

Case report

A rare case of pigmented mammary Paget's disease without underlying malignancy

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Abstract

Mammary Paget's Disease is reported in 1-3% of all breast cancers. More than 90% of them are associated with underlying malignancy. Pigmented variant of the condition is a very rare entity which is often misdiagnosed as malignant melanoma due to its clinical resemblance. Here we present an extremely rare case of pigmented mammary Paget's disease without underlying malignancy that was managed with conservative surgery.

Key words: mammary Paget's disease without malignancy, pigmented Paget's disease

Introduction

Mammary Paget's Disease [MPD] is an uncommon form of breast cancer mostly associated with underlying ductal carcinoma in situ [DCIS] or invasive breast cancer¹. MPD without underlying breast cancer is rare, and the pigmented variant of MPD is extremely rare, mimicking cutaneous melanoma both clinically and histopathologically². We present here a case of Pigmented Mammary Paget's Disease [PMPD] without underlying malignancy.

Case report

A 60-year-old woman presented with a pigmented lesion on the right breast, gradually enlarging over five years. She had no similar lesions elsewhere on her body and was otherwise in good health with no family history of malignancies.

On examination, she had a 4 cm × 5 cm ill-defined, irregular, indurated lesion over nipple areola complex with thin plaques of hyperpigmentation

(Figure 1A and B). There was no nipple retraction, oozing, ulceration, or bleeding. No palpable masses were detected in both breasts, and both axillae were clear. Systemic examination was normal.

Dermatoscopically, the lesion revealed blue-gray areas, brown dots, and globules distributed in an irregular and reticular pattern. Tentative diagnoses included melanoma, breast CA, pigmented basal cell carcinoma, and morphoea.

A punch biopsy of the lesion was performed. Histology showed large cells with hyperchromatic nuclei, vacuolated, abundant cytoplasm. [Figure 1C Hematoxylin and Eosin × 200 and 1D H&E × 400 pagetoid cell shown by arrow] Some cells contained melanin granules within the cytoplasm and also in the dermis.

Immunohistochemistry revealed positivity for CK-7 marker, favouring the diagnosis of MPD. HMB-45 was negative, ruling out melanoma. HER 2 was also positive.

Further investigations included an ultrasound scan of the breast, mammogram, MRI [Magnetic Resonance Imaging] scan of the breast, and a CT scan of the chest, abdomen, and pelvis. All of the above tests showed no evidence of underlying malignancy. Serum biochemistry was normal.

The patient underwent a wide local excision. Histology of the specimen was compatible with pigmented Paget's disease of the nipple areolar complex, and the remaining breast tissue was free of atypical ductal hyperplasia, DCIS, or invasive malignancy.

