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Case Presentation

Adenoid Cystic Carcinoma of the Base of the Tongue Metastasizing to the Scalp

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Abstract

Adenoid cystic carcinoma is a rare neoplasm that originates from secretory glands, most commonly from the salivary glands. We present a 76 year-old white man with a history of adenoid cystic carcinoma from the base of the tongue diagnosed 15 years prior to the development of the metastatic lesion on his mid-posterior scalp. The present case represents the second reported instance of an extracutaneous adenoid cystic carcinoma metastasizing to the scalp. Differentiating between a primary cutaneous adenoid cystic carcinoma and an extracutaneous adenoid cystic carcinoma metastasizing to cutaneous structures is crucial in determining prognosis and management.

Keywords: adenoid cystic carcinoma; cutaneous metastasis; scalp; tongue; salivary glands

Introduction

Adenoid cystic carcinoma (ACC) is a rare neoplasm that originates from secretory glands and comprises a large portion of malignant neoplasms originating in both major and minor salivary glands. Origin from other sites, such as excretory glands of the genital tract, lacrimal glands, and ceruminous glands, has been documented [1]. Adenoid cystic carcinoma is characteristically associated with a prolonged clinical course and slow, progressive growth. It tends to recur locally and can metastasize regionally and systemically years after the initial diagnosis. Approximately 35-50% of salivary gland ACCs metastasize distantly 3.1 to 7.6 years after diagnosis. The lungs are the most common sites of distant metastasis. Bone, liver, and brain are also common sites [2, 3]. Adenoid cystic carcinoma is generally treated by surgical excision with attention to margins and the presence of perineural invasion. This is typically followed by adjuvant external radiation of the surgical bed. Results from studies evaluating the efficacy of chemotherapy in the treatment of ACC remain inconclusive [1].

Primary cutaneous ACC is best classified as a neoplasm with sweat gland differentiation. These neoplasms can occur anywhere on the body, but have a predilection for the scalp and chest [4, 5]. Wide local excision with free margins is the treatment choice [6], and Mohs surgery is frequently employed [7]. Cutaneous ACC is locally invasive and 76% of the cases show perineural invasion, resulting in local recurrence in 44% of the cases [5, 7]. Primary cutaneous ACC, however, is less aggressive than salivary gland ACC, and distant metastases, typically to lung, are rare. Cases with metastatic spread to the pleura and lymph nodes have also

been reported. Metastases have been diagnosed from 8.2 to 16 years after diagnosis of the primary tumor [4-6]. Even though primary cutaneous ACC has a predilection for the scalp and chest, to the authors' knowledge, only one case of metastases from extracutaneous ACCs to the scalp has been reported [8].

Methods

A search of PubMed performed on November 2, 2013, with the following terms – "adenoid," "cystic," "carcinoma," "cutaneous," "metastasis," "metastases," "scalp" yielded only one report of metastases to the scalp from a primary ACC of lacrimal gland origin.

Case synopsis

A 76 year-old white man presented to the dermatology clinic with a firm, pink papule on the mid-posterior scalp that had been slowly growing for an indeterminate period of time (Figure 1). He had a history of non-melanoma skin cancer (NMSC) and right base of tongue ACC with pulmonary metastases. The primary ACC was diagnosed 15 years prior to this visit and the tumor had peri- and intraneural invasion (Figure 2). The initial treatment was surgical excision, followed one month later by a re-excision to obtain negative margins. He received no further treatment and was followed by his oncologist. A computed tomographic (CT) scan of the thorax with contrast, performed 2 years prior to the current date, revealed diffuse pulmonary metastases, bilaterally, confirmed by biopsy. The biopsied metastatic lesion was diffusely positive for CD117 (c-kit). A new thoracic CT scan showed that one of the metastases was possibly increasing in size and that the other nodules were relatively stable. The patient deferred oncologic treatment, but agreed to be monitored via thoracic CT scans. During his visit to the dermatology clinic for an unrelated reason, a firm, pink papule was discovered on the mid-posterior scalp and a shave biopsy was performed

