International Journal of Medical Science and Clinical Research Studies

ISSN (print): 2767-8326, ISSN (online): 2767-8342

Volume 04 Issue 12 December 2024

Page No: 2460-2463

DOI: https://doi.org/10.47191/ijmscrs/v4-i12-58, Impact Factor: 7.949

Primary Cutaneous Apocrine Carcinoma in an 82-Year-Old Male

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ABSTRACT

Primary cutaneous apocrine carcinoma is a rare adnexal malignancy, often presenting a diagnostic challenge due to its resemblance to metastatic ductal carcinoma of breast. We report the case of an 82-year-old male with a painless, progressively enlarging swelling in the left axilla over a year. Clinical impression was a sebaceous cyst. Following an excision biopsy, histopathology demonstrated a well-circumscribed, unencapsulated neoplasm in the dermis with apocrine differentiation. The hallmark feature of decapitation secretion was observed. Immunohistochemical studies with GCDFP 15, CK7, ER, PR, HER2 and S100 antigen supported the diagnosis.

The patient underwent complete excision and is doing fine without recurrence since January 2024. This case highlights the importance of meticulous morphological and immunohistochemical evaluation to distinguish primary cutaneous apocrine carcinoma from metastatic ductal carcinoma of breast.

KEYWORDS: Primary cutaneous apocrine carcinoma, skin adnexa, sweat gland, axilla, malignant adnexal neoplasm.

ARTICLE DETAILS

Published On:
31 December 2024

Available on:
https://ijmscr.org/

INTRODUCTION

Primary cutaneous apocrine carcinoma (PCAC) is a rare malignant adnexal tumour originating from apocrine sweat glands. Most commonly affecting older adults, PCAC typically arises in areas of high apocrine gland density, such as the axilla, and often presents as a painless, slow growing mass.

The clinical presentation of PCAC frequently mimics a sebaceous cyst. In addition, its histological features and immunohistochemistry can closely resemble metastatic ductal carcinoma of breast. Although most cases have a slow course, some may present with nodal or distant metastases, making the early detection vital for prognosis.

In this report, we describe the case of an 82-year-old male diagnosed with PCAC in the left axilla. We aim to highlight the distinctive features of this rare neoplasm and emphasize the diagnostic challenges it poses.

CASE REPORT

We would like to report a case of cutaneous apocrine carcinoma. An 82-year-old male presented with complaints of painless swelling in the left axilla of 1 year duration, which was progressively increasing in size. On physical examination a 2 x 2 cm firm mobile mass with surface induration was palpable in the left axilla. No breast mass was identified clinically.

Following this an excision biopsy was carried out. On gross examination, it is a skin covered nodulo-cystic mass with dimension of 1.5×1.5×1cm. Surface skin shows a grey white indurated area. On cut section, mass shows a circumscribed grey white lesion with faint lobulations and a cystic area filled with solidified yellowish green amorphous material (*Figure.1*).

Figure 1: grey white lesion with faint lobulations and cystic area filled with yellowish green material

On histopathological examination, skin with epidermal ulceration shows a circumscribed unencapsulated neoplasm in the deep dermis, composed of cells arranged in papillary, tubular, solid trabecular and cystic pattern (*Figure 2*). Individual cells are columnar with granular eosinophilic cytoplasm and basally placed enlarged ovoid vesicular nucleus with clumped chromatin and prominent nucleoli (*Figure.3*). Mitosis is brisk. Occasional cells show decapitation secretion. Tubules and cysts are seen filled with amorphous eosinophilic material. Stroma is inconspicuous. Periphery of the neoplasm shows dense lymphocytic infiltration. Margins were free.

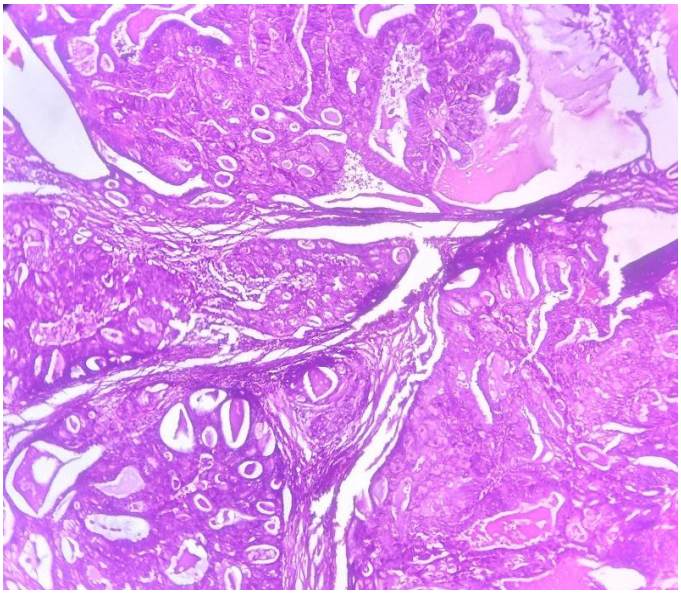

Figure 2: tumour cells arranged in papillary, tubular, solid trabecular and cystic pattern.

