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Case Presentation

Syringocystadenocarcinoma papilliferum: a rare tumor with a favorable prognosis

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Abstract

Syringocystadenocarcinoma papilliferum (SCACP) is a rare adenexal carcinoma with only 21 cases reported in the literature. Most patients describe a long-standing mass with recent change, supporting the idea that SCACP arises from malignant transformation of pre-existing syringocystadenoma papilliferum (SCAP). Syringocystadenocarcinoma papilliferum is generally treated with wide surgical excision of the lesion and patients do exceeding well and require no systemic therapy.

Case synopsis

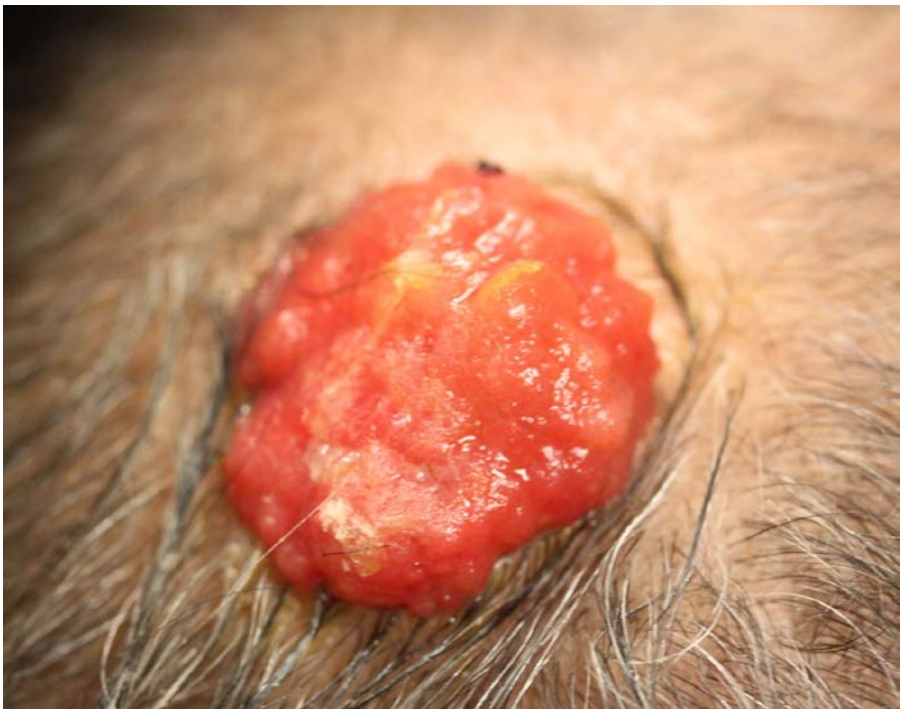

Figure 1. A 65 year-old Hispanic male presented with a flesh-colored, exophytic 3x3 cm tumor on the right posterior scalp with serosanguinous exudate.

A 65-year-old Hispanic male presented with a hairless, exophytic tumor on his scalp that had been present since birth. The patient reported that the lesion had been rapidly growing over the past year. The patient's past medical history was significant for type 2 diabetes and a comprehensive review of systems was unremarkable. Physical exam demonstrated a flesh-colored, exophytic 3x3 cm tumor on the right posterior scalp with serosanguinous exudate.

