BRIEF ARTICLE

A Young Woman with a Cystic Mass in the External Auditory Canal

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ABSTRACT

Introduction: Adenoid cystic carcinoma (ACC) is a malignancy that primarily affects the salivary glands. Primary cutaneous ACC (PCACC) is a rare variant that arises from ceruminous glands of the ear and other glands of apocrine origin.
Case Presentation: We presented a case of a 26-year-old woman evaluated for a slow-growing mass in her ear for several years that was painful with bloody discharge. Physical exam demonstrated a subcutaneous nodule with overlying telangiectasias protruding from the external auditory canal. Excisional biopsy revealed a dermal proliferation of basaloid cells forming islands in a cribriform architecture.
Discussion: PCACC grossly mimics other cysts and tumors, so diagnosis must be histologically confirmed. Basal cell carcinoma, mucinous carcinoma, and spiradenoma can mimic PCACC on histopathology, making differentiation important. PCACC is not associated with common risk factors for malignancy, such as lifestyle habits or known environmental exposures, thus it is important to include on the differential with any auricular tumor presenting as a pearly nodule with blue and purple hues. Despite treatment with wide local excision, postoperative radiation, and less commonly chemotherapy, recurrence rates are high.
Conclusion: It is important to accurately diagnose PCACC, due to the need for long term follow up, as these tumors commonly recur decades later. This case contributes to the growing literature on PCACC presentation, and how the selected treatment shapes the trajectory of patient follow up given there is no consensus on definitive treatment.

INTRODUCTION

Primary cutaneous adenoid cystic carcinoma (PCACC) of the ear is an extremely rare malignant tumor arising from the secretory cells of the ceruminous glands with an incidence of 0.004 per 100,000 people.[1] These tumors are slow growing, commonly misdiagnosed, and often mistaken for nonmelanoma skin cancer or spiradenoma.[2,3] The disease presents as a slow growing ear canal mass that tends to grow along nerves, lending to early symptoms of otalgia, hearing changes, and serous/clear otorrhea. As the disease

progresses, recurrent otitis externa, bleeding, and neuropathy may occur.[2] PCACC exhibits perineural invasion in 76% of cases and local recurrence in 44% of cases, with distant metastasis being less common.[4] Surgical resection with wide margins may be curative; while PCACC has a 5-year survival rate of 96% it has a long term recurrence rate of 44% at up to 20 years post excision.[4,5]

PCACC occurs equally in males and females at an average age of 64 years old.[5] We herein present a case of a middle-aged female who presented with a slow growing mass in her ear for several years that was originally misdiagnosed and undertreated prior to presenting to the clinic.

CASE REPORT

A 26-year-old woman presented for evaluation of a lump in her ear canal that had been present for a few years. Over the preceding few months, the lesion had grown, became painful with intermittent bloody discharge and associated neck pain. Of note, the patient had undergone incision and drainage of the lesion by a doctor in China a few years prior to presentation, without improvement of the mass or symptomatology. Physical examination revealed a skin-colored, firm, subcutaneous nodule with overlying telangiectasias protruding from the external auditory meatus (**Figure 1**). A complete palpatory lymph node evaluation was negative.

Excisional biopsy revealed a dermal proliferation of basaloid cells, forming islands and cords, without epidermal connection (**Figure 2**). The islands of cells had a cribriform architecture and demonstrated basophilic mucin in between the cells. The cells were cuboidal, had a high nuclear-to-cytoplasmic ratio, and did not demonstrate peripheral palisading. Mitotic figures were not evident. CK7 and c-kit highlighted inner ductal cells, while S100 and p40 highlighted the outer myoepithelial cell layer. The cells were negative for CK20.

The patient underwent wide local excision by otolaryngology. She had no evidence of recurrence at her three-month follow-up appointment.