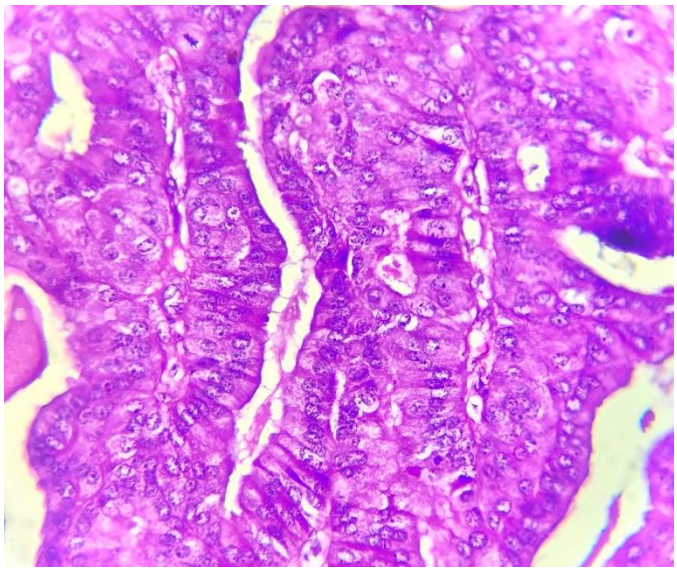

Figure 3: columnar cells with granular eosinophilic cytoplasm and enlarged ovoid vesicular nucleus with clumped chromatin and prominent nucleoli.

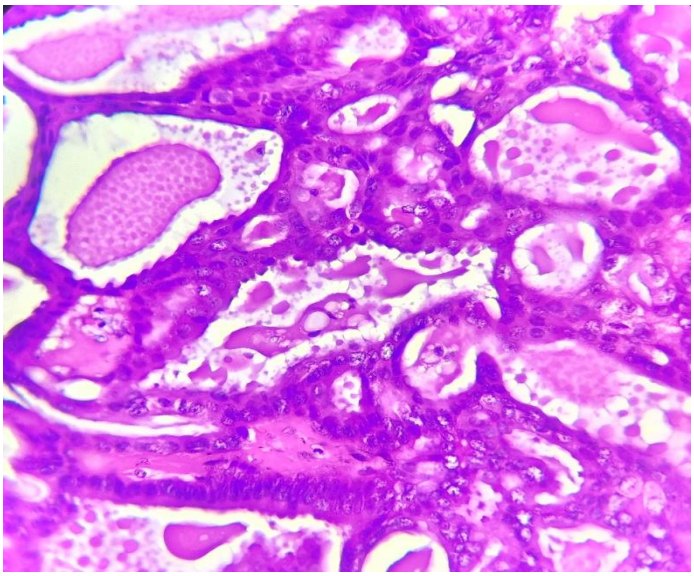

Figure 4: cells showing decapitation secretion.

On immunohistochemical examination *(figure. 5-10*), staining was positive for GCDFP 15 and CK7. Stains for S-100, ER, and PR was negative. HER2Neu showed unequivocal expression. Patient had uneventful post-operative period and was discharged and is currently doing fine without recurrence since January 2024

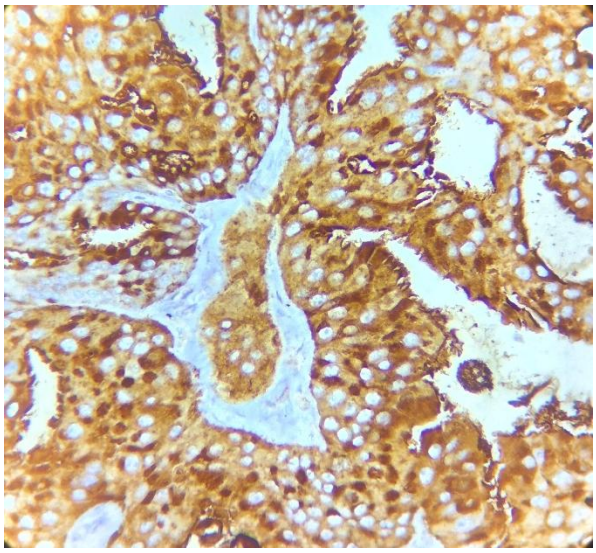

Figure 5: GCDFP 15 shows positive expression.

