Case Report

Apocrine carcinoma of the scalp with neck lymph node metastasis: A case report and review of the literature

Hadi Al-Hakami[1], Baraa I. Awad[1], Mohammed Al-Garni[1], Haneen A. Al-Maghrabi[2], Noura Al-Shareef[1]

[1]Department of Otolaryngology-Head and Neck Surgery, King Abdullah International Medical Research Center, National Guard Health Affairs, King Saud bin Abdulaziz University for Health Sciences, Jeddah, [2]Department of Anatomic Pathology, King Faisal Specialist Hospital and Research Center, Jeddah, Saudi Arabia

Abstract

Primary cutaneous apocrine carcinoma (PCAC) is an extremely rare malignancy. Distinguishing apocrine carcinoma from breast carcinoma metastasis is difficult. Only a few cases reported as PCAC of the scalp and primarily treated by wide local excision. The usual presentation is a skin lesion that rapidly progresses over the duration of a few weeks to few months. We reported a 56-year-old man with a right scalp ulcerative lesion diagnosed as a CAC. The patient underwent wide local excision. 1.5 years later, the patient developed neck lymph node metastasis that treated with neck dissection and adjuvant radiotherapy. No chemotherapy was given due to limited literature, suggesting substantial benefits of adjuvant chemotherapy for such cases. Review literature was performed to assess the clinical presentation, treatment, and prognosis of such malignancies. PCAC of the scalp is a challenging malignancy in the diagnosis and management.

Keywords: Apocrine carcinoma, scalp, sweat gland, treatment

Introduction

Primary cutaneous apocrine carcinoma (PCAC) is a rare cutaneous malignancy with the incidence of 0.005–0.017 per 100,000 patients per year.[1] Approximately 200 cases were reported in the literature and most of those cases were originated in areas with large numbers of apocrine glands as the axilla. Only a few cases were originated from the scalp.[2,3] These malignancies have an equal distribution between males and females, with the peak of presentation in the 6th and 7th decades of age and the predominance to Caucasian ethnicity.[4] Due to the similarity of the morphological profile, it is difficult to differentiate from metastases breast adenocarcinoma. It is crucial to perform detailed clinical presentation and thorough examination to search of a primary site in such cases.[5] Frequently, it infiltrated the epidermal layer resulting in ulceration. PCAC has a diverse presentation, occurring as both uninodular and multinodular growths with varying color.[1,2] Often these tumors are indurated, painless masses, and can be associated with benign lesions, as a nevus sebaceous and most commonly seen with scalp lesions.[6] Development of these lesions typically occurs within a year before diagnosis.[7] However, several cases have reported longer durations with a period of rapid growth.[8] Of the 200 cases of PCAC, few cases have reported detailed accounts of scalp primaries.[1] We report the case of a 56-year-old man who presented with scalp ulcerative lesion treated with wide local excision. Later, he developed neck lymph node metastasis treated with surgery and adjuvant radiation. We review reported cases in the literature on the clinical presentation, treatment, and outcomes of such rare tumors.

Address for correspondence: Dr. Hadi Al-Hakami, MMedEd, KSAU-HS, KAIMRC, NGHA, P.O. Box 9515, Jeddah - 21423, Saudi Arabia.
E-mail: abuhussam004@gmail.com

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Methods

Case report

A 56-year-old male patient presented to head and neck clinic with an ulcerative lesion in the right temporal area of the scalp since the last 3 years that gradually increased in size and associated with mild pain. He denied any history of other skin lesion or swelling in the neck. He has no history of smoking, alcohol consumption, or sun exposure. On physical examination, the lesion was an ulcerative measuring approximately 3 × 3 cm. There was no palpable cervical lymphadenopathy. No pre- or postauricular lymphadenopathy. No other suspicious skin lesions of the face or the neck were found. The rest of the examination was unremarkable. Head and neck computerized tomography (CT scan) did not show any evidence of pathologic lymph nodes or masses. Staging with CT scan of the chest, abdomen, and pelvis was also negative for metastatic disease. The patient underwent complete local excision and split-thickness skin graft. Histopathology assessment revealed skin tissue infiltrated by neoplastic growth composed of tubules, cords, and nests of malignant cells. The extensive perineural invasion was seen. There was no lymphovascular invasion. The resection margins were clear. The pathology confirmed an apocrine cutaneous carcinoma [see Figure 1]. The case was reviewed in the head and neck tumor board and the decision of no adjuvant treatment. 1.5 years later, the patient presented to the combined head and neck oncology clinic with right neck lymphadenopathy for the last few weeks. On physical examination, there was around 2 × 2 cm mass in the right posterior neck looks like a lymph node. CT scan of the head and neck revealed 1.8 × 1.5 cm right neck lymph node at the level Va with central necrosis [see Figure 2]. FNA biopsy was taken and histopathology reported malignant cells to consist with metastatic of cutaneous malignancy. The patient underwent right modified radical neck dissection and the histopathology reported the diagnosis of metastatic cutaneous apocrine carcinoma. The case again discussed in the tumor board and the plan was to give adjuvant radiation therapy. No chemotherapy was given. During his follow-up for the last few years, the patient was from the disease on the clinical and radiological evaluation.

