Syringoid Eccrine Carcinoma
-A Case Report

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Syringoid eccrine carcinoma is a rare cutaneous malignant tumor thought to be derived from eccrine sweat apparatus. They may be locally aggressive and have an infiltrative growth pattern. Because of their rarity, the origin and differentiation of this tumor has not been confirmed completely. Variable clinical appearance and diverse histologic findings often makes diagnosis difficult. We report a 71-year-old female with an asymptomatic brownish nodule located on the medial side of right knee for years. Histopathology showed a dermal tumor consisted mainly of tubulocystic proliferation embedded in a fibrous stroma, invading the whole thickness of dermis extending close of the subcutaneous tissue. The ducts are lined by one or two layers of cuboidal cells showing variable atypia and nuclear pleomorphism. Immunoperoxidase stains with epithelial membrane antigen (EMA), carcinoembryonic antigen (CEA), CK 7 and S100 protein were all positive. Such histopathologic features and immunohistochemical profile suggest that a syringoid eccrine carcinoma. The tumor was excised completely with free margin and systemic survey did not reveal visceral involvement. (Dermatol Sinica 25: 124-127, 2007)

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INTRODUCTION

Primary eccrine carcinomas are uncommon and account for less than 0.01% of all skin cancers. Because of their rarity, clinical appearance is not well characterized and their biological behavior is not clearly defined. Limited experience with these neoplasms often lead to difficulty with differentiation from their benign counterparts, from other cutaneous neoplasms, and from adenocarcinomas with skin metastases. We report a case of syringoid eccrine carcinoma occurring in a 71-year-old woman.

CASE REPORT

A 71-year-old Chinese female was examined for an asymptomatic brownish nodule located on medial side of right knee. (Fig. 1) The lesions were noted for years with slow gradual growth. Physical examination revealed a hyperpigmented, non-tender, indurated nodule about 1cm in diameter. Patient denied local trauma history and no regional lymph nodes were palpated. Under the initial impression of dermatofibroma, patient underwent total excision of the tumor and the specimen was sent for histologic examination. Pathology revealed that the tumor consisted mainly of numerous small cords and nests from the reticular dermis extending close to subcutaneous tissue. (Fig. 2a) Most of the tumor nests and cords formed luminal or tubular structures and the tumor cells had relatively uniform round nuclei with some showing variable atypia. (Fig. 2b) Multiple sections were done and neither keratinous cyst nor follicular differentiation was noted. Immunohistochemical analysis of the tumor was also performed. Immunoperoxidase stains with epithelial membrane antigen (EMA), carcinoembryonic antigen (CEA), Cytokeratin 7 and S100 protein were all positive. (Fig. 3) Syringoid eccrine carcinoma was diagnosed and extensive survey for other malignancy or possible metastasis was arranged. Routine laboratory examination, abdominal echogram, chest x-ray, and Ga-citrate
scintigram showed no remarkable findings suggesting visceral malignancy. Subsequent follow up did not reveal any clinically obvious local recurrence or distant metastases.

**DISCUSSION**

Syringoid eccrine carcinomas (SEC) are relatively rare adnexal neoplasms and only a dozen of cases have been reported. It was first described by Freeman in 1969 as a basal cell tumor with eccrine differentiation. Other synonyms also have been used previously, including malignant syringoma, sweat gland carcinoma with syringomatous features, adenocarcinoma of eccrine glands, and malignant chondroid syringoma. The majority of current literature favors to use the term syringoid eccrine carcinoma for describing this distinctive neoplasm.

The clinical appearance of SEC is not specific; they may present as slow growing solitary painless nodules or indurated plaques that often found on the head or extremities. These tumors are usually indolent, existing grossly several years before diagnosis. While SEC may be locally destructive with frequent recurrence, metastases are seldom reported; however, few cases have been described with lymph node metastases.

Histologically, SEC varies from well-differentiated glandular lesions to anaplastic neoplasms with little evidence of eccrine differentiation. SEC is characterized by a tubulocystic proliferation set in a fibrovascular stroma, invading the deep dermis and, in some cases, the subcutaneous tissue. Mitotic figures may be seen in the cells surrounding the ducts, which are lined by one or two layers of cuboidal cells. Diagnosis of malignancy is based on histologic and cytologic features including signs of invasion of surrounding tissues, the presence of perineural infiltration, mitotic activity, and nuclear pleomorphism. In our case, the tumor infiltrates the subcutis closely but vessel or perineural invasion was not noted.

SECs may need to be differentiated from basal cell carcinoma (BCC), primary cutaneous adenoid cystic carcinoma (PCACC), microcystic adenoid carcinoma MAC, and visceral adenocarcinoma with skin metastases. When histologic examination does not establish the diagnosis, immunohistochemical techniques may be done for further differentiation of the tumors.

SECs may sometimes be confused with BCCs. Histologically, SEC may resembles an adenoid basal cell carcinoma, but CEA is expressed in sweat gland neoplasms and is lacking in BCC.

Similarities with other variants of eccrine carcinomas can be confusing as they often have comparable immunohistochemical features. SEC may need to be differentiated from MAC. MAC contains both area of eccrine and follicular differentiation and are composed of nest and strands of basaloid cells with keratinous cysts. Unlike MAC, SEC do not usually form keratinous cysts and lack follicular component. SEC may also be confused with PCACC, which is another rare neoplasm that may arise from eccrine ducts but most often arises from the intercalated ducts of salivary glands. PCACC may histologically be similar to SEC but it usually shows evidence of mucin accumulation while SEC does not.

It may be difficult to differentiate SEC from cutaneous metastasis of adenocarcinomas by histologic features alone as immunohistochemical stains can not provide useful data in distinguishing primary eccrine carcinomas from metastases of visceral neoplasms. However, lack of CK20 expression may be of diagnostic help since colorectal carcinomas often expressed CK20. In our case, metastatic carcinoma from the extracutaneous organs was ruled out by clinical and instrumental examinations including various imaging techniques. CK20 stain is also performed and the result is negative, which further supports our case as primary eccrine carcinoma.

Most authors currently believe that SEC is of an eccrine nature, although there have been few reports suggesting apocrine differentiation. However, several authors have shown the lack of expression of apocrine markers HMFG and gross cystic disease fluid protein 15.
(GCDFP-15) in their cases respectively, weakening the support for apocrine differentiation. In our case, there are hints of eccrine features, such as absence of horn cysts, no connection with the epidermis, ducts lined by a single layer of cells, and a compatible immunohistochemical profile.

In summary, our case exhibited the histologic and immunocytochemical characteristics of SEC. It has been suggested that primary eccrine carcinomas are histopathologic entities with a spectrum of differentiation in which SEC is between the well differentiated MAC and the poorly differentiated PCACC. However consensus has not been reached and further unifying of classification of eccrine carcinomas is still needed. Complete excision was done with histologic free margin and successive follow-up did not showed local recurrence. Some authors have suggested wide local excisions as the primary treatment of eccrine neoplasms, while others prefer the Mohs microsurgery as a safer and more effective alternative. In all cases, follow ups should always be arranged for surveillance of future recurrence. We hereby present this distinctive sweat gland carcinoma and points out primary adnexal carcinomas such as SEC should be placed in the list of differential diagnosis for solitary nodule or plaque on head and extremities.

**REFERENCES**