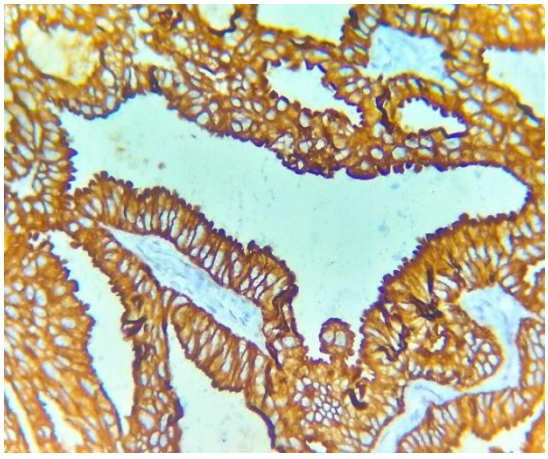

Figure 6: CK7 shows positive expression.

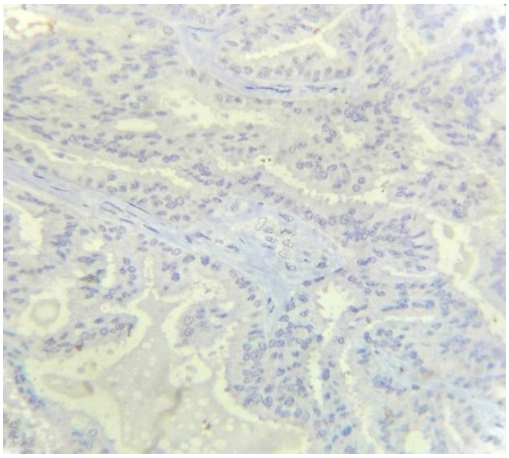

Figure 7:S100 with negative expression.

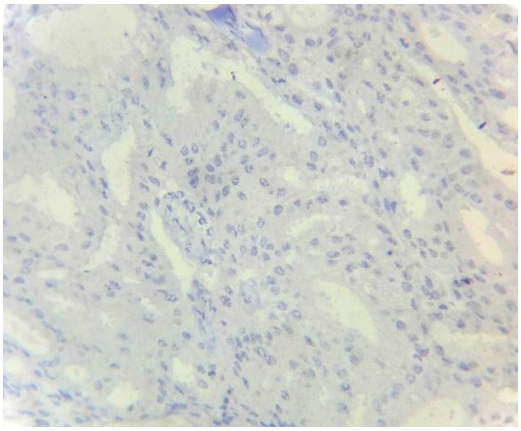

Figure 8: ER showing negative expression

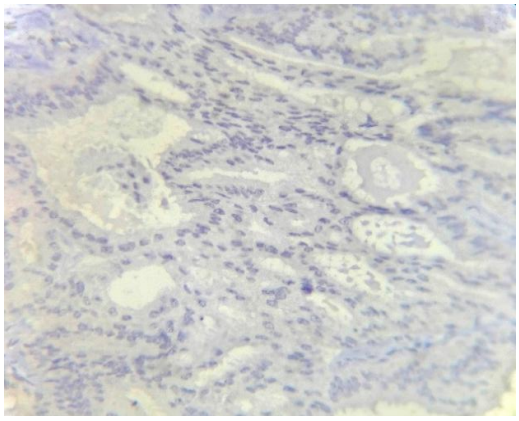

Figure 9: PR showing negative expression.

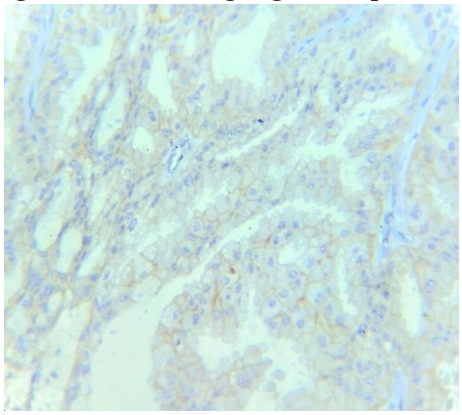

Figure 10: Her2/neu shows weak incomplete membrane staining, score 1+ - negative.

