Giant Primary Apocrine Carcinoma of the Frontal Region: Clinical Presentation, Histopathological Features, and Surgical Treatment

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ABSTRACT Primary cutaneous apocrine carcinoma (PCAC), a subtype of sweat gland carcinoma, is an extremely rare malignant neoplasm. Distinguishing an apocrine carcinoma from a breast carcinoma metastasis is difficult even for a pathologist. Most arise in regions of high apocrine gland density like the axilla, and rarely on the scalp and eyelid, but they can occur elsewhere on the skin. Primary cutaneous apocrine carcinoma of the scalp is a rare malignancy most often reported in the literature as case reports or small case series. The giant form of primary cutaneous apocrine carcinoma in the frontal region has not been described in the literature, to the best of our knowledge. There are no established protocols for treatment of primary cutaneous apocrine carcinoma. We report a case of a giant primary cutaneous apocrine carcinoma localized in the frontal region. A definitive diagnosis of a primary cutaneous apocrine carcinoma was established by biopsy with microscopic and immunohistochemical analysis. Wide surgical excision and reconstruction with large local transposition flap and split thickness skin grafts for secondary defect were our therapy of choice. Primary cutaneous apocrine carcinoma is a very rare malignancy, and the giant form has not yet been described. Surgical treatment provided the patient with tumor-free status as well as satisfactory aesthetical appearance and quality of life.

KEY WORDS: primary cutaneous apocrine carcinoma, diagnosis, surgical treatment, reconstruction

INTRODUCTION

Primary cutaneous apocrine carcinomas (PCAC) are rare and incompletely studied neoplasms, mostly arising in axilla, but they can be found elsewhere on the skin (1). The giant form of PCAC in the frontal region has not been described in the literature, to the best of our knowledge. PCAC is almost impossible to distinguish from cutaneous metastases of breast apocrine carcinoma on histopathological examination alone (2-4).

The standard treatment for PCAC is wide surgical resection and reconstruction (3-6). There is no clear consensus on management of regional lymph nodes, and the most important predictor of survival in a localized disease is lymph node status; therefore, a sentinel lymph node biopsy may be considered in management of this disease (4-6). The role of adjuvant chemotherapy and radiotherapy has not been established.

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Primary cutaneous apocrine carcinoma (PCAC) is a rare cutaneous malignancy with an incidence of 0.005-0.017 per 100,000 patients per year (3). Men and women are equally affected, with the peak of presentation in the 6th and 7th decades of age and a predominance among Caucasians (1,3).

Many of these carcinomas are indolent and slow to develop, but some are rapidly progressive. The majority occur in the genital skin and perineum (34.5%), followed by the trunk (26.4%), head and neck (18.3%), and lower extremities (13.9%) (3,7). They mostly present as nodules or masses 2-3 cm in size, without any additional symptoms, and can very rarely be larger, i.e. than 5 cm in diameter (1,8,9). The giant form, with a diameter of more than 10 cm, and a few centimeters in height, has not been described in the literature, to the best of our knowledge.

CASE PRESENTATION

An 80-year-old man presented with a giant, bleeding, malodorous, secreting tumor (about 10 cm in diameter and 7 cm over the skin level) on the right side of the scalp, with another tumor on the right temporal side (about 3 cm in diameter) of the scalp, and a tumor in the left mandibular area. The patient had noticed the giant tumor about 4 years before admission, while the two other tumors had been present for a year and a half (Figure 1).

Preoperatively, the patient was examined by a team consisting of a plastic surgeon, radiologist, neurologist, and cardiologist and adequately prepared for surgery with general endotracheal anesthesia. The laboratory analyses were within normal limits.

Based on the decision by the tumor board at our institution, the patient underwent radical tumor excision followed by soft tissue defect reconstruction. The wide excision was performed for all three tumors with free margins of 2 cm, followed by reconstruction with a large local transposition flap and split thickness skin grafts for the secondary defect (Figure 2).