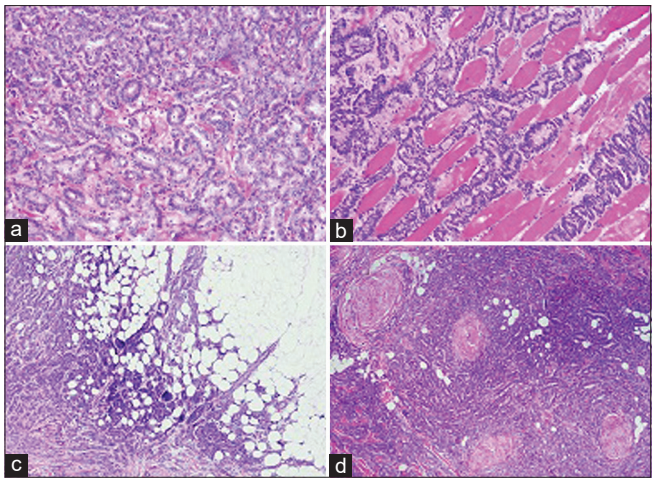

Figure 1: (a): Sheets of tumor cells composed of tubules, cords, and nests of malignant cells with high nuclear to cytoplasmic ratio and nuclear pleomorphism (H and E stain; magnification × 10); (b): Tumor cells invading skeletal muscles (H and E stain; magnification × 10); (c): Tumor cells extending deep into the fat and subcutaneous tissue (H and E stain; magnification × 4); (d): Extensive perineural invasion (H and E stain; magnification; ×4)

Discussion Summary of the Literature Review

We conducted a literature review through the PubMed engine to evaluate clinical presentation, the treatment options, and the prognosis for patients with PCAC of the scalp. Eighteen cases had detailed reports and were collected and summarized in Table 1.[1] We added our case, so the total is 19 cases. Out of the 19 cases, 11 females (57.9%) and eight males (42.1%) made up the cohort. The mean age was 57 years with a range from 20 to 85 years of age. The size of the scalp lesions varied among the cohort, ranging from 0.5 to 7.5 cm, with an average of 3.1 cm. There were seven cases, all of, with an average size of 2.2 cm at presentation. The average size of the metastatic lesions was 5.9 cm that was higher than the average size of non-metastatic lesions which was 4 cm or less. 12/19 (63.2%) patients were presented with the only local cutaneous disease. 3/19 (15.8%) patients were presented with cervical lymphadenopathy at the time of diagnosis. Metastatic disease was not present at diagnosis in any of the reported cases. No details of disease presentation were reported in four patients. The scalp PCAC lesions exhibited variable growth patterns among the 10 cases that had reported these details ranging from slow progressive to rapid and aggressive presentation. The primary treatment was the surgical excision (local complete vs. wide or radical), occurring in 18/19 (94.7%) patients. Details of the primary treatment were demonstrated in Table 2. Details regarding disease progression, local versus regional recurrence, and distant metastases were summarized in Table 3. For local recurrences without the involvement of regional lymph nodes, wide excision of the tumor was the main treatment. In case of regional lymph node metastasis, neck dissection or radiation or both were

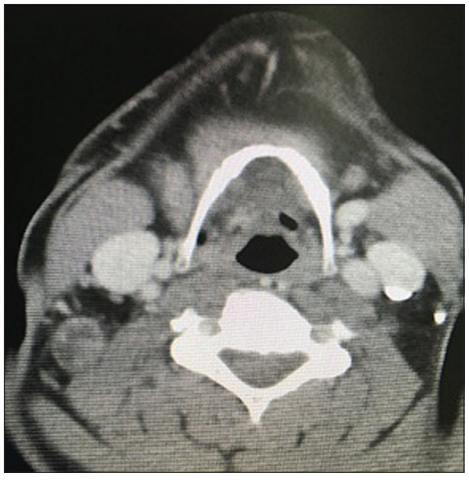

Figure 2: CT scan of the patient's neck revealed the right posterior triangle necrotic Lymph node measuring 1.8 × 1.5 cm