DISCUSSION

Our patient presented years after initial management and likely misdiagnosed PCACC. Earlier diagnosis would have spared the patient significant morbidity. Upon presentation and appropriate excisional biopsy, definitive diagnosis allowed for proper guidance and education to be given to the patient. Due to our limited chronicity, it will be critical to monitor the patient for an extended period of up to 20 years for recurrence.[5,6]

Studies have found that common risk factors for cancers, such as smoking and gender, do not hold true for ACC and should not be used to predict prognosis. Furthermore, unlike ACC, PCACC is much more likely to recur locally than metastasize to distant sites.[3,5,7] As seen in this patient's presentation, the histological appearance of PCACC is similar to basal cell carcinomas and spiradenomas, suggesting the importance of histologic differentiation. Histologically, our patient's biopsy had rare mitotic figures and abundant perineural invasion. Immunohistochemical analysis demonstrated two cell populations. The inner layer of ductal/epithelial cells stained positive for Ber-EP4, CEA, EMA, CD117, and CK7, while the peripheral myoepithelial cells expressed p63/p40, S100, and SMA, consistent with other documented cases of PCACC.[8] Basal cell carcinoma does

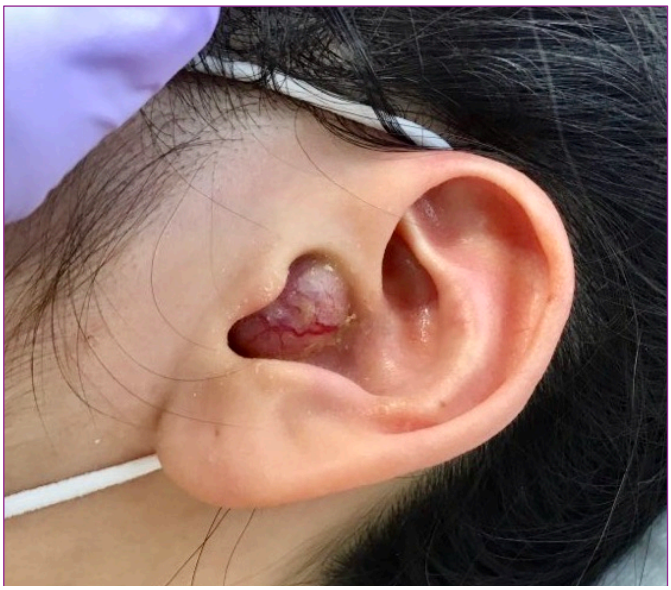

Figure 1. Skin-colored nodule protruding from the left external auditory canal.