The histopathological analysis showed an identical appearance for the giant tumor in the parietal region and the smaller tumor in the temporal area: a nodus with eroded surface, infiltrating dermis, subcutaneous fat and muscles, composed of confluent tumor islands with focal small ducts. Large epithelioid cells with intensely eosinophilic cytoplasm, moderate pleomorphism, and up to 17 mitoses per 10 high-power fields (Figure 3) were immunohischemically diffusely positive for pan-cytokeratin (AE1/AE3), CK7, and androgen receptor, and focally for Gross-cystic-disease-fluid-protein (GCDFP-15) and carcinoembryonic antigen (pCEA). Staining reaction with CK20, p63, mammaglobin, estrogen (ER), and progesterone (PR) receptors, thyroid-transcription factor (TTF-1), and prostate specific antigen (PSA) were all negative. Based on microscopic and immunohistochemical findings, a diagnosis of apocrine carcinoma was established, with clinical exclusion of a possible metastatic lesion being advised, preferentially from the breast and salivary glands. The head and chest X-ray and abdomen/pelvis ultrasound revealed no evidence of a primary tumor, whereas breast ultrasound was without suspicious changes. The tumor in the mandibular region was diagnosed as basal cell carcinoma (nodular type).

The case was reviewed at the Soft Tissue Tumor Board, and no adjuvant treatment, with a follow-up every three months, and then every six months was prescribed. Follow-up checks included complete clinical examination with abdomen and pelvis ultrasound, regional lymph node basin ultrasound, as well as lactate dehydrogenase (LDH) and protein S100 levels. The patient presented no evidence of recurrence

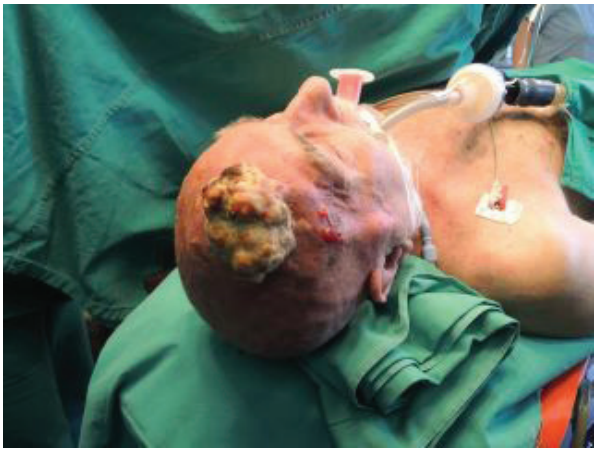

Figure 1. Preoperative view – the giant tumor in the frontal region a primary cutaneous apocrine carcinoma.

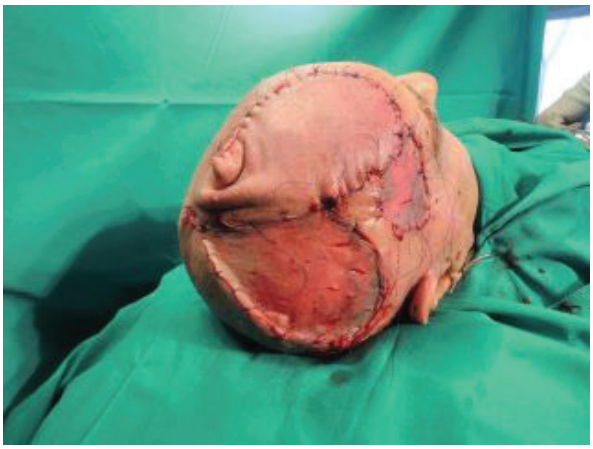

Figure 2. Postoperative view – wide excision and reconstruction with large local transposition flap and split thickness skin grafts for secondary defect.

