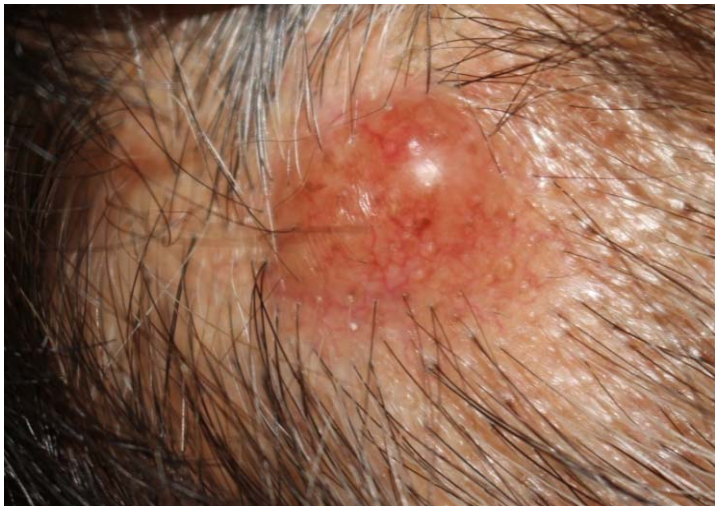
Case 28
A 58 year-old Thai man from Bangkok
Chief complaint: Asymptomatic slow-growing nodule on left temporal scalp for 7 years



Present illness:
The patient developed a solitary asymptomatic slow-growing nodule on left temporal scalp for 7 years. He denied history of trauma in this area and had no history of weight loss, fever, or fatique.

Past history:
He had no known underlying disease.

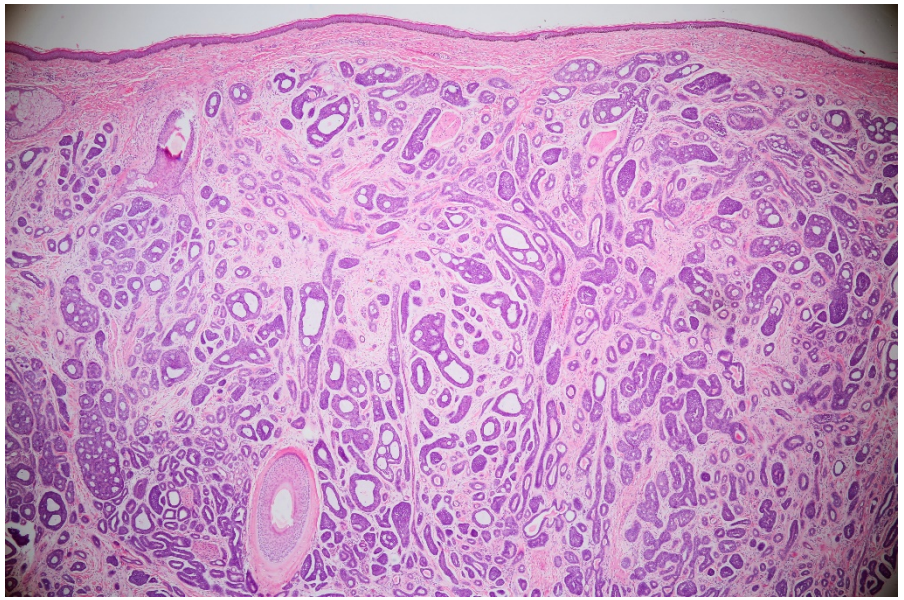
Family history:
There was no history of skin tumor in his family.

Skin examination:

- A solitary asymptomatic firm erythematous to skin-colored nodule with smooth surface and visible telangiectasia, 1.5 cm in diameter, located on left temporal scalp

Other physical examination: Unremarkable

Histopathology: (S16-01022A, scalp)



- Multiple ducts, cysts and variably sized islands of tumor cells in adenoid pattern, dispersed in a loose fibrous stroma

- Epithelium consisting of uniformly cells with hyperchromatic nuclei, conspicuous nucleoli and scant cytoplasm with ductal differentiation

- Mucin within ductal lumens

Investigation:
- Complete ENT examination was unremarkable

Diagnosis: Primary adenoid cystic carcinoma

Treatment:
- Wide excision with full-thickness skin graft

Presenter: Vipawee Ounsakul, MD
Consultant: Suthinee Rutnin, MD

Discussion:

Adenoid cystic carcinoma is a tumor most commonly diagnosed in the salivary glands and occasionally has been reported to arise in the breast, lungs, uterine cervix, prostate, lacrimal gland, external auditory canal, Bartholin glands, and the skin[1]. According to the rarity of primary cutaneous adenoid cystic carcinoma (PCACC), therefore the diagnosis of PCACC should only be established after excluding metastatic disease to skin from another primary site.