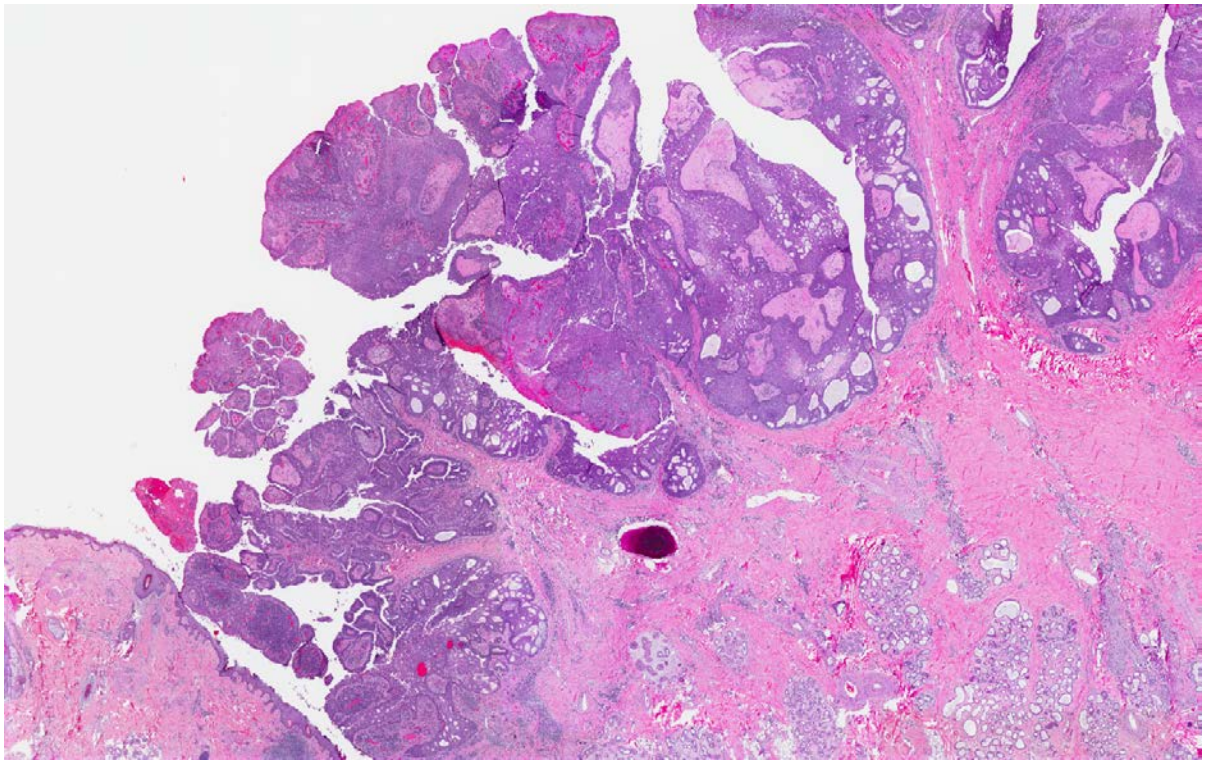

Figure 2. Excisional biopsy demonstrates an ulcerated exoendophytic neoplasm with prominent surface papillations lined by squamous epithelium superficially and a bilayer of secretory epithelium in the mid and deep portions (Hematoxylin & Eosin, 10x magnification)