Upon histopathology examination (Figures 3A and 3B), the lesion resembled a basal cell carcinoma, but a closer inspection revealed that the neoplasm formed multiple ductal structures expressing carcinoembryonic antigen (CEA), cytokeratin (CK) 8/18 (Figure 3C), epithelial membrane antigen (EMA) (Figure 3D), and pankeratin AE1/AE3. No connection to the epidermis was identified. At the time of this diagnosis, the pathologist was unaware of the patient's clinical history and the diagnosis considered was primary ACC of sweat gland differentiation. Once the pathologist learned about the patient's history of base of tongue ACC, the current tumor was compared to the tumor from the patient's previous excision. The histopathologic features of the original tumor and the scalp metastasis were almost identical, except that the metastasis lacked intraluminal globules of basement membrane material. Accordingly, a diagnosis of metastatic ACC was made.

The tumor measured 0.8 cm in greatest dimension and was excised with 0.4 cm negative margins. After the patient had a discussion with his oncologist, he decided that he wanted to defer workup for further metastatic lesions pending the results of future repeat CT scans.

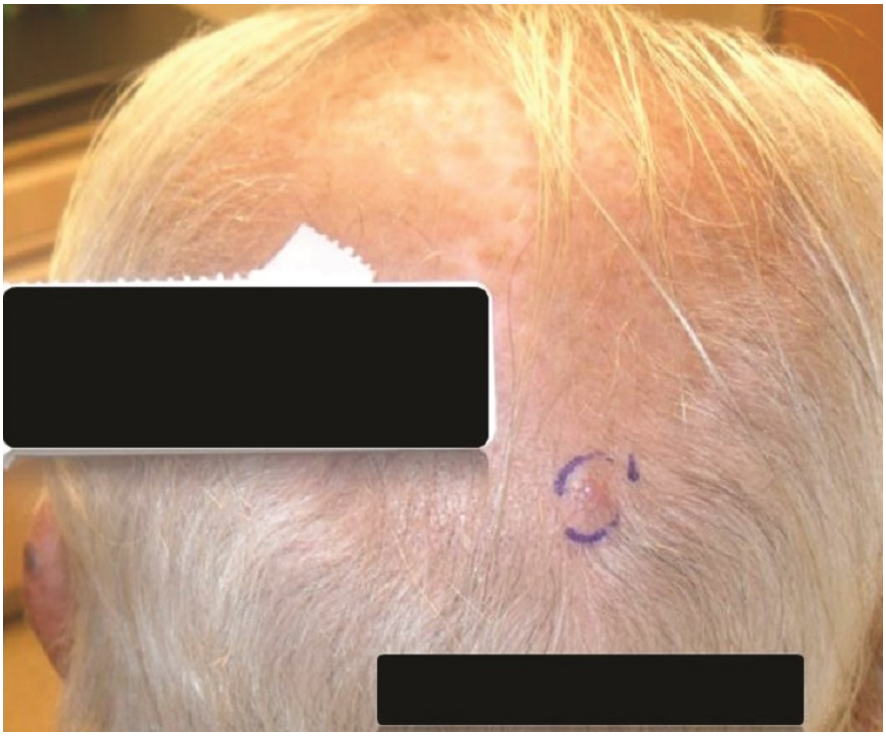

Figure 1. A 5 mm erythematous, slightly scaly papule on the mid-posterior scalp.