Primary cutaneous adenoid cystic carcinoma PCACC, a rare tumor first described in 1975 by Boggio[2], is classified by the World Health Organization within the category of adnexal skin tumors[3]. In United state population-based study of 152 PCACC patients diagnosed during the 30-year period, the overall PCACC incidence ratio was 0.23 per 1 million person-years[4].

Histogenesis of PCACC is still unknown, an origin from apocrine or eccrine gland is still debated[3].

PCACC commonly arises in middle-aged to elderly patients, with a mean age at diagnosis of 63 years[4]. Based on frequencies, a slight female predominance has been reported, however, the largest PCACC series found similar overall incidence ratio for men and women[4].

PCACC commonly presents as a variably symptomatic, slow growing, firm, skin-colored or erythematous, intradermal to subcutaneous nodule, varying in size from 0.5 to 9 cm. PCACC occurs predominates in the face, head, and neck region, with the scalp is the most commonly affected site[5,6]. Other reported locations include the chest, abdomen, back, eyelid, and perineum. The lesion may be asymptomatic, tenderness, or, rarely, pruritus[7]. Patients with scalp lesion may also present with local hair loss or scarring alopecia[8].

Diagnosis of PCACC is primarily based on histologic examination because of no distinguishing clinical features of the tumor. In addition, a thorough work-up should be performed to exclude metastasis to skin from another primary site, especially from salivary gland, before given the diagnosis of PCACC.

Histologically, PCACC is a poorly circumscribed tumor typically occupies the mid and deep dermis and may extend into the subcutaneous fat. The neoplasm consists of round to oval basaloid cells arranged in glandular, cystic, tubular, and a characteristic cribriform pattern. The tumors embed in a loose fibrous and sometimes mucinous stroma. Tumor cells have single or multiple hyperchromatic nuclei with sparse amphophilic cytoplasm. Mitotic figures are generally low, with no prominent nuclear atypia. True luminal spaces are surrounded by modified myoepithelial cells, often with prominent basement membrane material, which is periodic acid-schiff-positive, and diastase-resistant. The cystic spaces often contain alcian blue-positive mucin[5,8].

The microscopic differential diagnosis of PCACC includes metastatic cutaneous adenoid cystic carcinoma, adenoid basal cell carcinoma, cylindroma, mucinous apocrine carcinoma, and apocrine mixed tumor of the skin[5,8].

Immunohistochemical study of the tumor appears to shows two differentiated patterns, epithelial/ductal lines and myoepithelial line[9]. Epithelial/ductal cells are positive for EMA, CEA, cytokeratin, and S100 protein. Whereas myoepithelial cells are positive for SMA, S100 protein and cytokeratin. Cystic spaces are positive for type IV collagen and laminin, suggestive of basement membrane material[9].

In contrast to an aggressive behavior of salivary adenoid cystic carcinoma, PCACC is a more indolent low grade malignancy with a favorable prognosis. The overall 5-year survival rate is 96.1%[4]. Local recurrence is common, but lymph node metastasis is infrequent and distant metastasis is exceeding rare. The duration between surgery and recurrence that has been reported, ranged from 1 month to 35 years, with 44% of cases recur within an average of 58 months of follow-up[5]. Among 130 PCACC cases with specified stage, 70% presented with localized stage, 25% with regional metastasis and 5% with distant metastasis at diagnosis[4]. Although lymph nodes and lungs are the most common metastatic sites, pericardium, bone, nose, liver, and brain involvement have also been reported[10]. Metastasis may develop lately, up to 18 years in 1 case report[8].

5-year survival rate appeared slightly better among male than female, patients given a diagnosis before age 50 years than in older age, patients with localized and regional disease than metastatic disease and patients with PCACC located on the face, head, and neck region than on the trunk[4]. Perineural invasion is common, seen in 76% of PCACC, and associated with higher recurrence rate of 46% compared to only 22% of patients without perineural invasion[5].

An association of PCACC with subsequent development of lymphohematopoietic cancer and papillary thyroid cancers has been reported, with a significant standardized incidence ratio of 3.7 and 15.2, respectively[4].

The recommended treatment of PCACC based on the limited number of case reports is a wide surgical excision with at least 2-cm tumor-free margins and long term follow-up for possible recurrence[9-11]. Mohs micrographic surgery was also performed in several cases, without local recurrence during the follow-up peroid[12]. Lymph node dissection is not currently recommended unless there is evidence of lymphadenopathy[5]. Radiotherapy may have palliative value in cases that surgical removal is impossible or after local recurrence[13]. Chemotherapy is usually reserved for metastatic disease, in which cisplatin-based regimen reported to be relatively more effective than others[13].

References

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