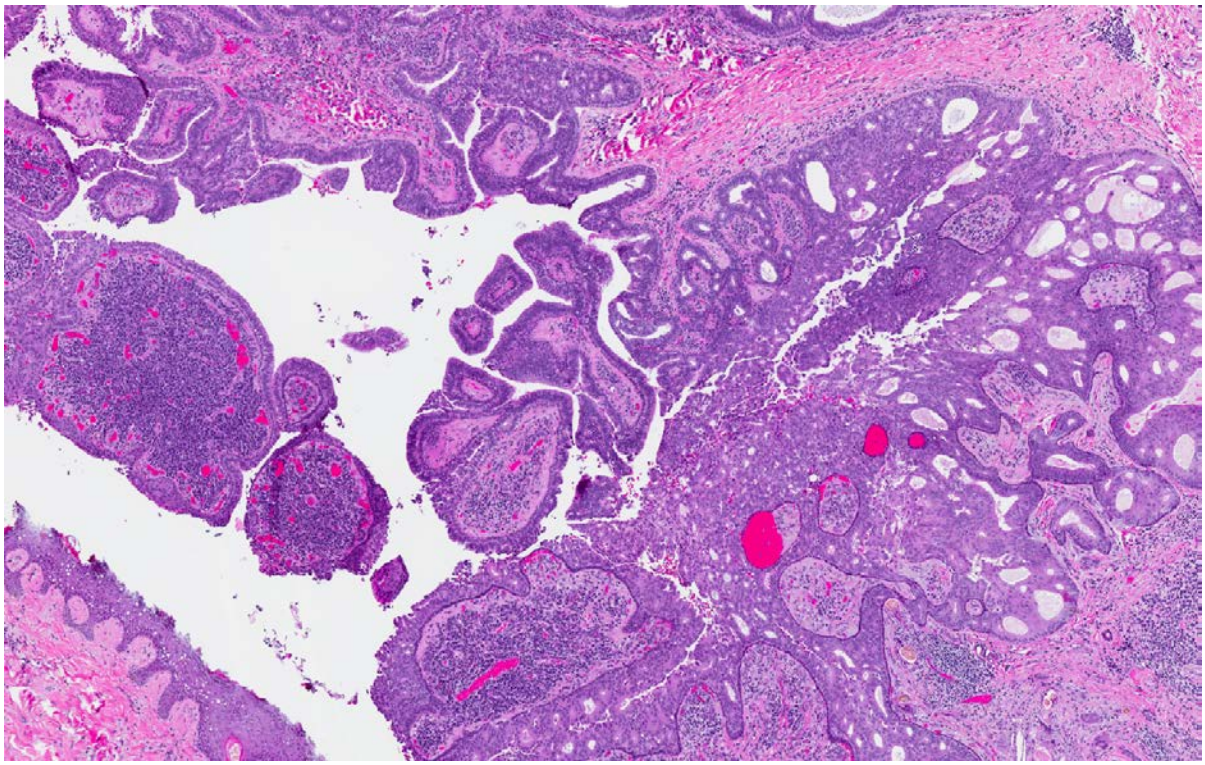

Figure 3. Excisional biopsy at 40x magnification accentuates the secretory epithelium consisting of an inner lining of columnar epithelium and a peripheral basal/myoepithelial cuboidal cell layer. (Hematoxylin & Eosin, 40x magnification)

