CASE REPORT

Primary apocrine carcinoma of an unusual site

Archit Gupta,¹ Mudita Gupta,² Kavita Mardi,³ Jagdish Gupta⁴¹Department of General Surgery, Indira Gandhi Medical College, Shimla, India²Department of Dermatology, Indira Gandhi Medical College, Shimla, Himachal Pradesh, India³Department of Pathology, Indira Gandhi Medical College, Shimla, India⁴Department of General Surgery, Indira Gandhi Medical College, Shimla, India

Correspondence to

Dr Archit Gupta,
archit9th@gmail.com

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SUMMARY

Primary apocrine carcinoma is a rare malignancy most commonly occurring in apocrine dense areas like axilla. There are only about 200 cases reported to date. We report a case of primary apocrine carcinoma present at an unusual site, that is, the arm. A wide local excision of the mass was done and was diagnosed as apocrine carcinoma on histopathological examination and was confirmed by immunohistochemistry. Wide local excision is the treatment required.

BACKGROUND

Primary cutaneous apocrine carcinoma (PCAC) is an extremely rare malignancy with only about 200 cases reported to date.¹ It most commonly occurs in apocrine dense areas such as the axilla and the anogenital areas² but has also been reported to arise from scalp, wrist, forehead and ear canal.³ The incidence rates of PCAC are estimated to range from 0.0049 to 0.0173 per 100 000 patients per year.⁴ We hereby report a case of PCAC of the arm who was treated by a wide local excision of the tumour.

CASE PRESENTATION

A 73-year-old male patient presented to us in surgery outpatient department with complaints of swelling in left arm since 5 years. The swelling was initially small but had started growing in size for the last 6 months. There was associated history of pain over the swelling from the last 1 month, which was dull aching, mild to moderate in intensity used to get aggravated with work. There was no other significant history. There was no family history of any similar lump or any malignancy. On local examination, an ill-defined swelling of size 3×3 cm was present on medial aspect of left arm approximately 7 cm from shoulder joint. It was irregular in shape with nodular surface. Skin over the swelling was intact with no surface changes or erythema. There was no evidence of muscle wasting and no evidence of similar lump anywhere else in the body. There was no evidence of generalised lymphadenopathy, axillary lymphadenopathy, organomegaly or any breast lump.

INVESTIGATIONS

All routine haematological and biochemical investigations were within normal limits. MRI of left arm had evidence of altered signal intensity lobulated mass of size 2.1×1.1 cm with craniocaudal extent of 3.2 cm seen in medial aspect of upper one-third of left arm in subcutaneous plane (figure 1). The mass was isointense to muscles on T1WI and

heterogeneously hyper intense on T2 and short TI inversion recovery images. The mass was causing buckling of underlying muscles of arm, though the fat planes were well maintained with the muscles.

Fine-needle aspiration cytological examination gave a possibility of malignant mesenchymal tumour left arm.

TREATMENT

With a provisional diagnosis of soft tissue tumour, the patient was taken up for surgery under general anaesthesia and a wide local excision of the mass was done. The excised tissue was sent for histopathology.

OUTCOME AND FOLLOW-UP

Histopathology revealed apocrine tumour cells with abundant granular eosinophilic cytoplasm with eccentric round vesicular nuclei with prominent nucleoli suggestive of primary apocrine carcinoma (figure 2). Immunohistochemistry revealed positivity for GCDPF-15. Oestrogen and progesterone receptors were not found.

DISCUSSION

PCAC is an extremely rare malignancy arising in areas of high apocrine sweat gland density.² There are only about 200 cases of PCAC to date. The first case was reported in 1944 by Horn.¹ This tumour has been more commonly reported in men in fifth to seventh decade of life. These lesions most commonly occur in the axilla but may also arise in the scalp, forehead, eyelid or arm.

PCAC is usually suspected to be a benign disease on initial clinical examination, leading to inadequate treatment and extensive metastasis.³ It usually arises de novo but has also been reported to arise in association with other benign tumours such as apocrine adenoma.

Most of these tumours are slow indolent tumours but can sometimes be rapidly progressive and aggressive also.⁵ They mostly present as masses of 2–3 cm in size but may also present with regional lymph node metastasis or even distant metastasis. These tumours cannot be diagnosed clinically as they only present as slow growing masses. Histopathological diagnosis is the investigation, which helps us to suspect the entity which can be confirmed by immunohistochemistry. PCAC has to be differentiated from metastatic deposits from adenocarcinoma breast. The latter may show positivity for oestrogen and progesterone receptor. In addition to these, PCAC may show higher positivity for CK5/6 and CK7. These immune markers were

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Figure 1 MRI of the patient showing a mass in the medial aspect of upper arm.

not done because of non-affordability by the patient. In addition, our patient did not have any lump in breast. This tumour can metastasise into regional lymph nodes; distant spread has been rarely reported.

Histological examination reveals characteristics similar to apocrine glands. It has ductal or glandular structures just like adenocarcinoma and can be well, moderate or poorly differentiated. Typically, it is composed of large

Figure 2 Histopathological image showing apocrine tumour cells.

cells with eosinophilic cytoplasm, which commonly has Periodic acid-Schiff (PAS) positive, diastase-resistant material.⁶ These cells also have hyperchromatic nuclei, mitotic figures and apocrine decapitation secretion. Immunohistochemistry may show positivity for GCDPF-15 and CD-15.⁷

The treatment of choice in primary apocrine carcinoma is wide local excision of the mass with margins of 1 to 2 cm with or without lymph node dissection.⁸ Sentinel lymph node biopsy can be done in patients with clinically negative nodes. There is not much role of postoperative chemotherapy or radiotherapy, though it has been used in moderately or poorly differentiated tumours.⁸ Antioestrogen therapy has also shown successful results as an adjuvant therapy in some case reports.⁹ Due to rarity of the tumour, no specific guidelines are available for treatment until now.

Learning points

- Primary apocrine carcinoma is an extremely rare malignancy and only about 200 cases have been reported to date.
- It most commonly occurs in apocrine-rich areas like the axilla but has also been reported from scalp, eyebrow or arm.
- Mainstay of diagnosis is histopathology, which shows characteristics similar to apocrine glands.
- Immunohistochemistry may show positivity for GCDPF-15 and CD-15.
- Treatment of choice is wide local excision with or without lymph node dissection. Adjuvant chemotherapy/radiotherapy can be used.

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