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Case Presentation

Syringocystadenocarcinoma papilliferum with locoregional metastases

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Abstract

Syringocystadenocarcinoma papilliferum (SCACP) is a rare malignant counterpart of syringocystadenoma papilliferum (SCAP). To date, less than 30 cases have been reported, and of those, only three had locoregional metastases [1,2]. Herein, we report a case of a 42-year-old man with a rapidly growing nodule on his right parietal scalp ultimately diagnosed as SCACP arising in association with a preexisting SCAP and nevus sebaceus. This case differs from prior reports in the tumor's rapid growth and aggressive course with the development of locoregional metastases within 5-weeks from initial presentation.

Case Presentation

A 42-year-old man presented with a 4-week history of a rapidly enlarging scalp mass that bled with minor trauma. He denied a history of a hairless plaque in this region, but stated that there may have been a smaller preexisting "lump" of uncertain duration. On exam, there was a 4.5 x 4 cm focally ulcerated exophytic nodule associated with a few small satellite papules on the right parietal scalp (Figure 1). No cervical lymphadenopathy was identified.



Figure 1. A 4.5 x 4 cm exophytic focally ulcerated nodule on the right parietal scalp.

An incisional biopsy at the periphery of the lesion showed epidermal acanthosis and papillomatosis accompanied by increased sebaceous glands that were unassociated with hair follicles, consistent with a nevus sebaceous. Additionally, there were duct-like invaginations that emanated from the epidermis associated with dermal cystic spaces containing papillary projections surrounded by a dense lymphoplasmacytic infiltrate, below which were dilated apocrine glands consistent with a SCAP (Figure 2). Since the biopsy was superficial and only a sample of a larger lesion, complete excision was recommended.

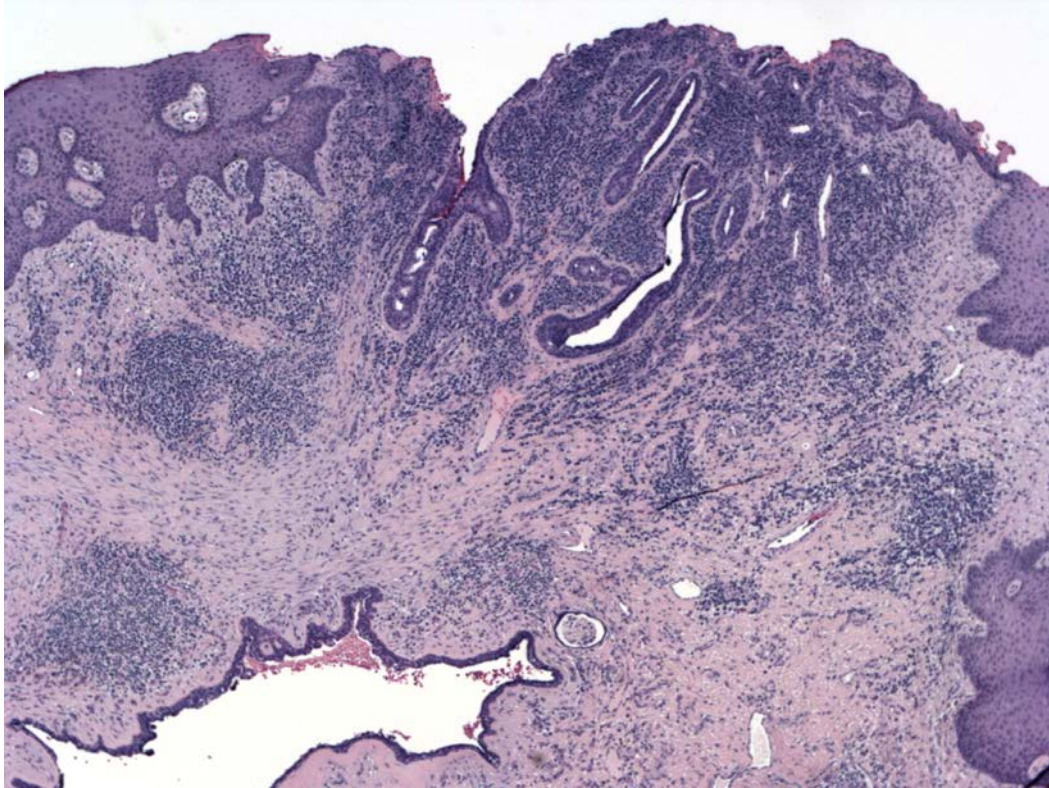


Figure 2. Initial Biopsy. Duct-like invaginations associated with a dense lymphoplasmacytic infiltrate (original magnification x 40).

