), alcian blue(+), aldehyde fuchsin(+) | -              | 1 local recurrence |
| Left arm  | 68            | F   | Excision  | PAS(+), alcian blue(+), aldehyde fuchsin(+) | -              | None               |
| Left thigh | 50            | M   | Excision  | PAS(+), alcian blue(+), aldehyde fuchsin(+) | -              | None               |
| Right knee | 60            | M   | Excision  | PAS(+), alcian blue(+), aldehyde fuchsin(+) | 3              | None               |
| Our case  | Left thigh    | 94       | M   | Excision  | CK7(+), EMA(+), S-100(+), CEA(±), Ki67: less than 10% positive | 80             | None               |

SEC: syringoid eccrine carcinoma, F: female, M: male, CEA: carinoembryonic antigen, EMA: epithelial membrane antigen, CK: cytokeratin, -: not described.
matrix, sometimes with a tadpole appearance, small neoplastic ducts, solid islands, cellular cords, and keratinizing and nonkeratinizing cysts\textsuperscript{14}. Cells show mild to marked nuclear atypia, usually of moderate atypia and a variable number of mitoses. SEC also presents with an infiltrative growth pattern with deep invasion and often extends into the subcutaneous tissue\textsuperscript{3}. The differential diagnosis of SEC includes cutaneous adenoïd cystic carcinoma, metastatic adenocarcinoma, sclerosing basal cell carcinoma, and syringoma\textsuperscript{15}. SEC resembles syringoma by showing ductal, cystic and comma-like epithelial components. It differs from syringoma by its cellular ity, anaplasia and deep invasiveness\textsuperscript{1}. Immunohistochemical staining of SEC has been described as quite variable, although cytokeratins, EMA and CEA are routinely noted to be positive. In our case, the immunohistochemical analysis of CEA was slightly reactive, but the immunohistochemical analysis of CK7 and EMA were strongly reactive. Treatment of choice for localized lesions is the surgical excision with clear margin. Chemotherapy and radiation therapy should be considered as a treatment option for metastatic lesions. In our case, the lesion was localized and didn’t recur after treatment with surgical excision. In nine cases of SEC presented on extremities, 3 cases showed recurrence, 1 case distant metastasis. Therefore, it is thought that SEC of extremities also shows local recurrence and distant metastasis like SEC of other sites.

We have reported a case of SEC of the thigh presenting as a coin sized round shaped erythematous to pinkish ulcerative nodule with no metastasis for an 80-year history. Although SEC usually present on the head and neck, it also can be found on the extremity. To the best of our knowledge, only 9 cases were located on the extremities.

**CONFLICTS OF INTEREST**

The authors have nothing to disclose.

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