BRIEF ARTICLES

A Case Report of a Primary Cutaneous Adenoid Carcinoma: A Diagnostic and Management Challenge

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ABSTRACT

Primary cutaneous adenoid cystic carcinoma (PCACC) is a rare glandular neoplasm that mimics benign lesions both clinically and histologically making it a diagnostic challenge. Although indolent, ACC has potential for local regional and distant metastasis and, even after surgical excision, there is a high recurrence rate. This report outlines a case of a PPACC and reviews current therapeutic approaches.

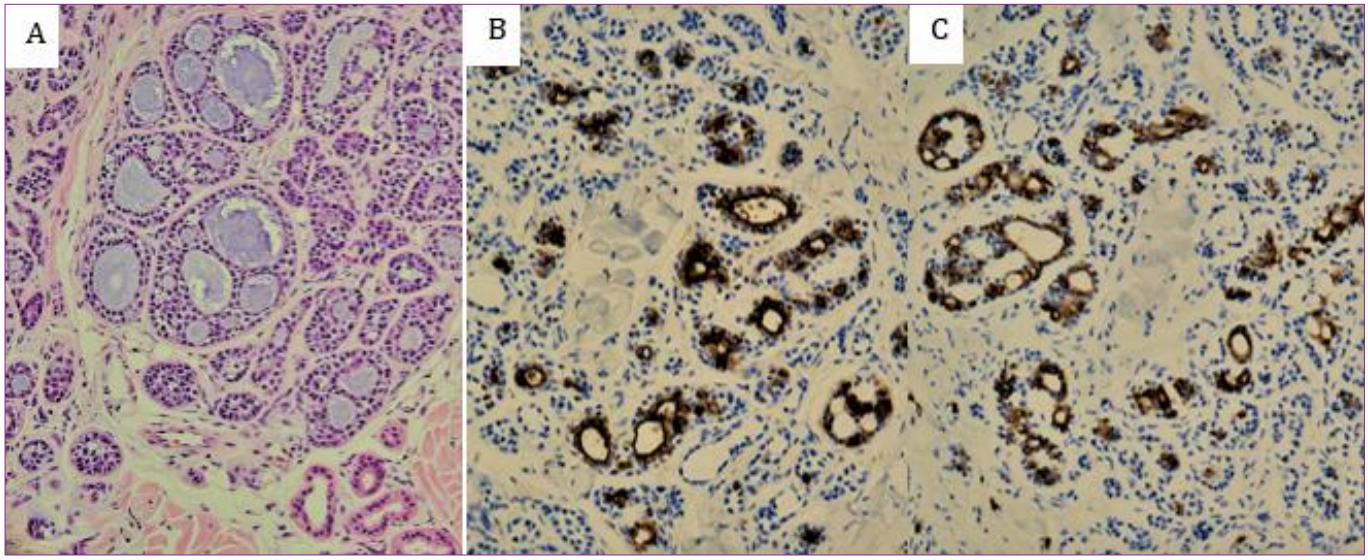
INTRODUCTION

Adenoid cystic carcinoma (ACC) is a slow growing, usually asymptomatic neoplasm most commonly associated with secretory glands. Rarely is it observed as a primary tumor on the skin.[1] It is unclear whether PCACC is of eccrine or apocrine origin.[2] Salivary gland ACC rarely metastasizes to skin.[3] However, ACC is also observed in the lungs, breasts, prostate and cervix and identification of cutaneous ACC should prompt investigations for metastasis or direct extension from salivary glands adjacent to the skin.[1] Further, the median age for developing PCACC is 61 years old with equal disease incidence in men and women. Primary neoplastic sites include the head and neck (46%), followed by upper limbs (17%), trunk (15%) and lower limbs (13%).[1,4] We report a case of PCACC that presents both as a diagnostic and management challenge.