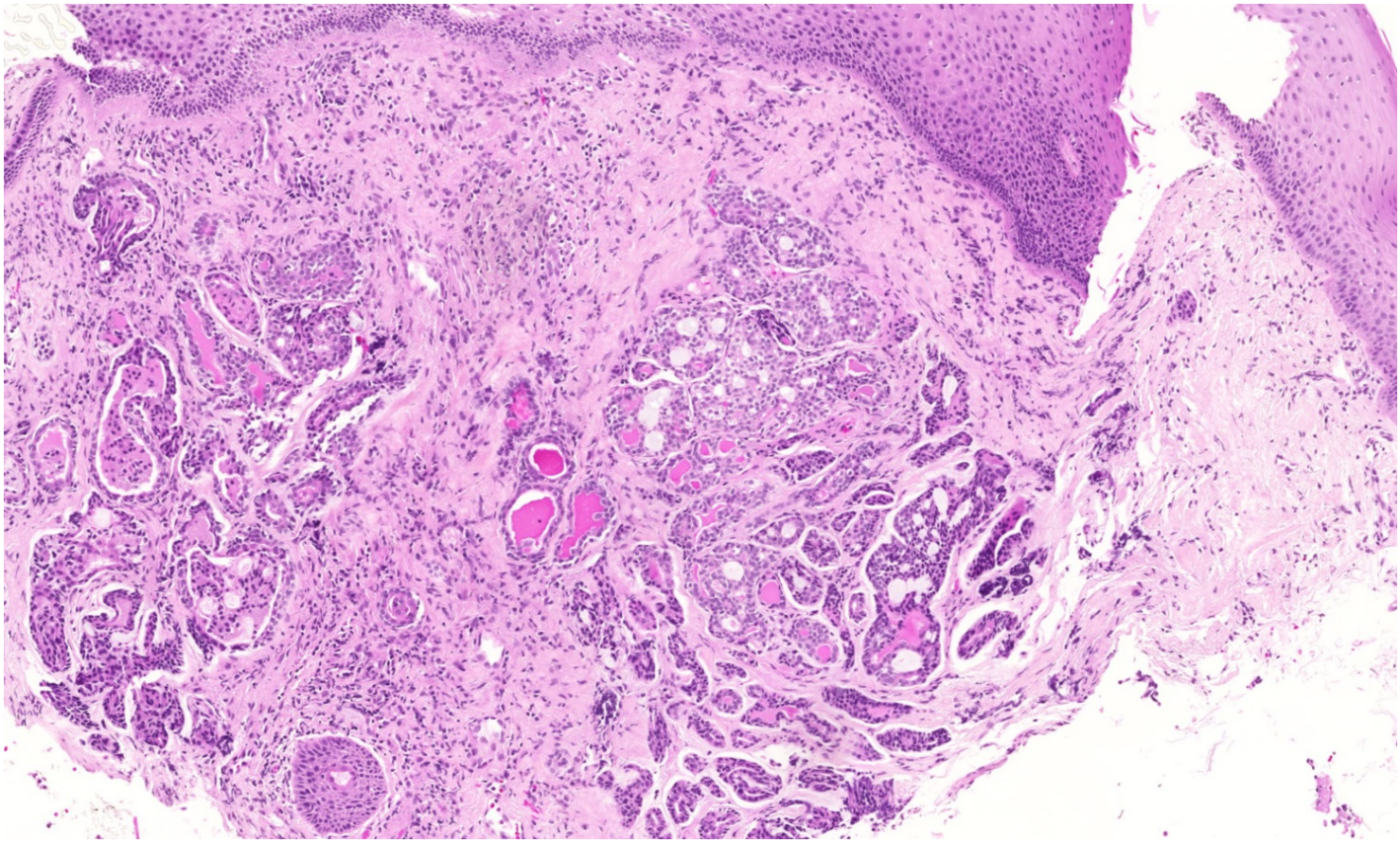

Figure 2. Primary adenoid cystic carcinoma at base of tongue; hematoxylin and eosin, original magnification x 10.

