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Primary cutaneous adenoid cystic carcinoma: A clinical and histopathological mimic: A case report

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A B S T R A C T

We report a case of cutaneous adenoid cystic carcinoma arising on the left supraorbital region in a 65 -year-old woman. No primary was detected elsewhere in the body. Initial histopathology examination had reported it to be a basal cell carcinoma. The patient was treated with wide local excision and reconstruction with local flaps, followed by adjuvant radiotherapy. The patient remained tumor-free at 24-month follow-up. Till date, 64 confirmed cases of PCACC have been reported in detail in the English literature along with one population based study from the United States, which described 152 cases of PCACC diagnosed over a period of 30 years. Cutaneous adenoid cystic carcinoma has potential for distant metastasis as opposed to basal cell carcinoma so strict vigilance has to be maintained in post-operative period. We propose that possibility of such tumor should be kept in mind during treating skin malignancies such as basal cell carcinomas. Also wide local excision followed by adjuvant radiation is an ideal method to achieve margin-free removal of cutaneous adenoid cystic carcinoma and prevent distant metastasis.

1. Introduction

Adenoid cystic carcinoma (ACC) is primarily recognized as malignant neoplasm of the major and minor salivary glands, it may also infrequently arise from a variety of other rare sites specially those having mucous glands, like respiratory tract, external auditory canal, breast, lacrimal gland, uterus, vulva, cervix, prostate, thymus, skin, and esophagus [1,2]. Primary cutaneous ACC (PCACC) arises directly from the skin and must be differentiated from a metastasis from other primary sites or direct extension of a salivary gland ACC to the skin. This entity was first reported by Boggio in 1975 [3]. Till date, 64 confirmed cases of PCACC have been reported in detail in the English literature along with one population based study from the United States, which described 152 cases of PCACC diagnosed over a period of 30 years [4]. However, no distinction was made between primary and secondary ACC in this population based study. ACC of salivary glands is an aggressive tumor in which local recurrence and widespread metastases result in death in the majority of patient; whereas PCACC follows an indolent course even though it has propensity for local recurrence [1,5]. There is a slight female predominance with mean age of 59 years [5]. Incidence rate per million population per year was highest for PCACC occurring on the face/-head/neck (0.16), followed by trunk (0.04) and extremities (0.02). Among PCACC occurring on the face/head/neck, most cases occurred on the scalp and neck (33.6%), followed by external ear (29.8%), face (23%), eyelid (8.6%), and lip (4.8%) [4].

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[1] The contribution of the other authors as mentioned below with their responsibility in the research.
[2] Contribution- Second Author and contributor as pathologist.
[3] Contribution- Proof reading, Logistic support and analysis.
[4] Contribution- Planning, proof reading and final approval.

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2. Case report

A 65-year-old female was referred to the Plastic surgery outpatient clinic for treatment of a non healing ulcer over the right supra orbital region for the past 7 years. General physical examination revealed a healthy-appearing elderly female having an approximately 30 × 15 mm, non tender, ulcerative lesion with everted margins and focal crusting. The ulcer had now infiltrated the periosteum of the frontal bone. There was no lymphadenopathy or organomegaly. An attempt was made one year back to excise the lesion completely but it recurred. The initial biopsy had been reported as "adenoid basal cell carcinoma" however the histopathology slides were not available for review. Initial PET scan did not identify any other neoplastic focus in the body. The patient underwent wide local excision with 1 cm margin. The involved part of supraorbital rim was excised along with the specimen. Lateral canthus was reattached to the lateral rim. The final defect of 4 × 2 cm was reconstructed with rotation-advancement flaps from cheek and forehead. The resected specimen was sent for histopathological examination which on microscopy revealed a poorly circumscribed infiltrative dermal tumor lacking any connection with the overlying skin and comprising of basaloid and polygonal epithelial/ductal cells, where the former cell type was lying at the periphery and the later occupied central portion of the nodules, glands, tubules, cribriform areas and cystic spaces containing mucinous materials, and cellular debris. Stroma comprised of loose fibrous tissue. Areas with prominent perineural invasion were seen. Figs. 1 and 2.

Immunohistochemistry for epithelial membrane antigen (EMA) and carcinoembryonic antigen (CEA) were positive. Based on the histopathological and immunohistochemical findings, a diagnosis of cutaneous adenoid cystic carcinoma was made. A tumor-free plane was achieved and adjuvant radiation was given in post-operative period. To further rule out an extracutaneous primary site of the tumor and search for evidence of metastasis, additional work-up was completed including a post-operative PET scan and thorough examination of all other systems. All were within normal limits. The patient is now 24 months postoperative, and there is no sign of local recurrence or distant metastasis.

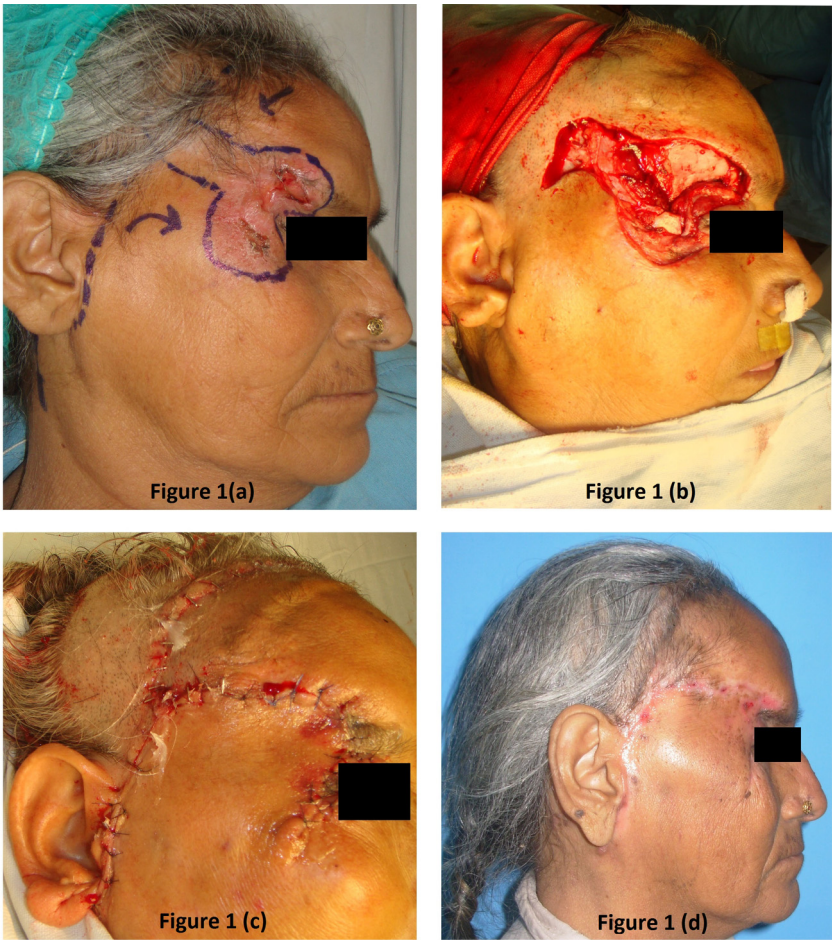

Fig. 1. The figure shows 65 YO female with (a) showing a 30 × 15 mm, non-tender, ulcerative lesion with everted margins and focal crusting infiltrating the periosteum of the frontal bone. (b) The patient underwent wide local excision with 1 cm margin and removal of part of supraorbital rim. (c) Reconstruction was done with local flaps. (d) Postoperatively, six months after radiotherapy.