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CASE REPORT

Primary apocrine carcinoma of the axilla in a male patient: a case report

Rehan Zahid[1,2], Madiha E. Soofi[2,3], Hind Elmalik[2,4] & Kulsoom Junejo[2,5]

[1]Plastic and Reconstructive Surgery Department, Rumailah Hospital, Hamad Medical Corporation, Doha, Qatar
[2]Weill Cornell Medical College in Qatar, Doha, Qatar
[3]Pathology Department, Hamad General Hospital, Hamad Medical Corporation, Doha, Qatar
[4]Medical Oncology, National Center for Cancer Care and Research, Hamad Medical Corporation, Doha, Qatar
[5]General Surgery Department, Hamad General Hospital, Hamad Medical Corporation, Doha, Qatar

Correspondence
Rehan Zahid, Plastic and Reconstructive Surgery Department, Rumailah Hospital, Hamad Medical Corporation; and Weill Cornell Medical College in Qatar, Doha, Qatar. Tel: +97433640861; Fax: +97444425550; E-mail: rehanzahid@hotmail.com

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Key Clinical Message

Primary apocrine sweat gland carcinoma is a rare neoplasm. It is usually slow growing and is often suspected to be a benign disease at initial assessment. A thorough clinical and histological workup is required for diagnosis. Treatment of choice is wide local excision with clear margins.

Keywords

Axillary carcinoma, cutaneous apocrine carcinoma, sweat gland carcinoma.

Introduction

Primary apocrine carcinoma of the sweat gland is an extremely rare neoplasm with around 50 cases reported in the literature so far [1]. It's usually slow growing, giving the clinical picture of a benign lesion but can sometimes progress rapidly and aggressively [2–5]. Clinically, it often presents as painless, indurate, nodules, or plaques [6, 7]. Mostly it affects the axilla and anogenital region but it can also affect the scalp, eyelid, ear, lip, chest, nipple, fingers, and toes [1, 2, 4]. Apocrine carcinoma frequently affects the elderly population with a median age of 67 [1]. There is no racial or gender predisposition noted [4].

The established treatment is wide local excision with or without lymph node dissection depending on the node status, while the use of adjuvant chemotherapy remains controversial.

We present a 56-year-old male patient with papillary adenocarcinoma of the apocrine gland affecting the axilla, and review the existing literature about the disease and its management.

Case Report

A 56-year-old male patient was first seen in September 2013 in a foreign medical facility presenting with a right axillary mass with overlying skin inflammation unchanged for 1 year. A diagnosis of hidradenitis suppurativa was made, treated with antibiotics, and the patient was lost to follow-up. He presented again in December 2014 with the right axillary mass increased in size. The mass was 5 × 4 cm in size, attached to the overlying skin which was erythematous. There were no palpable lymph nodes or other masses. A clinical diagnosis of inflamed sebaceous cyst was made and the mass was surgically excised as a whole with the overlying skin. Postoperative recovery was uneventful. Histopathology of the specimen reported papillary adenocarcinoma with positive margins favoring

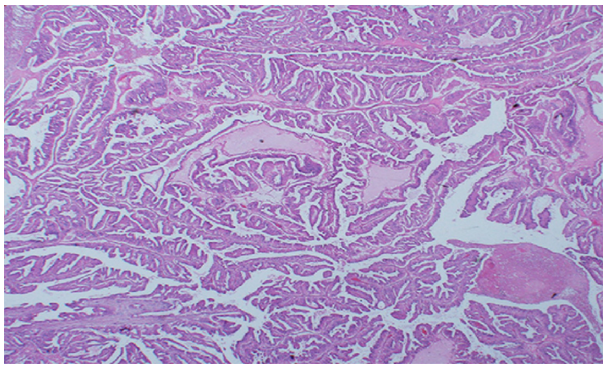

Figure 1. Low power (2x) view of the tumor with papillary architecture showing fibrovascular cores lined by eosinophilic epithelial cells.

a metastatic lesion. Patient was referred to us for further management.

