ADVANCE RESEARCH JOURNAL OF MULTIDISCIPLINARY DISCOVERIES
ISSN : 2456-1045
VOL.44 I ISSUE-1 I CHAPTER-1
PUBLISHER : INTERNATIONAL JOURNAL FOUNDATION (IJF)



# Primary Cutaneous Adenoid-Cystic Carcinoma - A rare case report

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## ABSTRACT

Primary cutaneous adenoid-cystic carcinoma (PCACC) is a rare slow-growing neoplasm of disputed histogenesis characterized by nest of basaloid cells forming tubular and cribriform with mucin in cystic space pattern at histology and local aggressive behavior. This tumor is most common in the scalp, affects middle-aged and older individuals (mean age 59) and has predilection for women. We describe an unexpected case of PCACC in a 75-years-old man referred to our clinic for a subcutaneous nodule in the scalp showing a slow growth and indolent course. The differential diagnosis and the clinical management of this PCACC patient, successfully treated with a wide local excision, are presented and discussed.

**KEYWORDS:** Rare skin tumors, primary cutaneous adenoid-cystic carcinoma, differential diagnosis

## ORIGINAL RESEARCH ARTICLE

**ISSN :** 2456-1045 (Online)
**(ICV-MDS/Impact Value):** 72.30
**(GIF) Impact Factor:** 5.188
**Publishing Copyright @** International Journal Foundation
**Journal Code:** ARJMD/MDS/V-44.0/I-1/C-1/DEC-2019
**Category :** MEDICAL SCIENCE
**Volume :** 44.0/**Chapter**- I/**Issue** -1(DECEMBER-2019)
**Journal Website:** www.journalresearchijf.com
**Paper Received:** 23.12.2019
**Paper Accepted:** 05.01.2020
**Date of Publication:** 20.01.2020
**Page:** 01-03

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## CITATION OF THE ARTICLE

**Athar M., Kumar A.** (2019) Primary Cutaneous Adenoid-Cystic Carcinoma - A rare case report; *Advance Research Journal of Multidisciplinary Discoveries;* 44(1) pp. 01-03



## I. INTRODUCTION

Primary cutaneous adenoid cystic carcinoma (PCACC) is a rare tumor that affects middle aged and older individuals. It can occur in the scalp (32–41%), chest and abdomen and is characterized by indolent course and local aggressive behaviour.[1]-[7] The average duration of the tumor prior to diagnosis is about 3 years. Patients typically present with slowly expanding skin colored nodules from 0.5 to 5 cm in size.[1]-[7] If the scalp is involved, alopecia is generally an associated finding (figure 1).[3] In the past PCACC has been regarded as an eccrine lesion, but the possible origin from apocrine glands has been also proposed.[2]

**Case report:**

A 75 years old man presented with a subcutaneous nodule of the scalp with associated alopecia. 3 years prior to the presentation she noted an indolent, slowly expanding firm nodular lesion. In her medical history there was no evidence of previous skin lesions or systemic tumor diseases. At physical examination no signs of systemic disease were found and superficial lymph nodes were not palpable. Hematological tests were also in the normal range. The lesion was surgically excised and submitted for histological examination (figure 1-4).

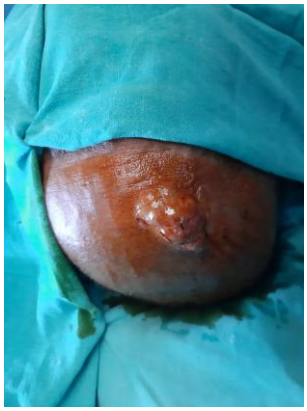

**Figure 1**

**Figure 2**

**Figure 3**

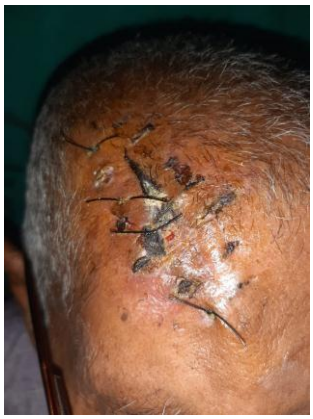

**Figure 4**

Macroscopically the lesion was a 5×3×2.5 cm grey-tan, flesh tumor, with poorly circumscribed borders.

