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Primary cutaneous adenoid cystic carcinoma of the abdomen: a rare entity

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Abstract

Adenoid cystic carcinoma is a rare neoplasm that arises from secretory glands, most frequently from the salivary glands. Primary cutaneous adenoid cystic carcinoma is microscopically identical to adenoid cystic carcinoma developing at other tissues. Therefore, differentiating between a primary cutaneous adenoid cystic carcinoma and an extracutaneous adenoid cystic carcinoma with cutaneous metastases is pivotal to determine its prognosis and management. We describe a case of primary cutaneous adenoid cystic carcinoma on the abdomen that was successfully treated with wide excision.

Keywords: adenoid cystic carcinoma, adnexal skin neoplasm, abdomen

Introduction

Primary cutaneous adenoid cystic carcinoma (PCACC) is a rare, slow-growing, malignant adnexal skin neoplasm that was initially described by Boggio in 1975 [1]. Few cases of PCACC have been reported in the literature [2]. It characteristically consists of a mixture of basaloid cells that include myoepithelial cells and ductal cells that together form a distinct adenoid or cribriform pattern in the mid-to-deep reticular dermis [1]. Definitive diagnosis of PCACC is made when typical histologic features are present and metastatic disease is excluded.

Case Synopsis

A 79-year-old man, who was otherwise healthy, presented with an 18-month history of an asymptomatic slow-growing skin lesion on the abdomen. There was no history of trauma and his medical history was irrelevant. Physical examination revealed a hard, erythematous nodule with 1.1×0.8cm (**Figure 1**). There were no palpable lymph nodes or organomegaly.

Excisional biopsy of the nodule was performed and revealed a multinodular cribriform tumor located in the dermis with mucin deposits, occasional hyaline, and marked ductal differentiation (**Figure 2**). Neoplastic cells were cuboid, basophilic, and monomorphic with low-grade cytologic atypia and low mitotic activity. Perineural or vascular invasion were not identified. Immunohistochemistry revealed

