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CASE REPORT

A rare case of primary cutaneous apocrine carcinoma of an unusual site

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ABSTRACT
Primary cutaneous apocrine carcinoma (PCAC), a subtype of sweat gland carcinoma, is an extremely rare malignant tumor that originates from either normal or modified apocrine glands and affects both genders equally, with no racial predilection. Most of these tumours arise in regions of high apocrine gland density, especially in the axilla and, to a lesser extent, in the anogenital region, and rarely in the scalp, face, chest, and distal upper extremities. This neoplasm is often difficult to diagnose from metastatic breast carcinoma. This requires clinic-radiological and histological correlation for final diagnosis.
Key words: Apocrine carcinoma, GCDFP-1, clinco-radiological

INTRODUCTION

Primary cutaneous apocrine carcinoma (PCAC), a subtype of sweat gland carcinoma, is an extremely rare malignant tumor that originates from either normal or modified apocrine glands and affects both genders equally, with no racial predilection. [1,2] Most of these tumours arise in regions of high apocrine gland density, especially in the axilla and, to a lesser extent, in the anogenital region, and rarely in the scalp, face, chest, and distal upper extremities.[2,3] Frequently,PCAC is indolent and slowly developing, but some are rapidly progressive and aggressive and mostly present as masses of 2-3cm in size but may also present with regional lymph node metastasis.[4] The clinical diagnosis of PCAC is challenging and necessitates histomorphological and immunohistochemical study and familiarity with the characteristics should help prevent misinterpretation. [1,5]Wide, local excision with clear marginswith or without lymph node dissection and/or sentinel lymph node biopsy remains the treatment of choice. [5]

CASE REPORT

We present a case of a 50-year-old lady who presented to Surgery outpatient department with a painful growth over right upper chest since last 5 to 6 years. The swelling was initially small (pea sized) and painless but started growing in size with surface ulceration for the last 15-20 days. On local examination, an ill-definedswelling of size 6x3cm was present over the right upper quadrant of the chest-infraclavicular area. The skin surface showed ulceration measuring 2x2cm.Rest of the systemic examination was within normal limits.

All routine haematological and biochemical investigations were within normal limits. Ultrasonography of bilateral breasts showed normal fibro-glandular tissue with normal nipple areola complex. No evidence of metastasis or axillary lymphadenopathy were seen. CECT chest showed a heterogeneously enhancing lobulated polypoidal growth in skin and subcutaneous plane involving right supra and infraclavicular region. Wide local excision of the mass was done and the specimen was sent for histopathological examination.

Grossly, the specimen included a skin covered tumor measuring 8x5x3cm.The skin surface showed an exophytic polypoidal ulcerated growth measuring 6x5cm. Cut sections showed a well circumscribed greyish white growth measuring 6x5x2.9cm.

Microscopically, showed keratinised stratified squamous epithelium with areas of ulceration. The underlying dermis shows an invasive malignant tumor arranged in tubules, cords and trabeculae. The cells are moderately pleomorphic with polygonal to oval to angulated cells with fine nuclear chromatin, prominent nuclei and moderate to abundant eosinophilic granular cytoplasm. Abundant