Microscopically it showed a nest of basaloid cells forming tubular and cribriform with mucin in cystic space pattern .cell show high N:C ratio, scanty cytoplasm and round to oval vesicular nuclei with conspicuous nuclei. The lesion invaded the mid and deep dermis but not the subcutaneous fat. The basaloid cells were never displaced in a palisades fashion and no connection to the overlying epidermis was observed. Mitotic activity was very low (1 mitosis ×10 high power field), whereas necrosis, perineural invasion, lymphatic and/or blood vessels infiltration were not present. ( figure 5)

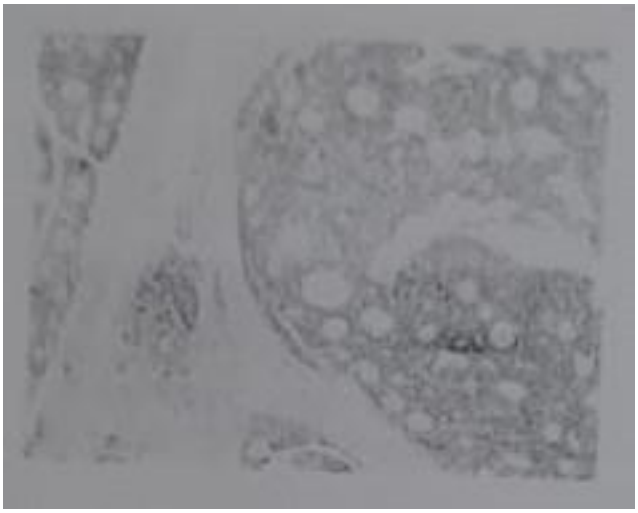

**Figure 5**

At immunohistochemistry, as expected, the tumor expressed low and high molecular weight keratins. A variably expression of S-100 protein and carcino-embryonic antigen (CEA) was also observed. The latter was restricted to the luminal spaces . To better analyze the tumor at phenotypic level, selected monoclonal antibodies to type IV collagen and epithelial membrane antigen (EMA) were also considered. The former highlighted the hyaline deposits among the basaloid cells and the latter the apical aspects of pseudo glandular areas (Figure 5). The basophilic intraglandular material, variably detected in the tumor, was also highlighted with alcian blue staining at pH 2.5 and showed immunoreactivity for laminin .

Both histological and immunophenotypical evaluation suggested the diagnosis of adenoid cystic carcinoma (ACC), but considering the rarity of this entity as a primary skin tumor it was necessary to rule out the possibility of a skin metastasis arising from other malignancies with histological features of ACC.

Adenoid cystic carcinoma, in fact, is most commonly seen as a neoplasm of the salivary glands and seromucinous glands of the upper respiratory tract. It has also been reported to occur in the breast, lung, uterine cervix, prostate, and lacrimal gland. [1],[8] As a consequence a careful breast and an otolaryngology examination was performed at specialist level, followed by head and neck ultrasonographic scan. A PET-CT scan was also considered to complete the oncological screening. Fortunately, the clinical examination was negative and all the radiological exams resulted in the normal range.

All together, the clinical, morphological and immuno-phenotypical features described above, supported the final diagnosis of a primary cutaneous adenoid cystic carcinoma (PCACC). To avoid local recurrences a wide surgical excision with 2 cm disease-free margins was finally performed.