Excisional biopsy showed focal residual SCAP adjacent to which was a dense proliferation of back-to-back glands admixed with solid islands and cords of markedly atypical cells containing numerous mitoses (Figure 3). Additionally noted were large dilated apocrine ducts lined by multilayers of pleomorphic epithelial cells, focal squamous differentiation, and areas of adenocarcinoma in situ (Figure 4). The tumor infiltrated the subcutaneous tissue, showed focal necrosis en masse, and exhibited perineural involvement and intravascular invasion.

Wide local excision (WLE) and sentinel node biopsy was planned. However, on the preoperative exam three discrete enlarged cervical lymph nodes were palpated. Fine needle aspirate confirmed the presence of malignant cells. Computed and positron emission tomography showed numerous enlarged lymph nodes, but did not identify distant metastases.

The patient underwent WLE and right posterior neck dissection. No residual tumor was identified, but 11/ 50 lymph nodes in the right neck at levels 2B and 5 were diffusely replaced by tumor. There was no evidence of recurrence or metastatic disease two months post-surgical intervention.

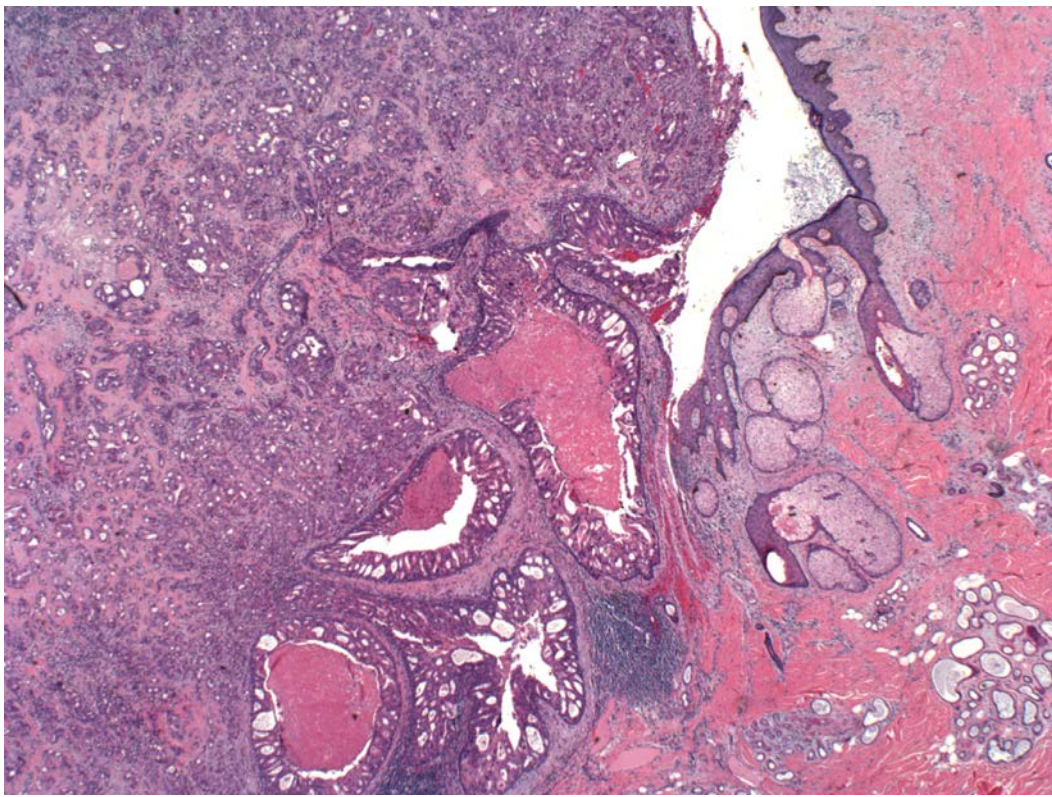


Figure 3. Excision. On the right side of the photomicrograph there is a back-to-back glandular proliferation of markedly atypical cells adjacent to a typical nevus sebaceous (original magnification x 20).