Table 1: Clinical Data of 19 Case Reports of Primary Cutaneous Apocrine Carcinoma of the Scalp. (Editing of the table that was done by Brown ZM *et al.* 2016 after permission)[1]

Reference	Age	Sex	Size (cm)	Time Evolved	Lymph Node Status at Diagnosis	Primary Treatment	Recurrence/Metastases and Treatment	Outcome (Follow-up)
Domingo and Helwig (1979)[6]	77	M	2×1.2	>17 y 1.5 m growth	NR	Excision (margins not specified)	1) LN Cervical (6 m) - ND 2) LR (1.5y) - Treatment NR	AWD (1.5 y)
Domingo and Helwig (1979)[6]	63	F	1.5	Birth	NR	Excision (margins not specified)	NED	NED (6 y)
Domingo and Helwig (1979)[6]	68	F	0.7	Unknown	NR	Excision (margins not specified)	NR	LTF
Domingo and Helwig (1979)[6]	65	M	7	Birth 1 m growth	NR	Excision (margins not specified)	1) LN Post-auricular, cervical and supraclavicular (6 m) - Treatment Radiotherapy 2) Distant Bone Mets. (9 m) - Treatment Radiotherapy	DWD (2 y)
Paties *et al.* (1993)[9]	85	M	3.5	20 y	NR	Excision (margins not specified)	LN Cervical posterior (2y) - Treatment NR	DOC (2.5 y)
Jacyk *et al.* (1998)[12]	54	F	4×1	Since Childhood Few month growth	No LN	Excision (margins not specified)	NED	NED (1 y)
Morbabito *et al.* (2000)[2]	46	F	NR	NR	NR	Excision (margins not determined)	1) LR (temporal) and LN Cervical (4 m) - Radical excision and ND, Radiotherapy, Chemo; 2) Cutaneous Mets. & Cervical LN (13 m) - Chemotherapy (17 m); 3) Cutaneous Scalp Lesion Progression (26 m) - Treatment NR	DOC (28 m)
Shimato *et al.* (2006)[13]	48	M	5	NR	Cervical LN	Wide excision (2 cm free margins) and ND	1) Lung Mets. (4y) - Chemotherapy; 2) Brain Mets. (6y) - Excision and Radiotherapy; 3) Brain recurrence (7y) - Excision; 4) Lung Mets. Progression (8y)	DWD (8 y)
Robson *et al.* (2008)[14]	73	F	0.5	NR	NR	Complete Local Excision (margins not determined)	NED	LTF
Robson *et al.* (2008)[14]	63	F	2.4	NR	NR	Complete Local Excision (margins not determined)	NR	LTF
Robson *et al.* (2008)[14]	70	F	1.9	NR	NR	Complete Local Excision (margins not determined)	NED	NED (2.5 y)
Robson *et al.* (2008)[14]	43	F	7.5	6 m	NR	Complete Local Excision (margins not determined)	Metastases (specific details of metastases and treatment not mentioned)	DWD (6 y)
Robson *et al.* (2008)[14]	31	M	1.4	3 m	NR	Complete Local Excision (margins not determined)	NR	LTF
Tlemcani *et al.* (2010)[10]	20	M	NR	Several Weeks	NR	Excision (margins not determined)	LR, LN, Bone and Lung Mets. (16 m) - Palliative Radiotherapy and Chemotherapy	DWD (55 m)
Kim *et al.* (2011)[15]	60	F	2×1.5	NR	NR	Chemotherapy	LR (7y) - Wide excision (2 cm free margins)	NED (8 y)
Paudel *et al.* (2012)[11]	45	M	2×2	Since Childhood 4 m growth	Cervical LN	Excision (margins not determined)	NR	NR

Contd...

Table 1: Contd...

Reference	Age	Sex	Size (cm)	Time Evolved	Lymph Node Status at Diagnosis	Primary Treatment	Recurrence/Metastases and Treatment	Outcome (Follow-up)
Vucinic *et al.* (2012)[5]	65	F	4	NR (Prior history of scalp mass & enlarged neck LN 3 years prior) (Treatment not reported)	Cervical LN	Radical Excision (2 cm free margins) and ND Chemotherapy and Radiotherapy	1) LR and LN (contralateral cervical) (10 m) -Re-excision & ND; 2) LR and LN (Cervical & Axillary) (12 m) - Re-excision, ND & chemo; 3) LN, Bone, Cutaneous & Lung Mets. (16 m) -Bisphosphonate & Supportive Therapy	DWD (3 y)
Brown ZM *et al.* (2016)[1]	42	F	3×2	NR	No LN	Local Excision (clear margins minimum 2 mm)	NED	NED (39 m)
Al-Hakami HA *et al.* (2018)	56	M	3×3	3 y	No LN	Local Excision (clear not specified margins) and STSG	LN (Cervical single right 1.8×1.5 cm) after 1.5 y - MRND & Radiotherapy	NED (2 y)

LN - Lymph Node; LR - Local Recurrence; ND - Neck Dissection; AWD - Alive with Disease; NED - No Evidence of Disease; LTF - Lost to Follow-up; DWD - Died with Disease; DOC - Died of other causes; NR - Not reported; MRND- Modified Radical Neck Dissection; STSG - Split Thickness Skin Graft.

