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A Rare Case of Primary Cutaneous Apocrine Carcinoma of the Forehead

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Abstract

Primary cutaneous apocrine carcinoma (PCAC) rarely occurs on the forehead, and it is easily mistaken for metastatic ductal carcinoma. The pathology of cutaneous apocrine carcinoma usually involves a mixed pattern of tubular and ductal structures containing poorly demarcated margins with cytonuclear pleomorphism, increased mitotic activity, increased tumor necrosis and micropapillary patterns infiltrating the stroma, and the pathological features are difficult to distinguish from those of metastatic ductal carcinoma. This case report describes a rare case of primary cutaneous apocrine carcinoma of the forehead, which was first considered to have originated from the breast. Furthermore, no standard therapy for the treatment of advanced PCAC has been established yet, and treatment depends on the characteristics of the tumor. A selective estrogen receptor modulator may be a suitable treatment for ER(+) and PR(+) PCAC patients.

Introduction

Primary cutaneous apocrine carcinoma (PCAC) is a rare subtype of sweat gland carcinoma, with a low incidence ranging from 0.0049 to 0.0173/100,000 per year. It occurs mostly in regions with a high density of apocrine glands such as the axilla and anogenital areas, although it has also been reported to occur in less typical locations, such as the forehead, wrists, ear canals, eyelids, trunk, feet, toes, and fingers [1], and only 34 cases on the scalp have been reported on PubMed and Google Scholar as of 2021. Although several case reports have described the successful treatment of PCAC with chemoradiotherapy or molecular targeted therapy, no standard therapy for the treatment of advanced PCAC has been established yet [2].

Case Presentation

A 27-year-old woman without systemic disease went to a previous hospital due to a 0.9 cm × 0.5 cm firm nodule over the left side of her forehead; the nodule was excised, and the pathology report indicated the following: invasive ductal carcinoma, ER: 3+ (90%), PR: 3+ (90%), HER2: negative, Ki67: 20%, and R1 resection. Breast sonography at the previous hospital revealed only one cyst over the left breast (BI-RADS Category 3) without a definite mass lesion over either breast except for fibrocystic changes. A whole-body positron emission tomography scan was performed at the previous hospital and revealed no primary tumor over the breast or other distal metastatic lesion. The patient then came to our hospital for wide excision and primary closure. We excised the safe margin of the tumor over the scalp region and sent it for pathology examination.

Pathological examination revealed hyperchromatic nuclei and enlarged nucleoli arranged in a solid pattern. The total Nottingham histologic score was 7 points (tubular formation score: 3; nuclear pleomorphism score: 2; mitotic score: 1); it was first considered metastatic ductal carcinoma, grade II with ER: 3+ (90%), PR: 3+ (90%), Her2/neu: 1+ (negative), and Ki-67: 10%. The histopathological sections revealed that the tumor cells had eosinophilic cytoplasm, hyperchromatic nuclei, and enlarged nucleoli arranged in solid nested, micropapillary, and focal glandular patterns. According to the other immunohistochemical profiles, the tumor cells were positive for TRPS-1, GATA-3 and GCDFP15/mammaglobin, and a tumor origin of the breast was considered. However, a primary tumor not identified. Therefore, we held a pathology conference to discuss the case. Pathology specialists mentioned that on morphological examination, the glandular structure showed apical snouting and decapitation, which are features of primary cutaneous apocrine carcinoma. Pathological interpretation in detail was mentioned in Figures 1-9. The After discussion with a

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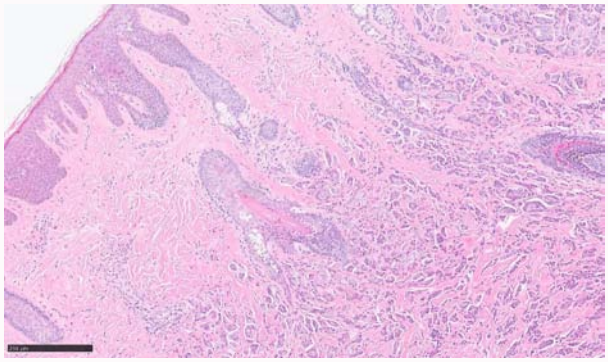

Figure 1: The lower-power field section shows tumor cells arranged in a solid nested pattern, with an infiltrative growth pattern in the dermal stroma (hematoxylin and eosin staining, 40x).

