Syringoid Eccrine Carcinoma: A Case Report and Review of the Literature

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Abstract
Syringoid eccrine carcinoma is a rare sweat gland carcinoma that can be difficult to diagnose. The clinical and histologic appearance is often nonspecific. Therefore, immunohistochemistry is often helpful with making the diagnosis. We report a case of syringoid eccrine carcinoma of the scalp and review the current literature.

Introduction
Syringoid eccrine carcinoma (SEC) is a rare, malignant adnexal tumor that can be challenging to diagnose both clinically and histologically. Clinically, the tumor has a nonspecific appearance and can often resemble basal cell carcinoma. Histologically, differentiation from other benign and malignant tumors can be difficult. Immunohistochemistry can be helpful in helping differentiate SEC from other neoplasms and adencarcinomas with skin metastases. We report a case of SEC that clinically presented and was treated as basal cell carcinoma and review the current literature.

Case Report
A 67-year-old white male presented with a several-year history of an enlarging, tender lesion on the posterior scalp. Physical examination revealed a pink, pearly, well-demarcated papule with overlying telangiectasia measuring 7 mm x 5 mm in diameter (Figure 1). No detectable lymphadenopathy was present upon examination. The lesion was clinically suspicious for basal cell carcinoma and was scheduled for surgical excision. The lesion was excised with 4 mm margins and sent for histologic examination.

Examination of the hematoxylin-and-eosin (H&E) stained specimen showed a small, well-circumscribed neoplasm of ductal structures with an infiltrating growth pattern surrounded by a desmoplastic stroma (Figures 2, 3). The tumor extended into the reticular dermis. Scattered mitotic figures were present. No evidence of perineural invasion was seen in this case. The tumor extended to the lateral tissue edges. Immunohistochemical analysis was performed, and there was found to be CEA, EMA, CK7 and p63 positivity.

Due to the positive margins, the patient was sent to plastic surgery for frozen section procedure. The tumor was excised with clear margins, and no signs of recurrence were noted at one-month follow-up.

Discussion
SEC is a rare type of sweat gland carcinoma originally described as basal cell carcinoma with eccrine differentiation (eccrine epithelioma) by Freeman and Winkelmann in 1969.1,1 There are various synonyms for and variations of this tumor, including syringomatous carcinoma, malignant syringoma, squamoid eccrine ductal carcinoma, sclerosing sweat duct carcinoma, sweat gland carcinoma with syringomatous features and, as previously mentioned, eccrine epithelioma.1,2

The clinical presentation of SEC is nonspecific and can vary, but most commonly the tumor presents as a solitary, firm nodule or plaque on the scalp that is sometimes painful. Ulceration is uncommon. SEC occurs mostly in the fifth and sixth decades of life and affects males and females equally. The tumor is slow-growing and locally aggressive with deep invasion. Multiple local recurrences are common, yet metastases are rare. Most reported cases of metastasis involve lymph nodes, with rare reports of metastasis to lung and bone.1,3,7,12

Histologically, SEC has a tadpole-like morphology composed of a basaloïd cell infiltrate with ductal differentiation surrounded by a dense fibrous stroma. Small epithelial cells are present with hyperchromatic nuclei, pale cytoplasm and indistinct cell membranes arranged in narrow cords. The tumor is deeply invasive, often extending into the subcutaneous tissue and muscle. Cytologic atypia and mitotic activity are variable. Perineural invasion is common and likely contributes to the tendency of local recurrence.1,3,4

The immunohistochemistry of SEC tumors is nonspecific and variable. Simple epithelial cytokeratins (CKs 7, 8, 18, 19) are expressed by most tumor cells, and a small number express stratified epithelial cytokeratins (CKs 5, 14).1,3 Tumor cells also commonly express EMA and CEA and occasionally express S-100 protein. Studies also demonstrate that the majority of primary adnexal tumors strongly express p63, as in our case. Other antigens reported to be positive in SEC include Ber-EP4, ER and PR.1,3,6

SEC can be difficult to differentiate from a variety of other tumors including syringoma, basal cell carcinoma, microcystic adnexal carcinoma (MAC), primary cutaneous adnexoid carcinoma (PCACC), and visceral adencarcinomas with skin metastases.1,3 Syringomas lack the cellularity, deep invasiveness and anaplasia that SEC demonstrates.3,5 SEC differs from basal cell carcinoma by the lack of retraction artifact, characteristic palisading arrangement, and by the presence of EMA and CEA positivity. Basal cell carcinoma rarely shows ductal differentiation as seen in SEC.1,3 MAC displays eccrine and follicular differentiation and is composed of nests and strands of basaloïd cells forming keratin-filled cysts, which are not present in SEC. SEC differs from PCACC in that it lacks the prominent cribriform pattern of tumor growth and mucin production demonstrated by PCACC. SEC and PCACC are similar immunohistochemically.1,2

Tumor morphology and immunohistochemistry

![Figure 1](image1.jpg)

![Figure 2](image2.jpg)

![Figure 3](image3.jpg)
distinguish SEC from skin metastases due to visceral adenocarcinomas, such as breast, lung and kidney. Immunohistochemical markers that can help differentiate these tumors from SEC include mammoglobin and gross cystic disease fluid protein for breast carcinoma, thyroid transcription factor 1 for lung carcinoma and CD10 and renal cell carcinoma marker for renal cell carcinoma.¹

Surgical excision with clear margins is considered the treatment of choice for localized SEC. Mohs’ micrographic surgery has been considered the method of choice for localized lesions if there are no “skip” areas or evidence of multi-focality.²⁻⁴ In our case, Mohs’ micrographic surgery was not an available option, and frozen section procedure was performed to ensure clear margins. Chemotherapy and radiation therapy have been used for metastatic sweat gland carcinomas with variable results.²⁻⁴ As previously mentioned, local recurrence is common, and approximately 40% to 60% of reported cases had one or more local recurrences within six months to 30 years after treatment with standard wide local excision.⁵

Conclusion
In conclusion, SEC can demonstrate variability in both clinical and histologic appearance. Immunohistochemistry, therefore, is crucial in differentiating SEC from other neoplasms. Due to the locally aggressive nature of the tumor, recurrence is common, though metastasis is rare. Excision with clear margins is the treatment of choice, and good results have been achieved by Mohs’ micrographic surgery.

References

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