CASE PRESENTATION

A 52 year old female presented to an outpatient dermatology clinic with an asymptomatic 5 mm skin colored, subcutaneous papule on the left mid back resembling a cyst. Given the clinical resemblance to a cystic lesion, it was treated with intralesional triamcinolone. Despite treatment, the lesion persisted and a biopsy was performed. Histologic examination showed a dermal nodular proliferation of basaloid islands and cords separated by spaces with an amorphous PAS^+ material. Initial pathology was interpreted as a benign sweat duct tumor, demonstrating the diagnostic challenges with these lesions. Cognizant that malignant sweat gland tumors can be misread as benign lesions, a second opinion from a more experienced dermatopathologist was sought. On second opinion the diagnosis was of a malignant, low-grade, sweat duct tumor. Staining of areas of ductal

Figure 1. Histopathologic diagnosis of PCACC. (A) Hematoxylin and eosin staining showing an infiltrating intradermal basaloid tumor with a cribriform pattern and mucinous secretions (H&E, 20x). (B) Positive luminal and pseudocystic spaces staining with EMA (10x). (C) Positive c-KIT (CD117) staining around the pseudocyst (10x).



differentiation showed CEA$^+$, EMA$^+$, S100+ and LMWCK$^+$ and c-Kit$^+$ (cd117) favoring the diagnosis of an ACC (Figure 1). The lesion was excised by wide local excision with 1.0 cm tumor free margins. A complete lymph node examination did not show any lymphadenopathy. Whole body PET-CT did not show metastasis suggesting local, regional disease.

DISCUSSION

Management of ACC remains a challenge since there is a high incidence of recurrence especially if there is perineural invasion.[5] Recurrence following surgical resection is around 44% with an average follow-up time of 58 months.[3,5] There is no consensus on the optimal treatment of PCACC. Surgical excision with adjuvant radiotherapy is routinely used for PCACC with skin-limited disease, although comparative studies have not conclusively demonstrated that adjuvant radiotherapy is superior to surgery alone.[6] Reported surgical modalities include local excisions, wide local excision (WLE) with 1.0 – 2.0 cm safety margins and Mohs surgery.[2,5,7] Perineural invasion and local recurrence has been reported in 76% and 44%, respectively with traditional surgical excision.[5] Therefore, in or practice, a WLE with at least 1.0 cm margins is performed. On the head and neck, 23% of PCACC have been identified on the face and 8.6% on eyelids. Mohs surgery should be considered in these cases to spare tissue and preserve function.[2,5] However, PCACC can spread asymmetrically along nerves, therefore Mohs surgery can have limitations for identifying margin clearance when there is perineural invasion.[2]

Staging of PCACC should include routine lymph node examination with whole body imaging to assess for extracutaneous involvement. Lymph node dissection is suggested for nodal involvement[5]. Whole body imaging should include the head and neck since primary ACC accounts for 10% of salivary glands neoplasms. The most common areas for metastasis include the lungs, bone and liver. Since primary ACC can arise from breast and in the

genitourinary tract, an age appropriate mammogram, PAP smear and/or prostate examination should be included in the work-up.[2]

Chemotherapy is first line for advanced, metastatic ACC, albeit this is not standardized.[8] Classical chemotherapy agents including cisplatin, 5-fluorouracil, or a combination of cisplatin, doxorubicin and cyclophosphamide are utilized for metastatic disease. Several studies have shown that drugs targeting epidermal growth factor receptor or the downstream tyrosine kinases has shown some promise in treating advanced ACC in a cohort of patients.[8] More molecular, genetic and clinical studies are needed to identify therapeutic agents specifically targeting ACC.

There are no standard guidelines for follow-up of patients with PCACC. PCACC follows an indolent course and the reported time to recurrence following surgical excision ranges from 1 month to 35 years.[3] A review of fifty cases in the literature showed that on average the cases recurred within 58 months.[3] Follow-up frequency may also vary depending on the presence of any metastatic disease. Therefore, there is insufficient evidence to support any definitive recommendations for follow-up and this should be done on a case-by-case basis.

CONCLUSION

PCACC can be diagnostically challenging owing to its indolent growth and its ability to mimic benign entities. Although there is no consensus regarding management of PCACC, we suggest WLE of at least 1.0 cm margins, full lymph node examination and imaging to elucidate the etiology. Routine follow-up with imaging is suggested given the high incidence of recurrence.

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