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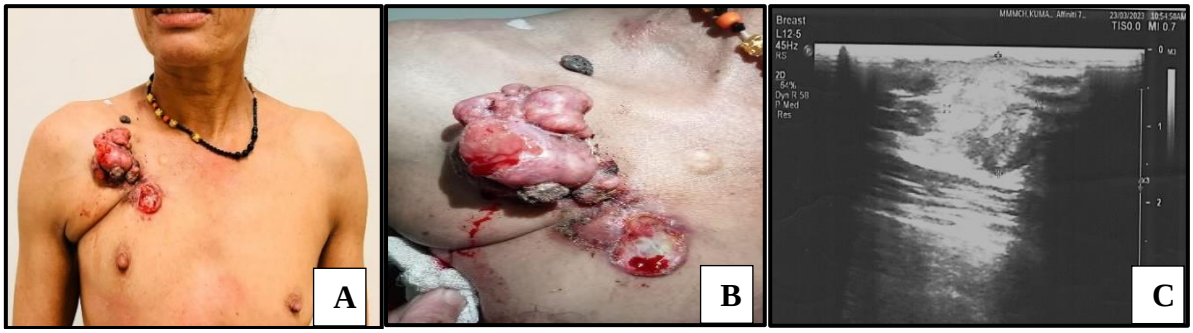
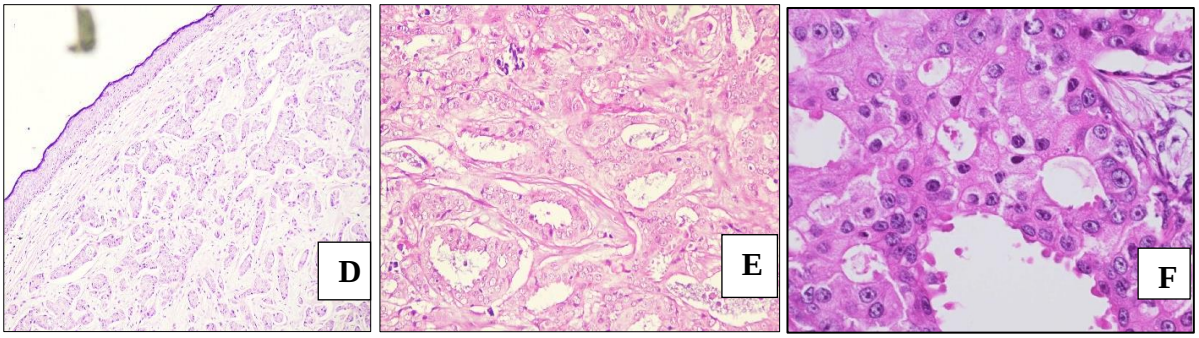
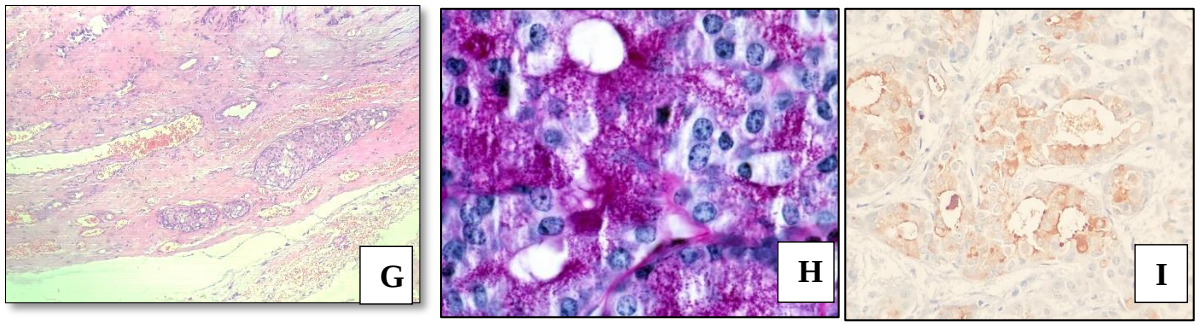
decapitated secretions and apical snouts were also present. There was moderate stromal desmoplasia. Lympho-vascular invasion was also present. Mitotic activity was increased. Fociof dystrophic calcification and PAS-positive eosinophilic material was also seen in the lumen of the glands. The tumor is infiltrating into the underlying fat. All the surgical resection margins were free of tumor involvement. Immunohistochemistry was done to support the diagnosis which revealed focal positivity for GCDFP-15. ER, PR and CK5/6 were negative. However, immunohistochemistry was overlapping with metatstatic breast carcinoma, with the above morphological features and clinco-radiological correlation final diagnosis of Primary cutaneous apocrine carcinoma was given.



A(Photograph) showing ulcerated and polypoidal lesion at infraclavicular region of the chest.B (Photograph) revealing a closer look of the tumor with adjacent ulcerated area.C(Radiograph) showing a hypoechoic lesion -? Likely neoplastic.



D(Photomicrograph,200x) Showing an invasive tumor lined by keratinised stratified squamous epithelium.E (Photomicrograph,200x)Revealing tumor arranged in tubules with decapitated secretions separated by desmoplastic stroma. F(Photomicrograph,400x)Showing tubule with apical snout.



G(Photomicrograph,200x) Showing Lymphovascular Invasion. H(Photomicrograph,200x)Tumor revealingcytoplasmic positivity for PAS-D. I(Photomicrograph,400x) Showing nuclear immunopositivity for GCDFP-15.

DISCUSSION

Primary cutaneous apocrine carcinomaisa rare adnexal neoplasm with less than 200 documented cases in the literature. It was first reported by Horn in the year 1944. It originates from either normal or modified apocrine glands and affects both genders equally, with no racial predilection.[1,2] The age presentation usually ranges from fifth to seventh decade of life. [1]The estimated incidence rate ranges from 0.0049 to 0.0173 per 100000 patients per year.[4]These tumours arise in the regions of high apocrine gland density, especially in the axilla and, to a lesser extent, in the anogenital region, and rarely in the scalp, face, chest, and distal upper extremities. [2,3]

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Frequently, these tumours are indolent and slowly developing, but in rare occasions can be rapidly progressive and aggressive. Up to one-third of cases have lymph node metastasis at the time of diagnosis.Because of their indolent course, apocrine carcinomas are diagnosed after many years and the patients have distant or localized lymph node metastases when they first show. There is a chance of metastasis to the lungs, liver, bone, brain and kidney. (3,4,5,6)

Histological examination reveals marked variation in the histologic patterns. The tumor reveals characteristics similar to apocrine glands. It includes papillary, complex glandular, anastomosing tubular, solid cellular sheets, or cord-like infiltration with desmoplasia. The tumor cells have large, polygonal, eosinophilic, or rarely, clear, granular, occasionally vacuolated cytoplasm, a large vesicular nucleus with chromatin in large clumps and a prominent nucleolus. Decapitation secretion in the luminal cells is seen. (4-5,7). In our case scenario this tumor presented at the rare location, which makes it a rarest of the rare kind. The main treatment is wide local excision with clear safety margins. There were no lymph nodes involved which was confirmed by ultrasonography and computed tomography. Patient was kept on follow-up for 6 months, there was no recurrence or metastasis.Clinico-radiological evidence is the most important clue in addition to histopathological examination to distinguish between PCAC and metastatic mammary carcinoma with apocrine differentiation.

CONCLUSION

Primary Cutaneous Apocrine Carcinoma is otherwise indistinguishable from apocrine mammary carcinoma metastatic to the skin or apocrine carcinomas arising in the ectopic breast tissue in the axilla. It is a challenging diagnosis for a pathologist due to its rarity, and lack of distinct clinico-pathological features, along with overlapping immunohistochemical findings with other tumors.

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