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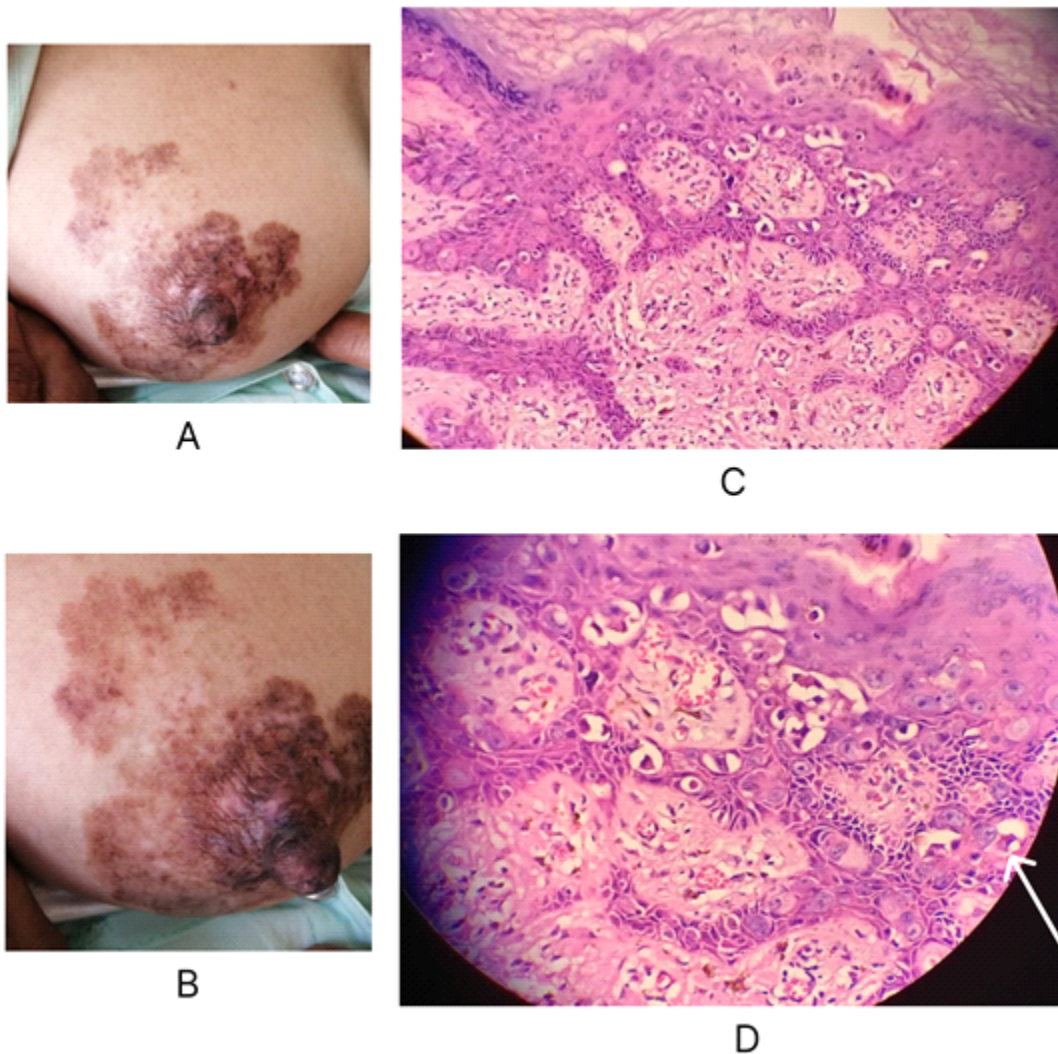


Figure 1.

Discussion

Mammary Paget's disease is reported in 1-3% of all primary breast cancers. Between 93-100% of MPD cases are associated with underlying breast cancer³. It is now recognised as a rare cutaneous intra-epithelial malignancy characterised by large epidermal adenocarcinoma cells, called Paget's cells, within the squamous epithelium of nipple, areola, or adjacent skin³. According to the most accepted epidermotrophic theory, MPD can arise from migrating cells of underlying ductal carcinoma or by malignant transformation of epidermal keratinocytes [in situ transformation theory]¹.

MPD often develops insidiously, evolving over years, typically presenting as a unilateral rash resembling eczema with fine scaling or fissuring. Advanced cases may ulcerate with nipple retraction. Hyper-

pigmentation may occur very rarely, as in our case. Palpable breast mass or bloody nipple discharge may be seen.

Dermatoscopic features of PMPD include an atypical pigment network, structureless blue-grey areas, brown or blue dots and globules, atypical pigment network and sometimes scar-like depigmentation, often mimicking the appearance of melanoma. Histologically, the presence of large epithelial cells [Paget's cells] within the epidermis containing melanin pigment, often with large hyperchromatic nuclei and pale cytoplasm, is seen in both PMPD and melanoma. Therefore, the key diagnostic role is played by immunohistochemistry: PMPD is positive for CK 7, EMA [epithelial membrane antigen], carcinoembryonic antigen, but negative for melanocyte markers like S-100, HMB 45, and Melan A⁴. The hyperpig-

mentation of PMPD is explained by both cytoplasmic deposition of melanin within Paget's cells and by proliferation of melanocytes, even though the exact mechanism of which is to be elucidated⁴.

Workup includes high clinical suspicion and high-quality diagnostic imaging like mammography, ultrasound scan, and MRI scan, which may be needed to rule out underlying malignancy.

Surgical management remains the main intervention for MPD, with more conservative options in cases without underlying cancer. Non-surgical options like radiotherapy alone and photodynamic therapy have also been utilised in selected cases³. Cryoablation of subareolar lesions combined with local excision has also been employed³.

Conclusion

This case emphasises the importance of recognising PMPD as a distinct clinical entity, particularly in the absence of underlying malignancy. Its rarity and

potential for misdiagnosis necessitate heightened clinical vigilance, reliance on histopathology, immunohistochemistry, and individualised management strategies to ensure optimal patient outcomes.

References

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