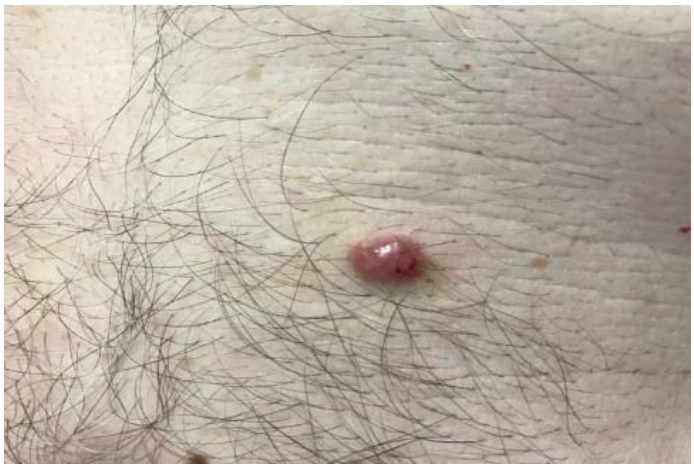

Figure 1. *Clinical aspect of the lesions - erythematous nodule on the abdomen.*

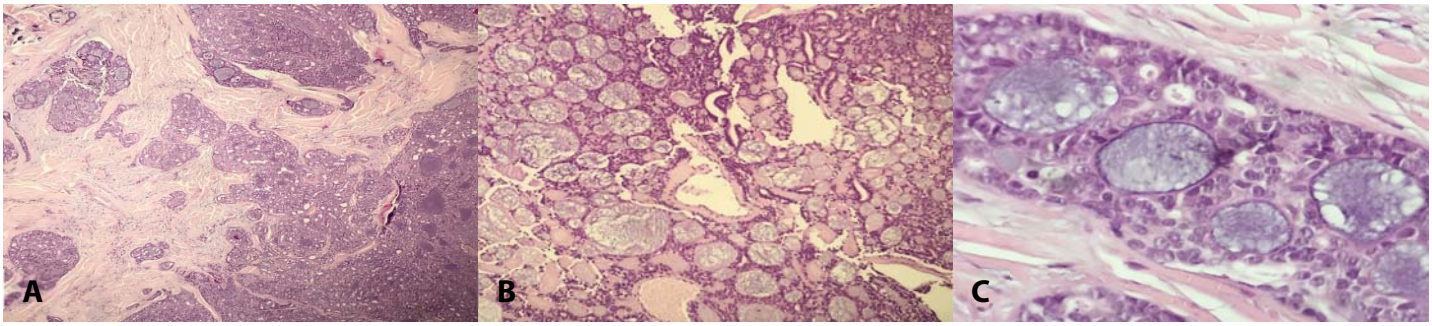

Figure 2. *Histopathology showed a multinodular cribriform tumor with marked ductal differentiation composed by cuboid, basophilic, monomorphic neoplastic cells, and mucin deposits. Hematoxylin and eosin stain, original magnification **A)** 40x, **B)** 100x, and **C)** 400x.*

two distinct cell populations: one that was luminal epithelial and positive for CAM5.2 and pCEA (apical membrane) and another myoepithelial (basal) which stained positive for p63 (**Figure 3**). These findings were consistent with adenoid cystic carcinoma (ACC).

The imaging study, including a computed tomography (CT) scan of the neck, chest, abdomen, and pelvis and a positron emission tomography (PET) scan, revealed no evidence of metastasis or other tumors. The parotid and salivary glands appeared normal on CT scan. Therefore, the tumor was diagnosed as PCACC.

Our patient's tumor was excised with wide local excision with 2cm lesion-free margins. There was no evidence of local recurrence or metastasis after the 24 months of follow-up since the tumor excision.

Case Discussion

Adenoid cystic carcinoma is an uncommon adenocarcinoma that can arise in a variety of gland-bearing organs, more frequently in the salivary glands, breasts, and in upper and lower respiratory tracts. Primary cutaneous adenoid cystic carcinoma is microscopically identical to ACC developing at other tissues, with most cases representing metastatic spread from a non-skin primary tumor. Therefore, all patients require an extensive workup, including CT and PET scans to exclude systemic disease before PCACC is diagnosed [3,4]. In our case no distant disease was observed. Therefore, the cutaneous lesion was determined to be the primary tumor and was classified as PCACC. This tumor occurs in both genders and the average age of onset is around 60 years [5]. It usually presents as a slow-growing, solid to cystic skin-colored tumor on the head, face, upper thorax, and upper extremities that can go undiagnosed for many years. Patients may be asymptomatic or may present with local hair loss, pain, tenderness, or rarely, pruritus [6].

The origin and etiology of PCACC are not completely clear [1]. Characteristically, the tumor is composed of a mixture of basaloid cells with hyperchromatic nuclei without prominent nuclear atypia and more subtle myoepithelial cells; the tumor cells are disposed in a mixture of three main architectural patterns: cribiform, solid, and tubular from the dermis to subcutis [5]. It is usually not associated with the overlying epidermis and rarely invades underlying tissues. Cystic spaces are filled with mucin, usually alcian blue-positive [7]. True lumina are surrounded by modified myoepithelial cells, often with prominent basement membrane material [1]. Histological differential diagnoses include spiradenocarcinoma, cylindroma, adenoid basal cell carcinoma, primary cutaneous cribriform carcinoma, mucinous apocrine carcinoma, and apocrine mixed tumor of the skin [2,5,8].

Primary cutaneous adenoid cystic carcinoma has a 5-year survival rate of 96%, which is higher in patients with primary neoplasms involving the face, head, or neck [8]. It is a locally aggressive disease with one half of patients presenting with a local recurrence after excision [2]. The presence of perineural invasion increases the risk of relapse and may be present in 76% of these neoplasms [2]. Although

