Primary Cutaneous Mucinous Carcinoma : A Case with Diagnostic Dilemma

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INTRODUCTION

Primary cutaneous mucinous carcinoma (PCMC) is a rare slow-growing neoplasm of the skin with an indolent behaviour. PCMC is slightly more common in men than women and typically affects people at 50 to 70 years old. The usual clinical presentation of PCMC in adults is asymptomatic, solitary erythematous nodule, papule, or cyst of variable sizes typically occurring on the face, scalp or in the axilla. Histologically, PCMC is indistinguishable from metastatic mucinous adenocarcinoma of breasts, gastrointestinal tracts, lungs, and salivary glands. This case report presents a case of PCMC, mimicking a skin metastasis from a male breast cancer.

CASE PRESENTATION

A 43-year-old gentleman had small bowel resection for small bowel Neuroendocrine Tumour (NET). He underwent a standard contrast-enhanced CT scan for staging at another hospital that revealed matted and enlarged lymph nodes at the left axilla with two small enhancing nodules in the upper-outer quadrant of the left breast. Subsequently, the whole-body positron emission tomography CT (PET-CT) scan confirmed the presence of high activity within the above-mentioned left axillary matted lymph nodes. Detailed history and clinical examination revealed that he had a painless left axillary skin lesion. He had it for many years and was not bothered by it. Therefore. he did not seek any medical advice. Recently, he noticed minimal serous discharge over this lesion after the Covid booster vaccination. Clinical examination showed a wart-like skin lesion (2x2 cm) at the medial aspect of the left axillary region, with multiple matted enlarged left axillary lymph nodes. The other axilla was unremarkable and there were no abnormalities on bilateral breast examination.

RADIOLOGICAL FINDINGS

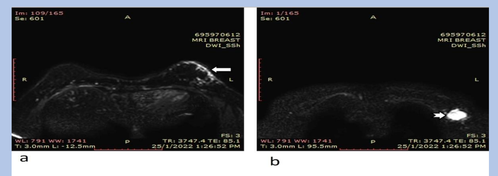


Figure 1 :
a) and b) DWI images showing prominent left intramammary lymphatic channels (arrow) and left axillary lymphadenopathy (notched right arrow), demonstrating restricted diffusion.
c) Post-gadolinium image and d) a corresponding DWI image, revealing no enhancement of the intramammary lymphatic channels and the skin (arrow), as well as the intramammary lymph node (notched right arrow) which is restricted on DWI.

HISTOPATHOLOGICAL FINDINGS

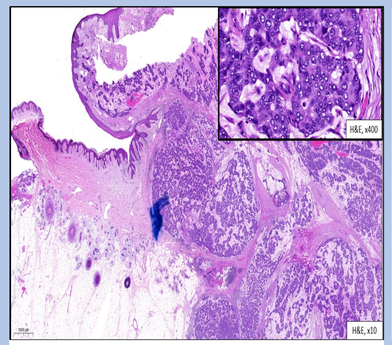


Figure 2. At the low power view, the tumour is seen at dermis, exhibits lobulated contour with expansile growth pattern. Adjacent normal skin with its adnexal structures is noted on the left (H&E, x10). The insert on the upper right shows moderate nuclear pleomorphism of the malignant cells (H&E, x400).

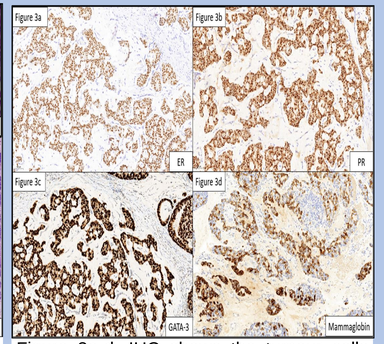


Figure 3a-d. IHC shows the tumour cells are positive for ER, PR, GATA-3, and Mammaglobin. (Magnification: x100)

CONCLUSION

PCMC is a rare tumour with challenging clinical presentation and histological diagnostic dilemma. Therefore, a multidisciplinary team approach with strong clinical, radiological and pathological correlation is warranted to differentiate PCMC from a case of metastatic breast carcinoma, as the morphological and IHC findings may overlap.

DISCUSSION

- PCMC of the axilla is a very rare tumour and only a few cases reported on this site[4].
- Morphologically, it is impossible to differentiate PCMC from metastatic mucinous adenocarcinomas of non-cutaneous organs such as gastrointestinal tract and breast[3].
- Challenges arise when the site of the tumour is within the chest and axillary region as these locations are strongly related to mammary origin[7]. Thus, the differential diagnosis would be PCMC versus metastatic breast carcinoma.
- To date, no immunohistochemical (IHC) panel has been identified to differentiate primary from metastatic mucinous carcinoma consistently.
- Immunohistochemistry is not particularly useful in distinguishing PCMC from mucinous adenocarcinoma of the breast as both tumors can express CK7, GATA 3, mammaglobin, estrogen receptor (ER), and progesterone receptor (PR)[3].
- Thus, the IHC staining cannot specifically diagnose PCMC. Hence the diagnosis was more challenging to confirm whether this lesion is a skin metastasis from occult breast cancer or PCMC[1].
- Generally, PCMC has a favorable good prognosis. It is a slow-growing tumor and have an indolent behavior[3] and often appears well localized. .
- PCMC is a rare disease and hence there is no established standard of management for it. Wide Local Excision (WLE) with or without regional lymph node dissection is the treatment of choice. As local recurrences are common, excision with 1-cm margin is recommended[4].
- Although PCMC has low metastatic potential (3%–11%), the local recurrence rate after surgery is high (29%–34%). Late recurrence and metastases have been reported probably related to incomplete excision of the tumor[9]. Such recurrences and metastatic disease have not responded well to treatment with radiotherapy or chemotherapy because they are highly resistant[9].
- There is limited data in the literature regarding the benefit of adjuvant endocrine therapy in PCMC. Tamoxifen has been successfully used in a few cases of metastasizing eccrine adenocarcinoma[12 13] and several case reports[10 11] have demonstrated the benefits of the endocrine therapy with an aromatase inhibitor for hormone receptor positive advanced PCMC. Knowing the fact that PCMC frequently expresses ER and PR, it might respond to endocrine therapies.

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