

# Primary cutaneous adenoid cystic carcinoma of shin

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Primary cutaneous adenoid cystic carcinoma (PCACC) is a rare variant of adenoid cystic carcinoma (ACC) which originates from the skin adnexa, most probably eccrine and apocrine glands. This tumor has a head and neck predilection and also occurs in the chest, abdomen, back, and perineum. We hereby present a 63-year-old man with PCACC located on the shin. No previous similar case has been reported in English literature; therefore, we decided to share our experience about its clinical behavior, histopathologic findings, and management.

**Keywords:** apocrine, eccrine, adenoid cystic carcinoma, adnexal tumor, leg, shin

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## INTRODUCTION

Adenoid cystic carcinoma (ACC) is a common neoplasm originating from salivary glands, especially in the head and neck <sup>1</sup>. It can also originate from other sites such as the external auditory canal, respiratory tract, breast, uterine, cervix, vulva, esophagus, thymus, prostate gland, and skin <sup>1,2</sup>. When the primary site of ACC is the skin, it is called primary cutaneous adenoid cystic carcinoma (PCACC). This variety of ACC was first described by Boggio in 1975 <sup>3</sup>. It should be differentiated from metastatic or direct extension of ACC to the skin tissue. PCACC is a very rare entity and its most common site is the scalp followed by the chest, abdomen, back and perineum <sup>4</sup>. Hereby, we present a case of PCACC of the shin which is a very unusual manifestation of this tumor.

## CASE REPORT

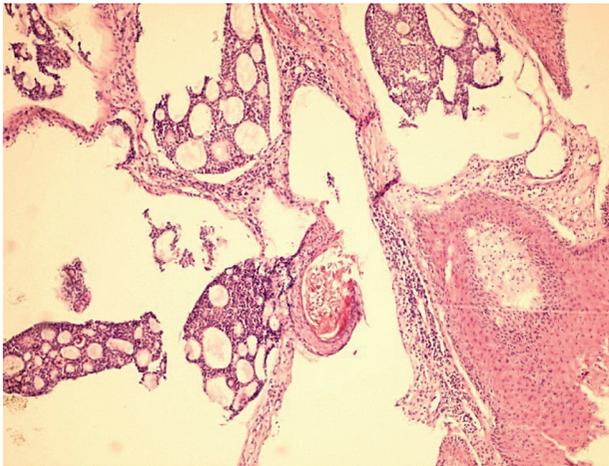
The patient was a 63-year-old man with a mass

on his left shin since 30 years ago. He underwent an emergency operation due to traumatization of the mass and its detachment. A general practitioner excised the remaining of the mass in the emergency operating room. The lesion redeveloped and began to grow slowly at the same site since 7 years before and gradually turned to a painful mass, especially on touch. Therefore, the patient consulted with a general surgeon due to tenderness, focal ulceration, and bleeding of the overlying skin. The mass was excised and sent for pathological evaluation.

Grossly, it was a grayish creamy poorly circumscribed round mass measuring 3.7×3.5×3 cm which was partly covered by ulcerated skin (Figure 1). In cut sections, it was solid and homogeneously creamy. Microscopic evaluation revealed a neoplastic growth of the basaloid cells arranged in a cribriform pattern without any obvious connection with the basal layer of the overlying epidermis (Figure 2). Microcystic spaces were evident in the tumor which were lined by an eosinophilic hyaline membrane and



**Figure 1.** Gross appearance: a poorly circumscribed grayish mass covered by an ulcerated skin.



**Figure 2.** Microscopic findings. This section shows a neoplastic growth of basaloid cells with a cribriform pattern adjacent to the epidermis and a hair structure without obvious connection to them (H&E×10).

contained mucin-like secretions. According to these findings, two differential diagnoses were taken into consideration including PCACC and adenoid cystic basal cell carcinoma (BCC). Immunohistochemistry was performed in the next step to determine the definite diagnosis. Smooth muscle actin, S100, carcinogenic embryonic antigen and CD117 were positive in tumor cells which were strongly in favor of PCACC (Figure 3 A-D).

