Adenoid Cystic Carcinoma of the Scalp as a Cause of Recurrent Operations

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Abstract
Introduction: Primary cutaneous adenoid cystic carcinoma (PACC) is a rare slow-growing tumor.
Presentation of the case: We report a 62 year-old woman presented with a nodular lesion on the scalp who had three operations before for recurring lesion at the same localization. In the operation, we detected a lesion which was adherent to the subcutaneous tissue. The lesion was firm and gray-white colored, and invading the surrounding soft tissues. According to histopathological findings, the patient diagnosed with adenoid cystic carcinoma.
Conclusions: Primary cutaneous adenoid cystic carcinoma is a rare tumor that the etiology, origin, and treatment are still controversial. The metastases must be ruled out from other sites, especially from the salivary glands after the its pathological diagnosis.

Keywords: Primary cutaneous adenoid cystic carcinoma; Scalp; Salivary gland histopathology

Introduction
Adenoid cystic carcinoma is known commonly as a tumor of salivary glands. However, it also may be seen rarely in lacrimal glands, mucosal glands of the upper respiratory tract, external auditory canal, breast, Bartholin’s glands of the vulva, uterine cervix, prostate gland, esophagus, and other sites. It may also involve the skin by direct or perineural invasion or as a site of distant
metastases [1]. Primary cutaneous adenoid cystic carcinoma (PACC) is a rare slow-growing tumor and firstly described by Boggio in 1975 [2]. There are about 60 cases in the literature. The most common site of PCACC is the scalp. The other sites are chest, abdomen, back, axilla, eyelid, and perineum. It may be asymptomatic or may present with local hair loss, tenderness, pruritus, or pain. It may recur locally and time interval for recurrence ranges between one month and 35 years [1].

Herein, we are presenting a new case with PCACC of the scalp that had three operations before for recurring lesion at the same localisation with no pathological diagnosis.

**Case Presentation**

A 62 year-old woman presented with a nodular lesion on the scalp. It was itching and mildly painful. Her medical history revealed that she was operated three times before for this lesion in the last 2 years. The last operation was about 6 months ago, but there was no histopathological diagnosis. There was no history of weight loss. In the physical examination, a firm nodular lesion was detected on the right parietal region. It was not mobile and was very close to the traces of the previous sutures. Neurological examination was completely intact. Brain computed tomography was performed that did not reveal any intracranial extension of the lesion or any other accompanying lesions. Our clinical approach revealed two suggestions about the nature of the lesion. First, as the lesion was recurring and had no prior diagnosis, it might be any type of tumor. Second, there might be a foreign body reaction due to the previous operations. At the operation, we determined that there was no foreign body. We detected a lesion which was adherent to the subcutaneous tissue. The lesion was firm and gray-white colored, and showed invasion to the surrounding soft tissues. It was excised broadly.

Grossly the biopsy was composed of skin and subcutaneous tissue that was measured as 3.2x2.3x0.8 cm. Macroscopic examination revealed a gray-white colored lesion about 1 cm with irregular borders. Microscopically, the lesion was localised in the dermis and subcutaneous tissue without an epidermal connection. The lesion was composed of myoepithelial and basaloid cells forming cystically dilated cribriform and tubular patterns, and focally solid areas (Figure 1). Some true lumina were seen. There were basal membrane like material, positive with PAS histochemically, among the tumor cells. Histochemically, mucin was detected in the cystic spaces and between the tumor cells with Alcian blue (pH: 2.5) (Figure 2). Immunohistochemical analysis revealed strong positivity with EMA, and weak positivity with CEA in true luminal cells. SMA, p63 and S100 immunostained myoepithelial cells. CD117, chromogranin A and synaptophysin were negative. Twenty two mitotic figures were detected in ten high power microscopic fields, and Ki-67 proliferation index was about 40-50%. The lesion was entirely examined but no perineural invasion or angiolymphatic invasion was detected. Coagulation necrosis was present focally in the lesion. Surgical margins were tumor free. The tumor was diagnosed as adenoid cystic carcinoma.

After the histopathological diagnosis, tumor markers were examined and found to be negative. Neck, thorax, upper and lower abdominal computed tomography images were performed, and no other lesion was detected. The metastatic tumor was ruled out by these additional examinations and we diagnosed the case as primary cutaneous adenoid cystic carcinoma of the scalp. The patient was advised to receive chemotherapy or radiotherapy because of the high recurrence risk, but she did not accept any therapies. She is still under clinical follow-up without recurrence for 8 months.
Figure 1 Dermal tumor showing cribriform, tubular and solid patterns (H&E, x40). Inset, infiltrating component of the tumor (H&E, x100).

Figure 2 Mucinous secretion in some ducts and between the tumor cells histochemically (Alcian blue pH 2.5, x200).
Discussion

For the diagnosis of PCACC, it is crucial to rule out metastasis because adenoid cystic carcinoma can occur at other sites mostly in salivary glands, palate, submaxillary glands, and rarely in nasopharynx, external auditory canal, breast, prostate gland, uterine cervix or lung etc [3].

PCACC is a slow-growing nodular malignancy and has an indolent course with local recurrences and rarely metastasis. The most common site of PCACC is the scalp and 41% of PCACC arises on it. PCACC shows a mild female predominance between 60-70 years [3,4,5,6]. The patients with PCACC are usually asymptomatic. However, local hair loss, tenderness, pain, and/or pruritus may occur [1]. Our patient had complaints of itching and mild local pain.

Although PCACC is known for more than 35 years, the origin and etiology are still uncertain [3]. There are some reports in the literature suggesting that the tumor may originate from eccrine glands, apocrine glands or modified apocrine glands.

Histopathologically, the PCACC is a poorly circumscribed tumor consisting of myoepithelial and basaloid cells with cribriform, tubular, and solid formations in a loose fibrous stroma with mucinous component [3]. The PCACC is reported to show immunoreactivity with pan-cytokeratin, EMA, CEA, CD117, keratin 7 in the basaloid cells, and show immunoreactivity with calponin, p63, CD10, SMA and S100 in the myoepithelial component in the literature. Also, PCACC shows weaker positivity with CEA than the extracutaneous adenoid cystic carcinomas, similar to our case [4]. Histopathological and immunohistochemical findings of the present case were consistent with the cases reported in the literature. Adenoid basal cell carcinoma, cribriform carcinoma, apocrine mixed tumor of the skin, mucinous apocrine carcinoma, cylindroma, and neuroendocrine carcinomas are the tumors that should be ruled out in the histological differential diagnosis [4].

The standard management of PCACC is the surgical excision [4]. Wide local excision with tumor-free margins is the established treatment of choice. Also, Mohs surgery is well defined in the literature with seven cases [7]. In rare instances, use of chemotherapy and radiotherapy has been reported [4,8].

PCACC tends to recur frequently after surgical excision. It is suggested that the main mechanism in the development of recurrence is perineural invasion that is seen in 76% of the PCACC. It is reported that the cases with perineural invasion...
have recurrence rate about 46%, however the cases without perineural invasion show recurrence rate about 22% [4]. No perineural invasion was detected in the present case but high recurrence rate suggests that the prior excisions might probably show perineural invasion. Recurrence time of the PCACC may vary between one month and 35 years [4,5,6]. Our patient was operated for three times in the last 2 years before the present excision. The last operation was about 6 months ago. We performed a wide excision.

Conclusion

The PCACC is a rare tumor that the etiology, origin, and treatment are still controversial. The metastases must be ruled out from other sites, especially from the salivary glands after the its pathological diagnosis.

References