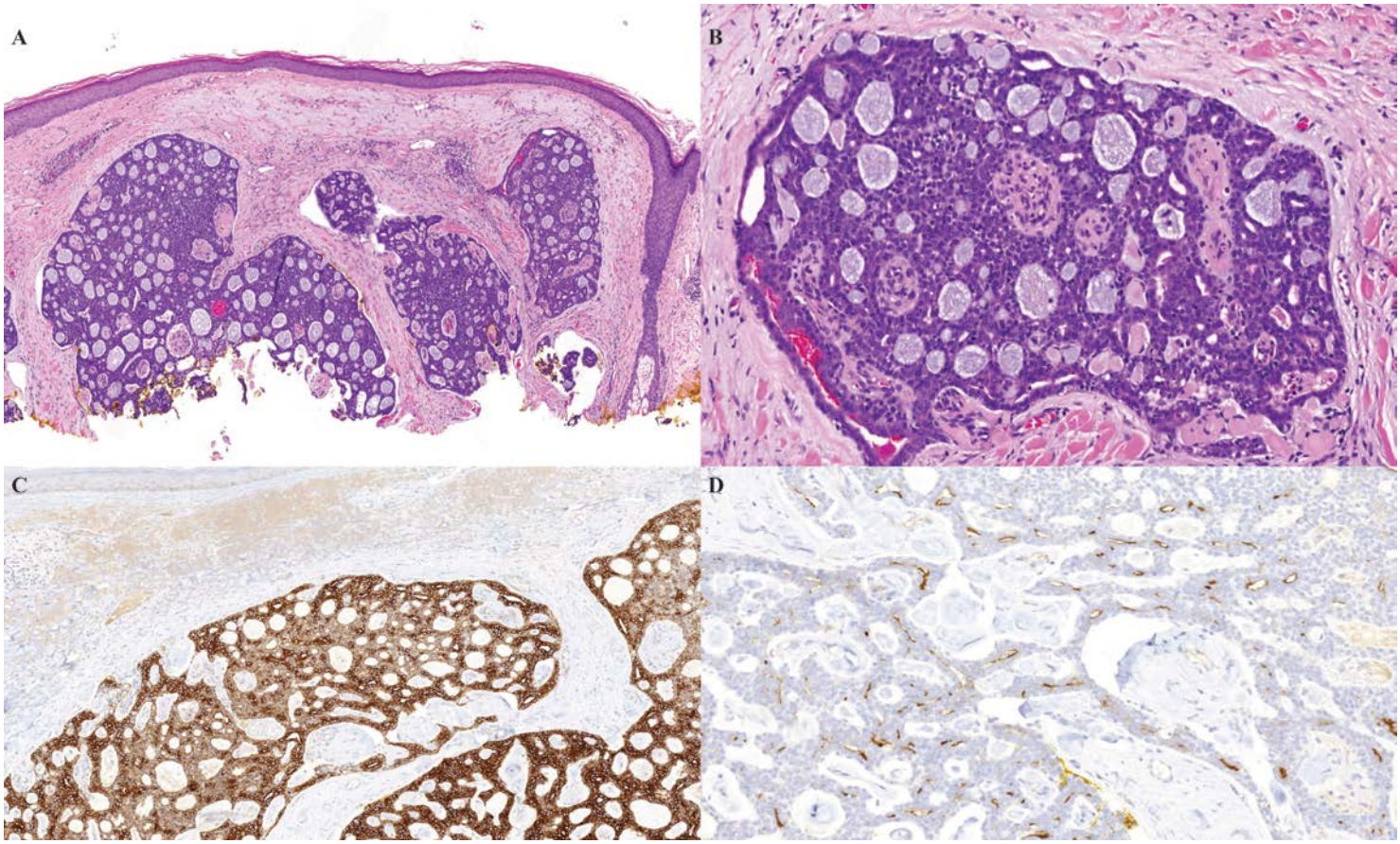

Figure 3. Metastatic adenoid cystic carcinoma to scalp. (A) No connection to epidermis observed; hematoxylin and eosin, original magnification x 5. (B) Cellular detail; hematoxylin and eosin, original magnification x 20. (C) Ductal components outlined with cytokeratin (CK) 8/18, original magnification x 10. (D) Ductal components outlined with epithelial membrane antigen (EMA), original magnification x 20.

Discussion

Differentiating between primary cutaneous ACC and cutaneous metastases originating from an extracutaneous ACC can be difficult, but it has important implications for management, specifically whether chemotherapy should be added to surgical excision [1, 6]. Distant cutaneous metastases from extracutaneous ACCs are rare and carry a poor prognosis [4, 5]. Table 1 presents all 16 cases of extracutaneous adenoid cystic carcinoma metastasizing to cutaneous sites that are documented in the available literature, including the present case. The present case is the second reported case of an ACC metastatic to the scalp.