Patient presented to us 7 weeks after the excision. He underwent a thorough workup. Patient's past history comprised of hypertension, dyslipidemia, and benign prostatic hypertrophy. He had a family history of his father diagnosed with lung cancer at the age of 75 and sister with breast cancer detected at the age of 60. He is a nonsmoker and nonalcoholic. On examination, the axillary wound showed normal postoperative healing with some firmness but no discrete swelling or signs of inflammation. The patient had no generalized lymphadenopathy, organomegaly, goiter, breast lumps, palpable axillary lymph nodes, skin lesions, or hyperpigmentation. ENT assessment including pan endoscopy showed no evidence of malignancy. PET–CT scan was negative except mild uptake of FDG at the site of surgery, which could be due to postsurgical changes. Breast ultrasound, mammogram, and MRI did not reveal any lesions either.

Microscopic reexamination for second opinion of the histopathology blocks at Hamad General Hospital revealed neoplasm with mostly papillary cystic architecture present in the dermis with epidermis evident in some sections. The papillary structures were lined by large apocrine cells with ample eosinophilic cytoplasm and large nuclei, some showing atypia (Fig. 1). However, the nuclear to cytoplasm ratio was low. Focal areas showed occasional mitosis. Prominent apocrine glands were noted close to the tumor in one section. Focal solid area showed glands with desmoplastic and inflamed stroma. Overall the lesion was well-circumscribed; however, it involved the inked margin focally. Areas of hemorrhage and necrosis were noted. No lymph node was identified. Immunohistochemistry with appropriate controls was positive in the tumor for CK7, E-cadherin, GCDFP, and focal CEA (Fig. 2). It was negative for S100, p63, ER, PR, TTF1, CK20, and PSA.

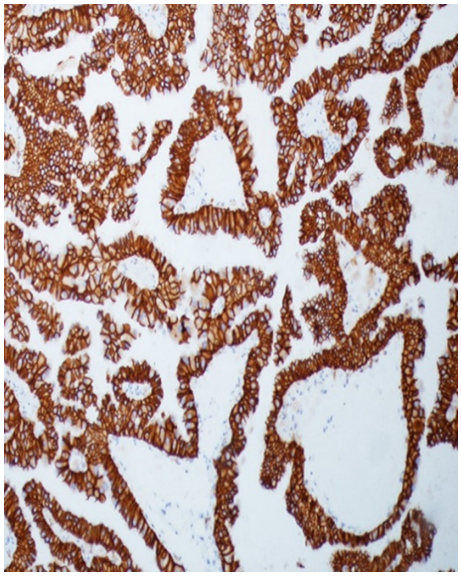

Figure 2. IHC was positive for GCDFP. In addition, E-cadherin and CK7 were also positive.

According to the pathology report, the differential diagnosis included metastatic carcinoma to the axilla. Based on the finding of a large single mass with prominent apocrine glands in its vicinity and no accessory breast tissue, primary apocrine sweat gland carcinoma of the axilla was favored as a diagnosis (Fig. 3). Due to the rarity of this tumor, histopathology blocks were sent to the Pathology Department of Mayo Clinic, Rochester, USA for another opinion, which labeled the specimen as a low-grade papillary adenocarcinoma compatible with an ectopic breast primary or possibly a cutaneous adnexal

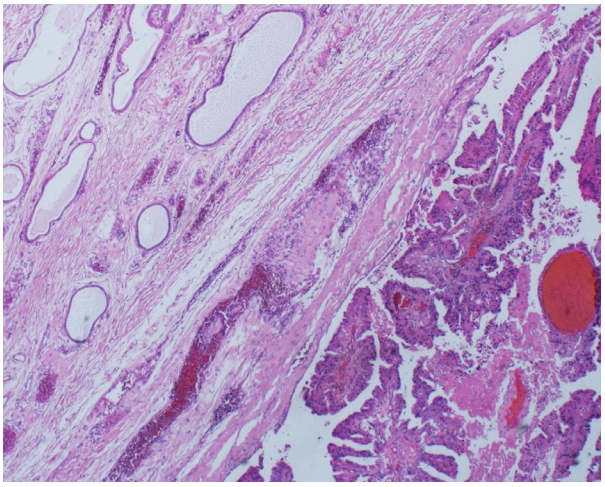

Figure 3. Prominent benign apocrine glands (left side) are noted close to the tumor (which is seen on the right lower side of the picture).

carcinoma. Her2 IHC stain was performed with appropriate controls, which was negative.