Table 2: Type of primary treatment among 19 cases of PCAC of the scalp

Primary Treatment	*n* (%) (*n*=19)
Surgery	**18 (94.7%)**
Local Excision	16 (88.9%)
Wide/Radical Excision	2 (11.1%)
Radiotherapy	**1 (5.3%)**
Chemotherapy	**2 (10.5%)**

Local Excision - complete excision with clear margins of less than 2cm or undefined margins; Wide or Radical Excision - complete excision with clear margins of 2 cm or more.

Table 3: Details of disease recurrence and metastases of 19 case reports

Recurrence/Metastases Outcome	*n* (%) (*n*=19)
No Recurrence	5 (26.3%)
Local Recurrence	6 (31.6%)
Regional Recurrence	7 (36.8%)
Distant Metastases	5 (26.3%)
Not Reported	4 (21.1%)

the treatment in most of the cases. For the patients that had developed metastatic disease, combinations of chemotherapy and/or radiotherapy were the palliative treatment in most of the cases. Common sites of distant metastasis occurred in the distant lymph nodes as the axillary lymphadenopathy, the bones, the brain, and the lungs. From the time of metastatic diagnosis, survival ranged from approximately 1–4 years, with an average of 2.25 years.[1-3,9-11]

PCAC of the scalp is a rare malignancy most often reported in the literature as case reports or small case series.[1] Approximately 200 cases were reported in the literature and most of those cases were originated in areas with large numbers of apocrine glands as the axilla. It is difficult to distinguish between PCAC and metastatic adenocarcinoma of the breast as both of them have the same morphological features. Limited work has been done analyzing the treatment options, prognosis, and outcomes available for the various stages of this malignancy. In our review, we were able to identify 19 cases of scalp primaries. At initial presentation, most cases had localized disease while regional lymph node metastases were less prevalent. Data suggests larger primaries at initial presentation may indicate the poor outcome, due to their tendency to metastasize; prognosis is often fatal upon the evidence of metastatic disease.[1] Our data were consistent with previous demographic and prognostic findings from other PCAC primaries.[1] A review of 186 cases was done by Brown *et al.*, analyzing several PCAC primaries, showed that similar to scalp lesions, patients most often present with localized disease, while metastases to the lymph nodes and distant regions is less common.[1] For the treatment of localized PCAC, the current consensus tends to support the use of wide surgical resection. Due to insufficient data, surgical margins have not been standardized; however, 1–2 cm may provide sufficient eradication of tumor cells. According to the aggressive nature of larger scalp lesions exceeding 5 cm, some reports have suggested the use of adjuvant radiation in the treatment protocol to improve survival rates.[16,17] Based on the low risk of occult lymph node metastases, there is no need to do neck dissection in case of node-negative malignancy (N0). Neck dissection followed by adjuvant radiotherapy was offered to the patient presented with cervical lymph node metastasis. Hollowell[4] recommended the use of sentinel lymph node biopsy (SLNB) to guide treatment planning, but due to the low incidence rate of PCAC, SLNB has not undergone prospective evaluation. Due to the low incidence rate of PCAC, individualized treatment should be addressed. Chemotherapy should be reserved for treating the advanced disease that often proves to be fatal and the initiation of palliative care in these circumstances is inevitable.[3] The prognosis among scalp cases is limited due to such a small sample size; however, survival seems to correlate with data from various primaries. In our case, our patient remains disease-free at 24 months post-surgery. These results are consistent with previous case reports in the literature.

Relevance to the healthcare practice

Primary apocrine carcinoma of the scalp is a rare malignancy. Following an in-depth review of the literature on the scalp

PCAC, it can be concluded that the recommendation for wide surgical resection with cleared margins seems to be appropriate among patients with local, node-negative disease. One to two centimeters surgical margins are generally accepted standards. Patients with metastases regional lymph node have lower median survival rates and may benefit from neck dissection and additional radiation. The use of chemotherapy and radiation may also be considered in patients with the advanced and distant disease, as well as recurrence, as palliative but should be decided on a case-to-case basis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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