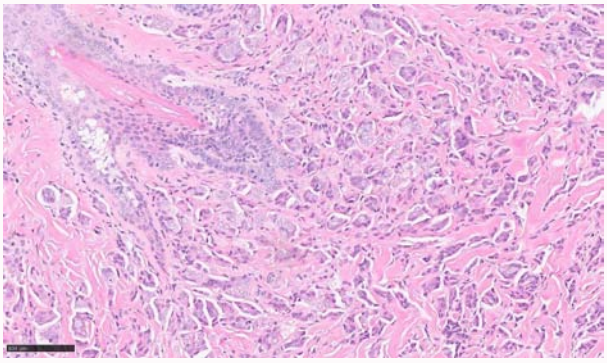

Figure 2: The section shows the tumor cells arising from skin adnexal structures (hair follicle in the left of the picture) (hematoxylin and eosin staining, 100x).

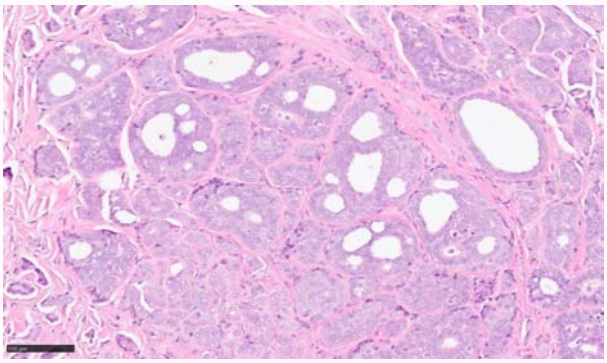

Figure 3: In some sections, the higher-power field included tumor cells with enlarged nuclei, increased N/C ratio, with eosinophilic cytoplasm, arranged in glandular and cribriform structure with focal apical snouting and decapitation secretion (hematoxylin and eosin staining, 200x).

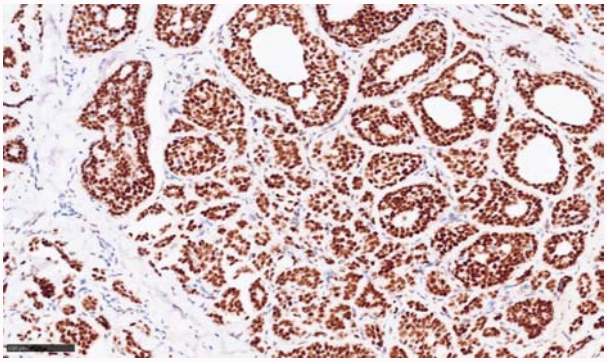

Figure 4: The image of immunohistochemical staining shows tumor cells strongly positive for TRPS-1, which are often positive in breast carcinomas and skin adnexal tumors (immunohistochemical staining of TRPS-1, 200x).

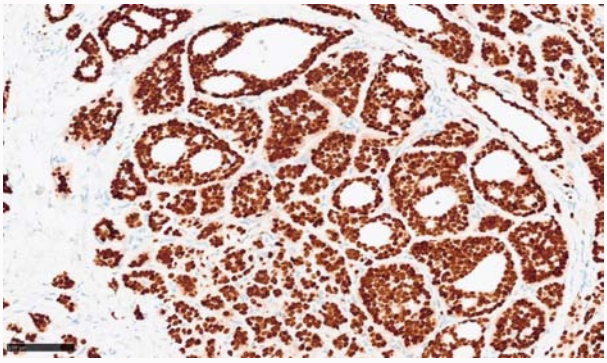

Figure 5: The image of immunohistochemical staining shows tumor cells strongly positive for GATA-3, which are often positive in breast carcinomas and skin adnexal tumors (immunohistochemical staining of GATA3, 200x).

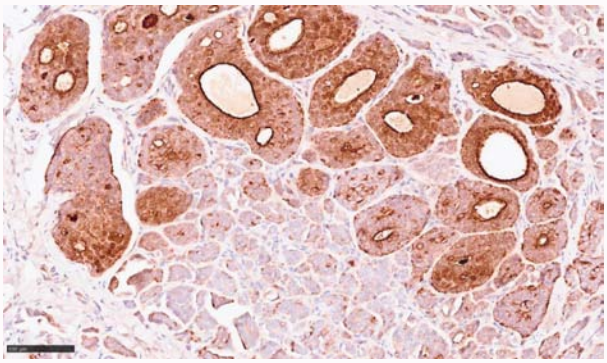

Figure 6: The image of immunohistochemical staining shows tumor cells strongly positive upon GCDFP15/Mammoglobulin double staining, which are often positive in breast carcinomas and some skin adnexal tumors (immunohistochemical staining with GCDFP15/Mammoglobulin double stain, 200x).

pathology specialist and matching clinical manifestations, we revised the diagnosis to apocrine carcinoma of the forehead, pT1N0M0, stage IA. Tamoxifen was prescribed for one year, leuprorelin was prescribed for three months, and no tumor recurrence was noted.