Ultimately, the patient was referred to a dermatologist with a diagnosis of PCACC, and he was underwent a wide local excision of 2 cm free margin and was discharged with a proper follow up policy.

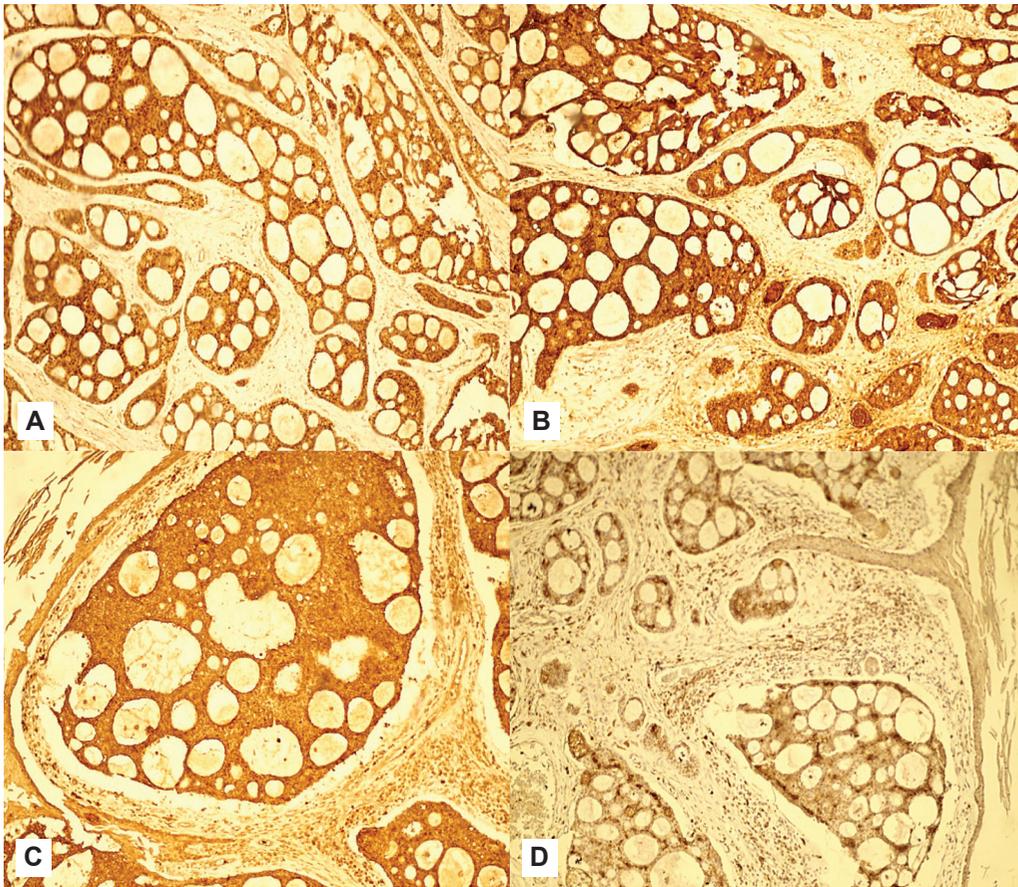
## DISCUSSION

PCACC is a rare variant of adnexal skin tumors with resemblance to ACC of salivary origin. It is suggested that it is derived from eccrine or apocrine glands<sup>5</sup>. Exclusion of metastatic extension of other more common sites is necessary for the diagnosis of this primary skin tumor<sup>6</sup>. According to the most comprehensive English literature review including 50 cases, there is a slight female predominance and the mean age at the time of diagnosis is 59 years old<sup>4</sup>. However, in a recent epidemiologic study, it showed no sex predilection and tended to occur in older age groups. The aforementioned study reported that its incidence rate was 0.24 per million per year, and the most common site of involvement of the tumor was head and neck<sup>7</sup>. Nevertheless, no previous well documented case of PCACC localized to the shin has been reported to date.

PCACC usually presents as a subcutaneous, firm, poorly circumscribed and slow growing mass. The size of the tumor ranges from 0.5 to 9 cm with an average of 3.5 cm. Patients may be asymptomatic or suffer from local hair loss or tenderness<sup>4,8</sup>. However, ulceration of the epidermis and its oozing is a very rare clinical manifestation of this tumor.