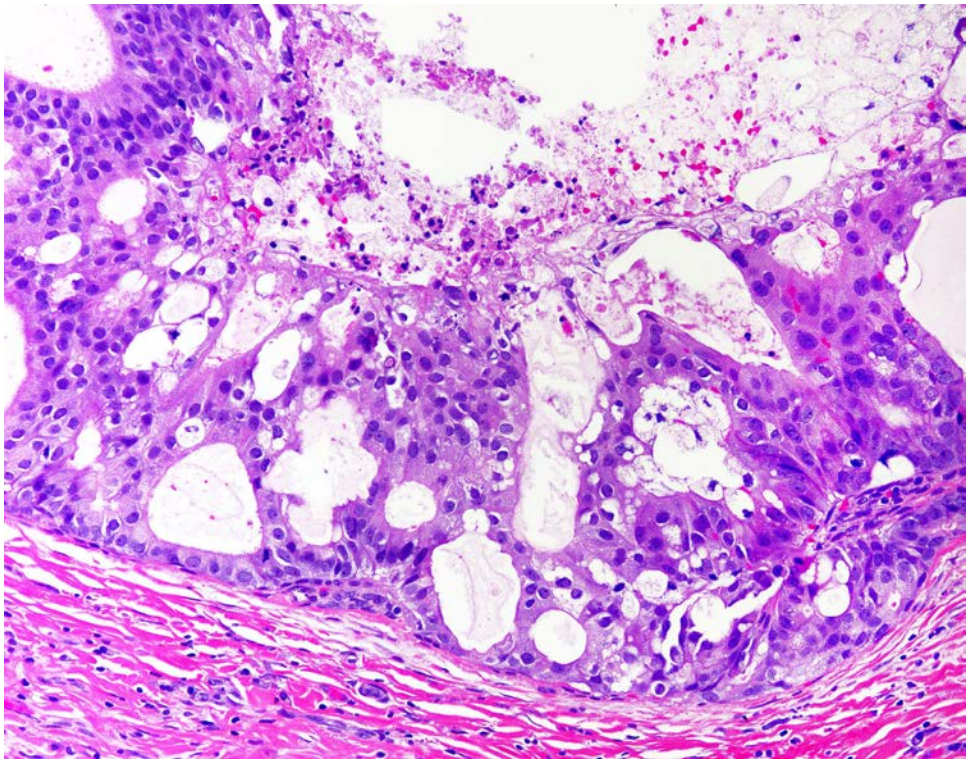

Figure 4. Excisional biopsy at 100x magnification demonstrates stroma containing infiltrative plasma cells and lymphocytes. There was pleomorphism and enlargement of the secretory epithelium in the deep aspect of the tumor, with formation of cribriform back-to-back glands with necrosis. (Hematoxylin & Eosin, 100x magnification)

Skin excision demonstrated an ulcerated exoendophytic neoplasm with prominent surface papillations lined by squamous epithelium superficially and a bilayer of secretory epithelium in the mid and deep portions. The secretory epithelium consisted of an inner lining of columnar epithelium and a peripheral basal/myoepithelial cuboidal cell layer. The stroma contained infiltrative plasma cells and lymphocytes. There was pleomorphism and enlargement of the secretory epithelium in the deep aspect of the tumor, with formation of cribriform back-to-back glands with necrosis. The basal layer was highlighted with cytokeratin 14 and p63. Rare foci of invasive adenocarcinoma were identified. We diagnosed syringocystadenocarcinoma papilliferum (SCACP) arising in a background of benign syringocystadenoma papilliferum (SCAP). The tumor was completely excised with 5 mm margins and the patient is alive with no disease recurrence.

Discussion

Including our case, the average age at diagnosis is 66 years and the male to female ratio is equal [1,2,3]. SCACP usually arises from longstanding benign SCAP, but has also been described within nevus sebaceus and linear nevus verrucosus [4].

SCACP is found on the head and neck but has also been found in the suprapubic region, arm, and calf [5]. Lesions range from 1.5 cm to 13 cm in size and present as exophytic papillated nodules associated with ulceration and pain that often have oozing from glandular secretions [2,5].

Although clinically SCACP can resemble other common types of skin cancer, the histopathologic differential is less broad. Hidradenocarcinoma papilliferum, apocrine ductal adenocarcinoma, and cutaneous metastases of visceral and breast carcinoma can resemble SCACP microscopically [2]. Hidradenocarcinoma papilliferum and apocrine ductal adenocarcinoma are dermal neoplasms that do not show an epidermal connection. SCACP is often positive for carcinoembryonic antigen, gross cystic disease fluid, and p63. Additionally, p63 expression favors a primary sweat gland neoplasm of the skin rather than a cutaneous metastasis of a visceral adenocarcinoma. [3]

Only three cases of SCACP have demonstrated lymphatic spread [2,3]. There was one putative case of SCACP from 1949 in which the patient died owing to disseminated disease, but because there were no histopathologic illustrations, confirmation is not possible [2]. No other cases of distant metastasis or death have been reported [1,2,3]. Treatment consists of surgical removal of the lesion with wide excision. Some authors advocate sentinel lymph node sampling, although because of the rarity of SCACP, general guidelines have not been established [3].

Conclusion

Our patient represents the typical presentation of SCACP with a long-standing tumor that showed recent rapid growth. Although SCACP is exceedingly rare, it can mimic other more common and serious forms of cancer. The unique histopathological features are key in the accurate diagnosis of SCACP. Fortunately, SCACP does not often metastasize and can be successfully treated with surgical excision alone.

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Acknowledgments

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