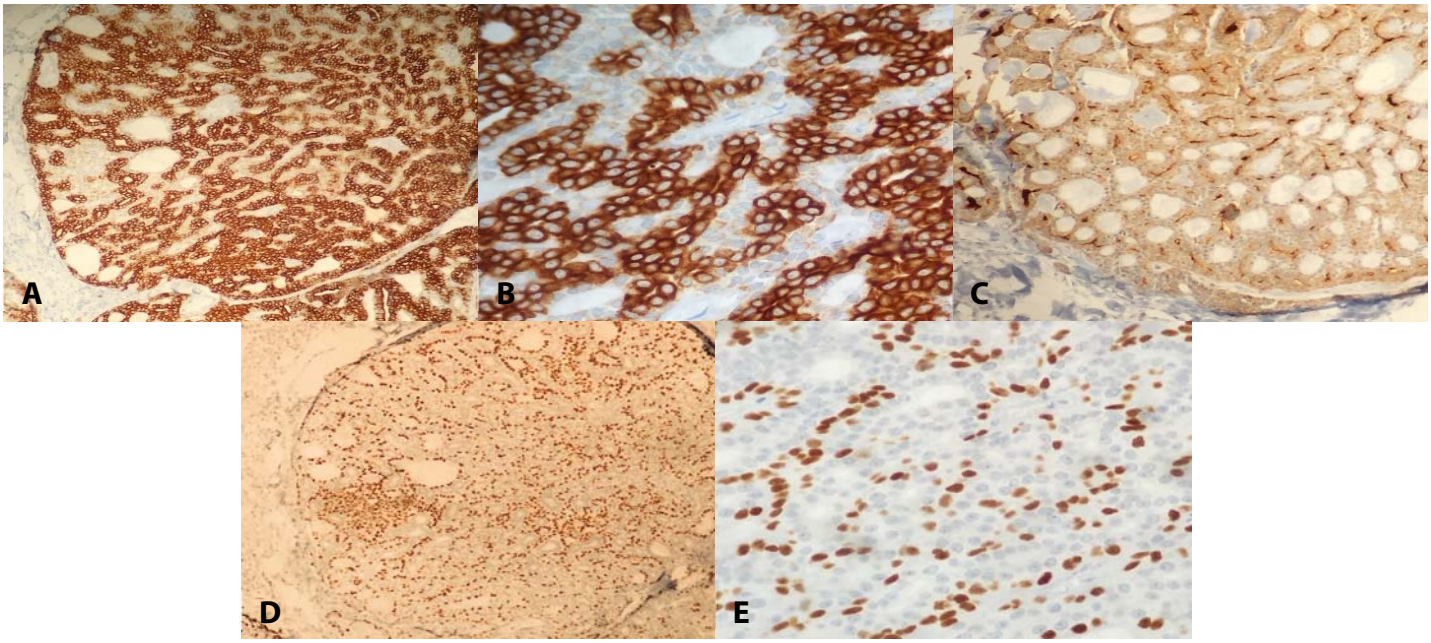

Figure 3. *Diffuse staining and highlighting of epithelial cells. CAM5.2 stain, **A)**, 100x, **B)** 400x. **C)** pCEA, 100x. Moderate staining of myoepithelial cells, p63 stain. **D)** 100x, **E)** 400x.*

ACC of the salivary glands is an aggressive tumor leading to local recurrence and widespread metastases that can cause death in the majority of patients, PCACC has a low metastatic potential since spread to regional lymph nodes and occurrence of distant metastasis are only occasionally observed [2,4,8].

The gold standard treatment of PCACC is wide local surgical resection with at least a 2cm margin around the tumor to exclude recurrences caused by extensive perineural extensions, which can be hard to detect [9]. Some patients also receive adjuvant or therapeutic radiation therapy. However, chemo-therapy is not a common therapeutic option in the treatment of this neoplasm [7,10]. Lymph node dissection is not recommended when there is no evidence of lymphadenopathy. Owing to its tendency for local recurrence, patients should have regular clinical follow-up.

Conclusion

Our report highlights a typical case of a PCACC, which is a very rare tumor and should be considered in the differential diagnosis for slow-growing solitary nodules, particularly if located on the face, head, or neck regions. Skin biopsy is necessary to confirm the diagnosis and an extensive workup is required to exclude metastatic disease.

Potential conflicts of interest

The authors declare no conflicts of interests.

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