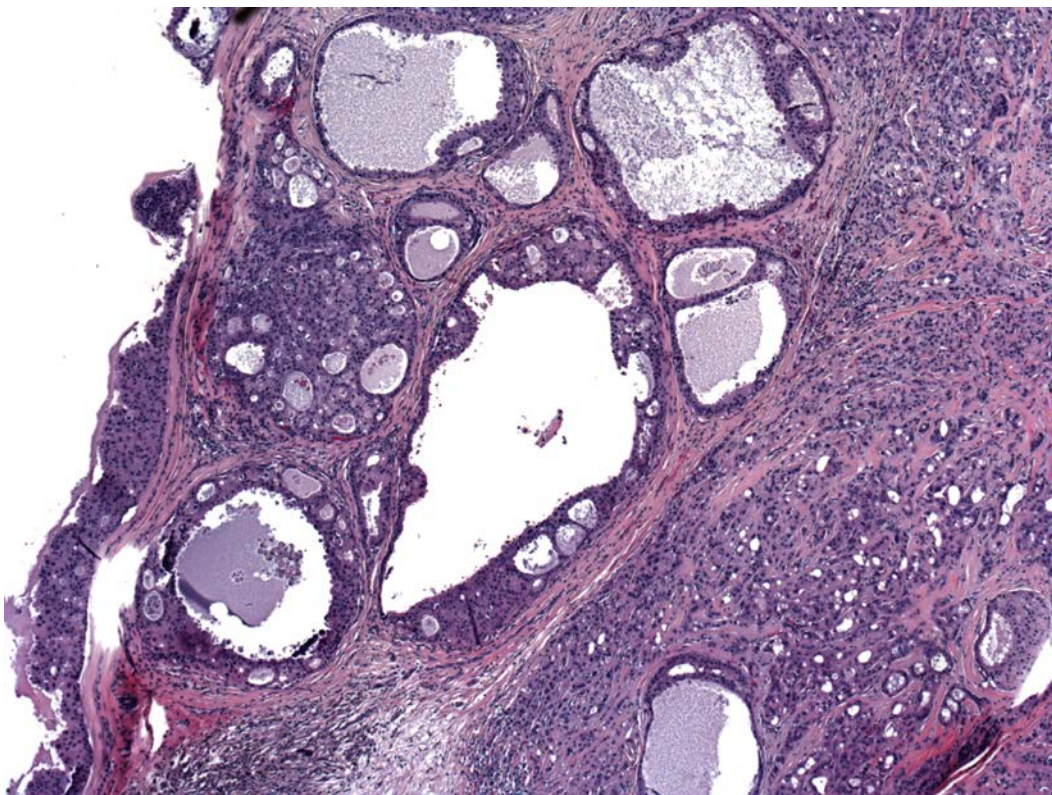


Figure 4. Higher power view of excision. Large dilated apocrine ducts lined by multilayers of pleomorphic epithelial cells admixed with back-to-back glands and cords of atypical cells (original magnification x 100).

Discussion

Syringocystadenocarcinoma papilliferum is a rare apocrine gland carcinoma often arising in association with a pre-existing SCAP and/or nevus sebaceous. To date, less than 30 cases have been reported, and of those, only three had locoregional metastases [1,2]. The majority of cases are located in the head and neck region, or less commonly the perineal area and extremities of middle aged to elderly individuals, median age of 65 [2-5]. Clinically, SCACP presents as an enlarging flesh-colored to hyperpigmented

exophytic nodule ranging in size from 0.5-13cm, which may be associated with pain, discharge, and/or ulceration [3,4]. Duration is variable, ranging from months to at least 30 years [3-5].

Although the majority of lesions consist of invasive adenocarcinoma, SCACP can exhibit morphologic diversity, with some tumors showing an admixture of invasive squamous cell carcinoma and/or syringocystadenocarcinoma papilliferum in situ, often with pagetoid spread [2-5]. Rarely the lesion has been reported to additionally contain a sebaceoma, sebaceous carcinoma, ductal carcinoma in situ, or a focal area resembling lymphoepithelioma-like carcinoma [3-5]. The histological features can be diagnostically challenging and evoke a differential diagnosis that includes metastatic adenocarcinomas arising from the breast, ovary, endometrium, thyroid, and gastrointestinal tract [3-5].

There are no clinical or histological features to assist in determining prognosis, but in general the tumor has low-grade malignant potential. Recurrence is rare with only two cases reported [2,3]. To date, there have only been four cases, inclusive of the one presented, that had locoregional metastases and there have been no reports of distant metastases [1-2].

Owing to its rarity no standard treatment protocol other than WLE exists. However, recently Mohs surgery has been advocated as an alternative [6]. Adjuvant radiation therapy has been tried in one case, but the patient subsequently developed locoregional metastases [2].

In conclusion, we report a case of SCACP that arose in association with a pre-existing SCAP within a nevus sebaceous. To the best of our knowledge this is the youngest case in the literature. The tumor differed from prior reports in its rapid growth and subsequent development of palpable lymphadenopathy within 5-weeks of initial presentation. Because of the aggressive nature of this patient's tumor, adjunctive radiation was considered. Unfortunately, the patient has been lost to follow-up.

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