Patient was discussed with the Breast Multidisciplinary Team (BMDT) and planned for a surgical staging procedure. In view of the histopathology report and positive margins after the initial surgery at another institute, as well as regional node metastasis not being uncommon in published literature, the BMDT recommended axillary clearance. With regard to the rarity of the case and lack of an established management protocol, the axillary dissection was an acceptable approach to prevent recurrence and/or regional spread. Re-excision of margins was done with dissection of level one and two axillary lymph nodes 2 weeks after his presentation to our facility. Adequate clearance margin of 1–2 cm was taken and direct primary closure of the defect was done without much difficulty or the need for a rotation flap. Histopathology report showed no residual malignancy. Thirty-three lymph nodes were identified and all were negative for malignancy. Numerous benign apocrine glands with focal inflammation were noted. Given the negative margins and node status he was not considered for adjuvant radiotherapy. Patient had an uneventful recovery postoperatively. He was planned for 3 months follow-up for the first year and then annually. He has been evaluated regularly for 9 months since the re-excision. Our patient has not shown any indication of recurrence or metastasis of the disease to date.

Discussion

Primary apocrine sweat gland carcinoma is a rare neoplasm with no distinctive clinical features. It usually occurs as a single or multinodular mass, or plaque, and is often misdiagnosed [2, 8]. It is difficult to distinguish it from a metastatic breast or lymph node carcinoma and requires a thorough history and clinical evaluation for breast cancer along with a meticulous histopathological workup in such patients. In this particular case, the patient had a large single mass proximate to the epidermis with abundant apocrine glands in the vicinity and no evidence of ectopic breast tissue. The supporting microscopic examination and immunohistochemistry favored the diagnosis of primary apocrine sweat gland carcinoma rather than a metastatic breast carcinoma.

Review of literature supports high local recurrence and regional lymph node metastasis but sparse evidence of distant metastasis [1]. Metastasis has been reported in about 30% of cases [2, 8]. Regional lymph node metastasis is considered as an important prognostic factor. Median overall survival and 5 year disease-specific survival were reported as 51.5 months and 88%, respectively [4].

Diagnosis of primary apocrine carcinoma is based histologically on apocrine differentiation of the tumor and clinically on a usual anatomic location where apocrine glands are numerous [8]. The histologic picture is similar to adenocarcinoma that may be well, moderately, or poorly differentiated [3, 5]. The adenocarcinoma contains ductal or glandular structures with apocrine features. The cytoplasm of the tumor cells contains periodic acid–Schiff-positive, diastase-resistant granules and often iron-positive granules [3, 5, 9]. Robson et al. [2] suggested that apocrine carcinoma can be diagnosed mainly by its hallmark histologic features, such as abundant granular, eosinophilic cytoplasm with luminal decapitation secretion.

The treatment of choice is wide local excision with clear margins, with or without regional lymph node dissection depending on the node status. Postoperative radiotherapy and chemotherapy has shown little benefit on outcome in patients with moderately or poorly differentiated tumors [5]. The rarity of this type of carcinoma and the lack of clinical trials precludes set guidelines for the management of such cancers.

Conclusion

In conclusion, we present the first case of primary apocrine carcinoma in the Middle East. The rarity of this type of carcinoma and the lack of clinical trials precludes set guidelines for the management of such cancers. However, wide local excision with clear margins, with or without regional lymph node dissection has been established as the treatment of choice. Further reporting and literature review will help in establishing a diagnostic criteria and the most effective treatment modality.

Conflict of Interest

The authors declare they have no conflict of interest.

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