Discussion

Primary cutaneous apocrine carcinoma (PCAC) is often mistaken for other types of tumors. On pathological examination, cutaneous apocrine carcinoma usually displays a mixed pattern of tubular and ductal structures containing poorly demarcated margins with cytonuclear pleomorphism, increased mitotic activity and tumor necrosis. Papillary projections within the lumina and decapitation secretion may be present. Periodic acid–Schiff (PAS)-positive granules are also observed in the lumen, and increased expression of low-molecular weight kininogen (LMWK) is found detected in the tumor cells. Immunoreactivity in the cytoplasm of tumor cells for gross cystic disease fluid protein 15 (GCDFP-15) is usually positive. The level of the tumor marker carcinoembryonic antigen (CEA) may be elevated [3]. Ductal carcinomas have a wide range of pathological features and are thus also referred to as “no special type.” Classically,

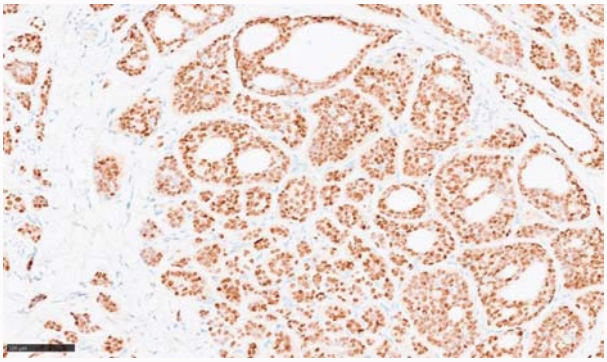

Figure 7: The tumor cells strongly expressed hormone receptors of ER (immunohistochemical staining of ER, 200x).

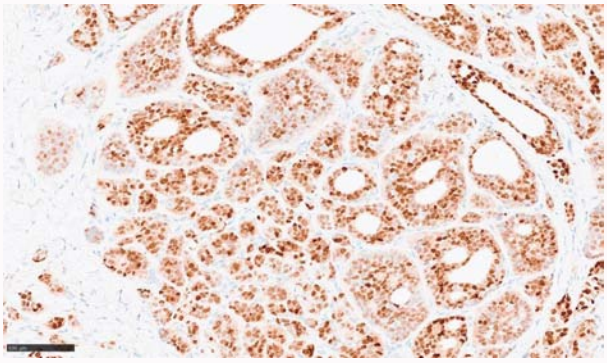

Figure 8: The tumor cells strongly expressed the hormone receptor PR (immunohistochemical staining of PR, 200x).

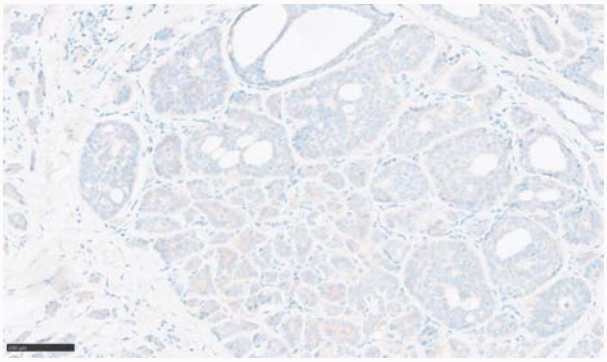

Figure 9: The tumor cells were negative for Her2/neu (immunohistochemical staining of Her2/neu, 200x).

well-differentiated tumors show infiltration of glands through the breast stroma; however, the more poorly differentiated the tumor is, the fewer tubules it contains, presenting more solid sheets of tumor cells [4]. Due to the pathological similarity of the two kinds of tumors, PCAC is easily mistaken for metastatic ductal carcinoma. There have been 4 cases of primary apocrine carcinoma that were initially misdiagnosed as metastatic adenocarcinoma where the primary site could not be identified, as in our case, according to a search of PubMed up to 2022 [2].

To date, no standard therapy for the treatment of advanced PCAC has been established, and treatment depends on the characteristics of the tumor. According to a previous report, a 53-year-old man with a lump over his left cheek was finally diagnosed with PCAC of the face with ipsilateral neck lymph node metastasis post-excision; the cytoplasm of tumor cells presented strong immunoreactivity for GCDFP-15 and strong nuclear staining for both estrogen receptor protein and progesterone receptor protein in most of the tumor cells. Tamoxifen was prescribed for 3 years, and the patient was disease-free during this period [5]. Although there is no standard therapy for the treatment of advanced PCAC, hormone therapy may be considered if it is compatible with the patient's tumor characteristics.

Conclusion

PCAC rarely occurs on the forehead, and it is easily mistaken for metastatic ductal carcinoma. Although there is no standard therapy for the treatment of advanced PCAC, hormone therapy may be considered if it is compatible with tumor characteristics.

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