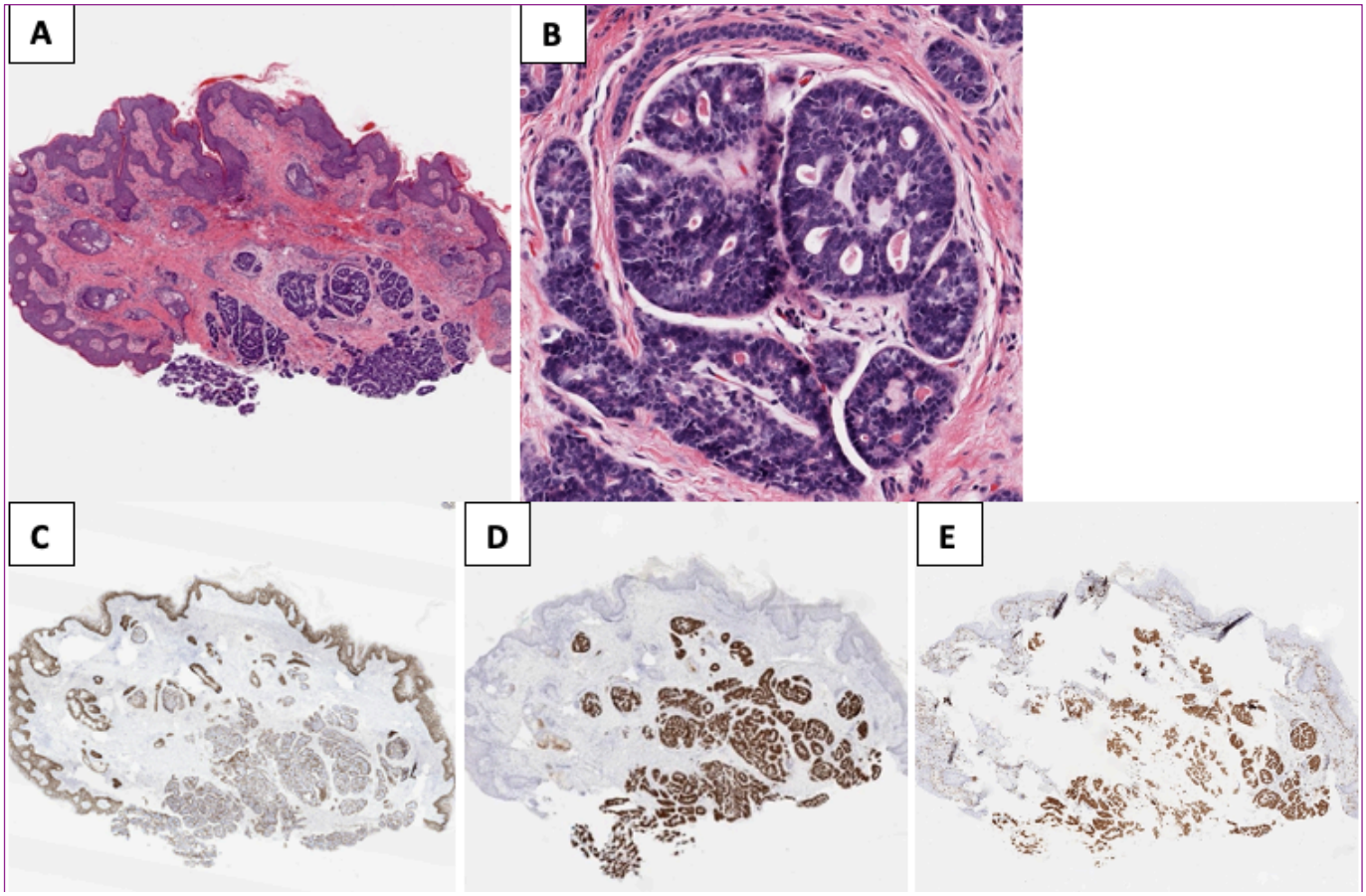

Figure 2. Excisional biopsy of nodule. **A)** Whole tissue section with dermal islands of basaloid cells. **B)** Tumor islands have a cribriform architecture with basophilic mucin in between the cells. **C)** Myoepithelial cells are positive for p40. **D)** Tumor cells are CK7 positive. **E)** Tumor cells are c-kit positive.

not display the two cell populations described above and is negative for CEA, EMA, and CK7. In addition, basal cell carcinoma is negative for CD117 in 80% of cases.[8] Cribriform foci can sometimes be seen in spiradenoma, but they represent a minor feature and are much less organized.[9]

In regards to treatment and follow up for this rare tumor, guidelines and supporting evidence is undetermined and the National Comprehensive Cancer Network states that all ACC be managed postoperatively with radiation.[10] Postoperative radiation therapy has proven to increase recurrence-free survival in ACC of the head and neck, although PCACC specific to the external ear may have the best outcomes with surgical excision alone.[1–3] Interestingly, a recent retrospective study conducted in 2023 found that, in contrast to most studies, there was no association between survival and radical surgery. Chemotherapy may be used in metastatic ACC but has not shown clear benefits; this may be due to its slow growth, making it less responsive to the treatment.[3]

CONCLUSION

PCACC represents a difficult tumor to diagnose and manage largely due to its slow growth, similarity to other common tumors, and delayed recurrence. It is important to continue including PCACC in the differential for cutaneous head and neck masses, report novel effective therapies, and provide treated patients with long term follow up. Given that there is no consensus on the treatment of PCACC, we hope this case will contribute the evidence for therapeutic options and follow up of this rare disease.

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