Unexpected Malignancy Presenting as a Cystic Scalp Lesion

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INTRODUCTION

- Primary cutaneous adenoid cystic carcinoma (PCACC) presents as a painless nodule that commonly arises on the head and neck.
- Over 50% of cases present with perineural invasion, leading to high recurrence rates.
- Metastasis from underlying adenocarcinoma must be excluded.
- Mohs surgery may be considered as first-line treatment due to high recurrence with wide local excision.
- Data on the utility of adjuvant radiation is limited, and this may be considered on a case-by-case basis.

Case Presentation

HPI: 52-year-old African American male presented to general surgery for a several-year history of a scalp lesion that had enlarged and become tender.

Examination: A poorly circumscribed subcutaneous, subcentimeter papule on the scalp was noted.

Histopathology: An excisional biopsy demonstrated a proliferation of epithelial cells with a ductal and cribriform pattern, basement membrane material, and blue mucinous material (Figure 1), confirmed as adenoid cystic carcinoma by immunohistochemistry. Extensive small and large caliber perineural invasion (PNI) was noted (Figure 2 and 3).

Additional workup: PET/CT and laryngoscopy ruled out metastasis from another site.

Treatment: The patient was referred to Mohs surgery, requiring 4 stages to achieve negative margins without perineural invasion. Due to extensive neurotropism, the patient underwent adjuvant radiation therapy.

Follow-Up: There is no evidence of recurrence for 2+ years now.

Discussion

- PCACC is a rare adnexal carcinoma of the skin that most commonly occurs on the head and neck of older individuals.[1] Forty percent of all tumors occur on the scalp.[5]
- PCACC typically presents as a painless, skin colored, nodule, with a mean size of 4.0 cm.[2]
- PCACC is present for an average of 10 years prior to diagnosis.[3]
- Current literature reports approximately a 1:9 ratio of African American to White patients.[1]
- **Approximately, 32% of PCACC present with regional or distant metastasis at the time of diagnosis.**[2]

Figures/Pathology

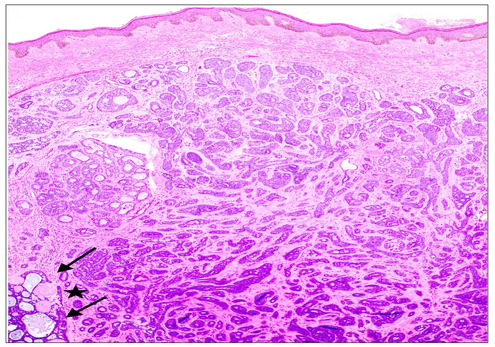
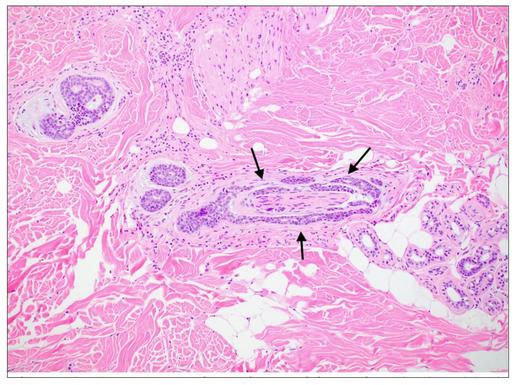


Figure 1. Histopathology of adenoid cystic carcinoma. Ductal and cribriform pattern of a basaloid cell proliferation noted. Arrows point to intraluminal blue mucinous material (lower left), and the star marks basement membrane material left (H&E 4X).

Figure 2. Arrows mark perineural invasion (H&E 10X).



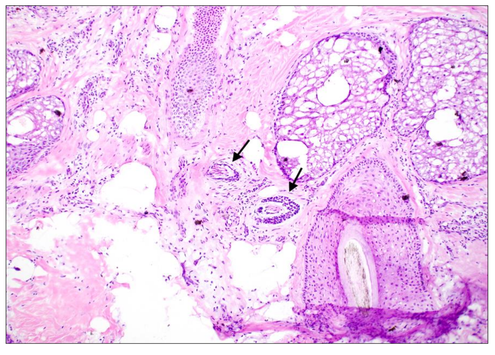


Figure 3. Mohs Frozen Section with arrows marking perineural invasion that required 4 stages to clear (H&E 10)

Histology

PCACC is a poorly circumscribed tumor of basaloid cells with hyperchromatic nuclei arranged in lobules, islands, and cords, often showing cystic and ductal structures with basement membrane or mucinous material. Myoepithelial differentiation, indicated by markers like SMA, MSA, calponin, p63, and S100, rims the basaloid cells, which also stain positive for keratin, EMA, CEA-m, and Ber-Ep4. Perineural invasion is seen in over 50% of cases, typically within the mid and reticular dermis, without epidermal connection. The differential diagnosis includes metastatic ACC (from salivary, lung, or breast origins) and adenoid BCC, the latter showing palisading and stromal clefting absent in PCACC. Benign adnexal tumors like cylindroma and spiradenoma are also considered but differ in cystic and cellular patterns and lack perineural invasion. Tissue biopsy with histological evaluation and immunohistochemistry remains critical for diagnosis.

Discussion (continued)

- PCACC is a form of adenoid cystic carcinoma (ACC) that occurs exclusively on the skin without metastasis from a distant organ. ACC may occur in the salivary glands, breast, lung, and external auditory canal.[1] **Diagnosis of PCACC requires a systemic workup to rule out cutaneous metastasis from an underlying tumor.**[3]
- Local recurrence from PCACC has been reported at 50-70%.[4]
- Although wide surgical resection with 2 cm margins has been advocated by some authors, a recurrence rate up to 50% has been reported.[6-8]
- Utilization of Mohs surgery has shown lower recurrence rates given the sensitivity for detecting perineural invasion.[6]
- Several studies have described the application of Mohs surgery in treating PCACC in cosmetically sensitive areas with recurrence free survival ranging from 10-28 months.[7]

CONCLUSION

- PCACC is a rare tumor that presents as a non-specific nodule most commonly on the scalp followed by chest sites. Although most cases of ACC occurring on the skin are of primary cutaneous origin, it is important to rule out metastasis from an underlying adenocarcinoma.
- Due to delay in diagnosis and the propensity for PNI, PCACC can behave more aggressively than initially thought.
- Several reports describe the benefit of Mohs surgery for PCACC given the high frequency of PNI.
- We opted to treat our patient with postoperative radiotherapy given the extensive neurotropism, although there is little data documenting its utility in reducing local recurrence. He has had no evidence of disease recurrence during 2 years of follow-up.

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