On microscopic examination, PCACC demonstrates basaloid cells with hyperchromatic nuclei, but without atypia, arranged in a cribriform, tubular or solid pattern. The tumor is usually located in the mid to reticular dermis without any obvious connection to the epidermis and is surrounded by a fibromucinous stroma. The tumor has an infiltrating margin and perineural invasion is seen in about 76% of the cases<sup>4</sup>. The immunohistochemical profile exhibits a strong positive reaction for epithelial membrane antigen, a usually positive reaction for smooth muscle actin, S100, cytokeratin and a variably positive reaction for carcinogenic embryonic antigen<sup>1,9</sup>.

The histopathologic differential diagnosis of PCACC includes adenoid-cystic variant of basal cell carcinoma (BCC), dermal cylindroma (DC), mucinous carcinoma (MC), and primary cutaneous cribriform apocrine carcinoma (PCCAC)<sup>10</sup>. The most important differential diagnosis is adenoid cystic BCC, especially in our case. This variant of BCC shows gland and cystic like formations, peripheral palisading of basaloid cells, retraction artifact, and



**Figure 3.** Immunohistochemistry profile. A-Smooth muscle actin diffusely stained tumor cells (Hematoxylin counterstain, magnification  $\times 100$ ). B- S100 reacted strongly with tumor cells (Hematoxylin counterstain, magnification  $\times 100$ ). C- Carcinoembryonic antigen stained tumor cells especially near the luminal surface and secretions of adenoid cystic spaces (Hematoxylin counterstain, magnification  $\times 100$ ). D- CD117 showed a diffuse reaction in tumor cells (Hematoxylin counterstain, magnification  $\times 100$ ).

the connection of tumor nests with the epidermal basal layer. Discrimination between PCACC and adenoid cystic BCC is significant, as the latter rarely metastasizes<sup>5</sup>. Immunohistochemically, epithelial membrane antigen, carcinoembryonic antigen, and S100 can be used for differentiating PCACC from BCC. All the mentioned markers are positive in the former (PCACC) and negative in the latter (BCC). DC shows a jigsaw pattern surrounded by a pink hyaline material. MC can be differentiated from PCACC on the basis of produced sialomucins<sup>10</sup>. Finally, PCCAC reveals features such as a diffuse cribriform pattern, interconnections of tumor cell nests and nuclear pleomorphism which are absent in PCACC. On the contrary, PCACC demonstrates uniform tumor cells with spaces of basement membrane material between tumor cell aggregates. In addition, neurotropism is usually found in PCACC while it is never observed in PCCAC<sup>4,10</sup>.

The prognosis of ACC generally depends on the tumor stage, its location, and its histologic pattern. Tumors originating from the breast and lung have a favorable prognosis and a slow infiltrative growth pattern with local recurrence, while salivary ACC has an aggressive course and leads to death due to local recurrence and metastases. In terms of the histologic pattern, those with a solid pattern have a worse prognosis when compared to cribriform and tubular patterns. Although PCACC has an indolent course, it tends to recur locally after excision. Perineural invasion in this variety is a significant indicator of tumor recurrence and the rate of recurrence is remarkably higher in cases with perineural invasion (46%) versus those without it (22%)<sup>4,11,12</sup>.

Wide local excision with tumor free margins is the treatment of choice in PCACC due to its high recurrence, especially in the context of perineural invasion<sup>4</sup>. Some authors have also suggested

Mohs surgery as an alternative treatment<sup>13</sup>. Radiotherapy has also been used as adjuvant therapy, but it has not been proved to reduce the rate of recurrence<sup>6</sup>. Pulmonary metastases have been treated successfully with cisplatin and doxorubicin hydrochloride<sup>14</sup>.

In conclusion, PCACC is a very rare variety of skin adnexal tumors and correct diagnosis of its unusual presentations requires clinicopathologic correlation and immunohistochemistry studies. Discrimination of this tumor from its more common mimickers in the skin such as BCC is important, as their prognosis and management differ.

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