## II. DISCUSSION

Primary cutaneous adenoid cystic carcinoma is a rare skin tumour. It was first described by Boggio in the 1975[3] and, This lesion frequently arises in the scalp of middle-aged or elderly patient, with a slight predilection for women with an average age of 59 years.[1] In about 76% of the cases perineural invasion may be observed and this feature seems to have a prognostic value, with a double increase of relative-risk of local recurrences when it is detected (46% *vs*. 22%).The lesion didn't show images of perineural invasion nor lymphatic or blood vessels infiltration (several histological preparations were examined). As aforementioned adenoid cystic carcinoma is relatively frequent in the salivary glands and in this contest it can be an aggressive tumour. ACC has been reported to occur also in the breast, lung, vulva, cervix and prostate.[1],[8] Considering the age of the patient and the rarity of this tumour in the skin it was imperative to rule out a skin metastasis from an ACC arising in other organs. This fact has important clinical implications because the occurrence of a skin metastasis from a "clinically occult ACC" requires a more complex oncological approach for advanced tumour disease. In our case the negative PET-CT scan and the careful clinical evaluation (in particular breast and salivary glands) allowed the diagnosis of PCACC. As aforementioned PCACC is characterized by a less aggressive behaviour compared to the analogous tumour arising in the salivary gland. The occurrence of lung and lymph nodes metastasis, in fact, is exceptionally rare in PCACC,[7]-[10] but, although infrequently, they can be found in ACCs arising from salivary gland and breast. Moreover perineural invasion, a hallmark feature of the salivary gland ACCs, may be absent in PCACC, as in the

case described here.[8] An indolent course is the major feature of PCACC with a high tendency of local recurrence.[1],[8] For this reason the recommended treatment for PCACC is a wide surgical excision with at least 2 cm tumour free margins.[6]-[11]

## III. CONCLUSION

Primary cutaneous adenoid cystic carcinoma is a rare skin tumor . PCACC is characterized by a less aggressive behaviour compared to the analogous tumour arising in the salivary gland. wide surgical excision with at least 2 cm tumour free margins.

## IV. REFERENCES


[1] **Naylor BA, Sarkar P, Perlis CS, et al.** Pimary cutaneous adenoid cystic carcinoma. J Am Acad Dermatol. 2008;58:636–41.

[2] **Van der Kwast TH, Vuzevski VD, Ramaekers F, et al.** Primary cutaneous adenoid cystic carcinoma: case report, immunohistochemistry and review of the literature. Br J Dermatol. 1988;118:567–7.

[3] **Boggio R.** Adenoid cystic carcinoma of the scalp. Arch Dermatol. 1975;111:793–4.

[4] **Barnes J, Garcia C.** Primary cutaneous adenoid cystic carcinoma: a case report and review of the literature. Cutis. 2008;81:243–6.

[5] **Seab JA, Graham JH.** Primary cutaneous adenoid cystic carcinoma. J Am Acad Dermatolol. 1987;17:113–18.

[6] **Irvine AD, Kenny B, Walsh MY, Burrows D.** Primary Cutaneous Adenoid Cystic Carcinoma. Clin Exp Dermatol. 1996;21:249–50.

[7] **Kato N, Yasukawa K, Onozuka T.** Primary cutaneous adenoid cystic carcinoma with lymph node metastasis. Am J Dermatopathol. 1998;20:571–7.

[8] **McKee PH, Calonje E, Granter SR.** Pathology of the skin. 3rd ed. Philadelphia: Elsevier-Mosby; 2005. pp. 1653–55.

[9] **Weekly M, Lydiatt DD, Lydiatt WM, et al.** Primary cutaneous adenoid cystic carcinoma metastatic to cervical lymph nodes. Head Neck. 2000;22:84–6.

[10] **Doganay L, Bilgi S, Aygit C, Altaner S.** Primary Cutaneous adenoid cystic carcinoma with lung and lymph node metastasis. J Eur Acad Dermatol Venereol. 2004;18:383–85.

[11] **Chesser RS, Bertler DE, Fitzpatrick JE, Mellette JR.** Primary cutaneous adenoid cystic carcinoma treated with Mohs micrographic surgery toluidine blue technique. J Dermatol Surg Oncol. 1992;18:175–76.


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