Table 1. Cases of extracutaneous adenoid cystic carcinoma metastasizing to cutaneous sites[a,b]

Year Published	Authors	Case Number	Primary ACC Origin	Local or Systemic Spread	Time from primary neoplasm to cutaneous metastasis
1939	Coley [12]	1. Phalanx	Unknown	Systemic	Unknown
1960	Fine et al. [12, 13]	2. Subcutaneous tissue of abdominal wall	Minor salivary gland	Systemic	5 years
1975	Weitzner [12, 14]	3. Distal phalanx and adjacent soft tissue of left great toe	Submaxillary gland	Systemic	5 years
1977	Osborn [12, 15]	4. Skin, unspecified	Unknown	Systemic	Not documented
1979	Vinod and Gay [12]	5. Cutaneous surface of the palm of patient's hand	Minor salivary gland in nasal cavity	Systemic	8 years
1986	Nascimento et al. [16]	6. Skin, unspecified	Salivary gland	Systemic	Not documented
		7. Skin, unspecified	Salivary gland	Systemic	Not documented
1999	Makdessian et al. [17]	8. Two cutaneous nodules on right neck	Parotid gland	Systemic	Unable to determine[c]
1999	Nakamura and Miyachi [18]	9. Upper lip	Lacrimal gland	Systemic	2 years
2003	Chang et al. [19]	10. Five subcutaneous nodules on the abdomen	Parotid gland	Systemic	4 years
2007	Perez et al. [9]	11. Subcutaneous nodules on left shoulder, right elbow, and mental region	Parotid gland	Systemic	Nodule on left shoulder – 10 years; nodules on right elbow and mental region – 11 years
2007	Seethala et al. [20]	12. Soft tissue of shoulder	Submandibular gland	Systemic	Not documented
2007	Yurut-Caloglu et al. [21]	13. Cutaneous lesions on plantar surface and fifth digit of left foot	Parotid gland	Systemic	19 years
2012	Park et al. [22]	14. Right upper eyelid conjunctiva and skin	Maxillary sinus	Systemic	9 years
2013	Jedrych and Galan [8]	15. Multiple cutaneous metastases to back and scalp	Lacrimal gland	Systemic	21 years
2014, current case	Saco et al.	16. Mid-posterior scalp	Base of the right tongue	Systemic	15 years

[a]ACC = adenoid cystic carcinoma.
[b]Excluding cases involving the external auditory canal.
[c]Patient had right neck mass for 50 years prior to diagnosis of metastatic skin lesions. However, a fine-needle aspiration biopsy performed 10 years prior to diagnosis of metastatic skin lesions indicated that the mass was benign. A biopsy of the primary tumor that was performed during the same time that the metastatic skin lesions were discovered revealed that the primary tumor was malignant, thus indicating a previously benign pleomorphic adenoma in the parotid gland that underwent malignant transformation to ACC. Histopathologic examination showed no evidence of right parotid ACC extending directly to the skin.

In general, there are no histopathological differences between a primary cutaneous ACC and a cutaneous metastasis from an extracutaneous ACC [9]. Immunohistochemical results in cutaneous ACC closely resemble those of salivary gland ACC. One noteworthy difference is that CEA is consistently positive in extracutaneous ACC, whereas it is only positive in half of the cases of cutaneous ACC [5]. As is typically observed in ACC, the present case was positive for CEA, EMA and cytokeratins AE1/AE3, CK7, and CK 8/18 [9, 10]. Interestingly, although basal cell carcinomas can stain positively for most of the aforementioned immunohistochemical markers, basal cell carcinomas are typically negative for EMA, except for focal staining of keratinizing/squamous areas. Thus, EMA can be very useful in differentiating between a basal cell carcinoma and ACC [11].

CD117 and focal expression of vimentin and S100 can also be observed in ACC [4, 5, 9]. Because primary cutaneous ACCs can be misclassified as metastases, one could argue that the present case is a primary cutaneous ACC rather than a metastasis, even in light of the nearly identical histopathology in the tumors examined. Primary cutaneous ACC, however, should only be diagnosed with certainty if the patient does not have a history of extracutaneous ACC [5].

Conclusion

In conclusion, the present case demonstrates a rare yet relevant scenario that clinicians must consider when dealing with patients who have a history of ACC. The distinction between primary cutaneous ACC and a metastasis originating from an extracutaneous ACC is difficult, but it has critical implications for prognosis and selection of therapy. Primary cutaneous ACC should only be diagnosed with certainty if the patient has no history of extracutaneous ACC. Furthermore, in addition to being monitored closely by oncologists, patients with a history of ACC should also be followed by dermatologists with regularly scheduled total body skin exams.

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