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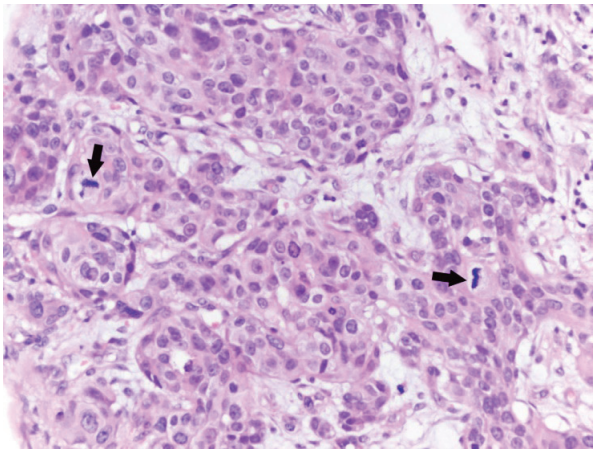

Figure 3. Apocrine carcinoma of the scalp, microscopic presentation with predominantly solid growth pattern and scattered mitoses (arrows) (hematoxylin and eosin, original magnification ×200).

or any local and visceral metastasis during a one-year follow-up. The Soft Tissue Tumor Board decision was based on the elderly age of patient; surgical treatment was performed at the initial stage (wide excision and clear margins) and with absence of intracranial invasion, and locoregional and distant metastasis. Adjuvant chemotherapy and radiotherapy were discussed in light of potential risks, cost-benefit, the patient's age, and absence of significant locoregional and distant tumors spreading. Due to low incidence of PCAC and the lack of large clinical trials, the use of sentinel lymph node biopsy (SLNB) in cases such as this is not well-established, and no guidelines are currently available. The role of SLNB in nonmelanoma skin tumors shows that early treatment of nodal metastases leads to substantial improvements in patient outcomes, including prolonged distant disease-free survival (DFS), but not overall survival (OS).

DISCUSSION

Primary cutaneous apocrine carcinoma (PCAC) as a subtype of sweat gland tumors is a very rare adnexal neoplasm that arises from apocrine glands and usually occurs in regions with a higher density of apocrine glands such as the axilla, groin, and anogenital regions, but can also be found in various body sites (1,3). A definitive diagnosis of PCAC is established by biopsy with histopathological and immunohistochemical analysis. Without the identification of an *in situ* component of the skin tumor, it can be impossible to distinguish PCAC from metastases, particularly of breast carcinoma, which should be excluded with breast examination, clinically, and using imaging methods (9-11). For lesions in other regions with less density of apocrine glands, it can be very difficult to establish a clinical differential diagnosis from more common skin tumors such as basal cell or squamous cell carcinoma.

We reviewed the literature to find the best surgical options and treatment modalities. PCAC of the scalp is a rare malignancy most often reported in the literature as case reports or small case series, and there are thus no established protocols for PCAC treatment, especially for the giant form of PCAC (3.5). Most authors advise clear margins about 2 cm in width. There is no clear consensus on management of at-risk lymph nodes. Since lymph node status is the most important predictor of survival in localized disease, sentinel lymph node biopsy may be considered in the management of this disease (6,8,9). When the tumor is localized on the limb, the role of neoadjuvant or adjuvant chemotherapy as well as radiotherapy or isolated limb perfusion has not been established, nor has the role of immunological therapy.

CONCLUSION

We described a giant form of PCAC of the scalp in an elderly patient, diagnosed with histopathological examination, immunohistochemical analysis, and clinical and imaging investigations for exclusion of a metastatic tumor. Wide surgical excision and reconstruction with a large local transposition flap and split thickness skin grafts for the secondary defect were our therapy of choice. The aforementioned surgical treatment provided the patient with tumor-free status as well satisfactory aesthetical appearance and quality of life.

Ethical approval:

This article was planned in compliance with the Patient Rights Directive and ethical rules by considering the principles of the Declaration of Helsinki. The Ethics Committee of the University Clinical Center of Serbia approved the study, No 11721/2020.

Consent for publication:

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of the present journal.

Availability of data and materials:

The data used during the current study are available from the corresponding author on reasonable request.

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Author contributions

DPP was a major contributor in writing the manuscript and reviewed literature, MN provided the idea for this manuscript, wrote the manuscript, and reviewed literature and interpreted the patient data, MS wrote the manuscript and supervised the manuscript, DB performed the pathohistological and immunohistochemical examination, established the definitive diagnosis, and supervised the manuscript, MPE wrote the manuscript and performed analyses that excluded other tumors. All authors read and approved the final manuscript.

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