DISCUSSION

Primary cutaneous apocrine carcinoma is an extremely rare malignant adnexal neoplasm of old age group, arising in high sweat gland density areas. With an incidence of 0.0049 to 0.0173 per 100,000 patients per year, only about 200 cases are reported in the literature till now (1,5). The median age of affected is 67 years and it shows equal preponderance for males and females (2). Most of the cases arise de novo but some have shown association with apocrine adenoma, apocrine hyperplasia, cylindroma and nevus sebaceus (4). Most common location of this tumour is axilla, making it a challenge to distinguish from metastatic apocrine carcinoma of breast (3). Patients usually present with a painless slow growing uninodular or multinodular mass (2). The growth period of the reported cases ranges from several weeks to 40 years (4). The surface skin of the tumour may show variable colour, induration, and ulceration (2). Even though most patient present with an asymptomatic mass, some have lymph node metastasis or distant metastases at presentation. Sites of metastases reported are lungs, bone, brain, and parotid gland. Relative asymptomatic presentation and slow growth of the tumour rarely raises suspicion of malignant neoplasm and are usually thought as benign preoperatively (4).

Histologically, tumour usually shows features characteristic of apocrine differentiation. Well differentiated cases show ductal structures and glands, which varies from papillary, cord like, trabecular, solid, and complex glandular patterns. The cells are large, have abundant granular eosinophilic cytoplasm and hyperchromatic nuclei. Characteristic luminal apocrine decapitation secretion seen in these tumours is a hallmark of apocrine differentiation (3,4). Vascular invasion suggests probability for lymph node metastasis. These tumours exhibit Intracellular and extracellular mucin and it is often PAS positive and diastase resistant (4). An in-situ component can be seen in some cases and this reinforces the diagnosis of primary PCAC (2). Poorly differentiated cases do not show obvious decapitation secretion and show higher grade of cellular pleomorphism. Presence of focal glandular differentiation, acrosyringial involvement and pagetoid spread within epidermis are some clues that help in diagnosis of poorly differentiated cases (3).

PCAC shows histologic similarity with metastatic breast carcinoma and in few cases with other tumours like metastatic bladder or salivary gland carcinoma (2). High grade poorly differentiated tumours can mimic metastatic adenocarcinoma of any organ (3). Immunohistochemistry aids for differentiating these tumours but significant overlap is seen between them, particularly metastatic breast carcinoma. So, the final diagnosis should always be backed with clinical and radiologic data. An eight test IHC panel is recommended by Fernandez-Flores group to differentiate PCAC from metastatic breast carcinoma, which includes p63, GCDFP-15, calretinin, D2-40, ER, PR and mammaglobin (2). PCAC shows positive expression for GCDFP 15, CK7, GATA 3, AR, CK 5/6 and EMA in most cases. Variable expression is seen with ER, PR, CEA, Her2, Mammaglobin and S100 staining and usually a negative expression is seen with p63, D2-40, CK20, and TTF1. In-situ

component can be identified with peripheral staining of basal and myoepithelial antigens like calponin and SMA (2,3,4,5,6).
No significant recurrent genetic alterations are defined for primary cutaneous apocrine carcinoma. Although hidradenocarcinomas show CRTC1-MAML2 fusion, no evidence of such is seen in PCAC (8).

Prognostic factors are not well established for this entity as only limited cases are reported. Due to its striking similarity with breast carcinoma, it has been proposed that, The Bloom-Richardson grading system for breast carcinoma may be used to determine treatment. Studies have shown that there is a statistically significant difference in survival between grade 3 tumours and grade1/2 tumours (9). Along with higher grade of tumour, positive lymph node status, positive margins and distant metastases suggests worse outcomes (5).

Treatment modalities include surgery, chemotherapy, and radiotherapy (7). Wide local excision without any adjuvant therapy is the treatment of choice for low grade and localized disease. A sentinel lymph node biopsy followed with lymph node dissection in positive cases is suggested to avoid poor prognosis. Studies propose that adjuvant radiotherapy may be offered for large tumours with high grade and for those with positive margins or lymph node involvement. Adjuvant chemotherapy using breast cancer regimens is proposed in some research for patients with poor prognostic factors. Endocrine therapies in ER positive tumours and targeted therapies for patients with HER-2 amplification, positive RANK-L tumours, and positive PDL-1 tumours has also been proposed (5).

CONCLUSION

Primary cutaneous apocrine carcinoma is a rare malignant neoplasm with an indolent course in most cases. The challenges in diagnosing PCAC are its rarity, lack of definite diagnostic criteria and its histologic and immunohistochemical overlap with metastatic breast carcinoma. A thorough clinical, radiological, and histopathological examination, along with aid of immunohistochemistry will warrant a definite diagnosis.

ACKNOWLEDGEMENTS

The authors wish to thank Oncore diagnostics, Bangalore for their IHC services.

CONFLICT OF INTEREST

None declared.

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