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(CASE REPORT)

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Pagets disease of the nipple with underlying occult invasive carcinoma discovered through lymph node biopsy: Case report

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Abstract

Paget's disease of the nipple (PDN) is a rare and often misdiagnosed condition characterized by the infiltration of adenocarcinoma cells into the nipple epidermis. It poses substantial diagnostic and therapeutic challenges due to its similarity to benign dermatological conditions and its association with in situ or invasive carcinoma. This case study presents a rare and clinically challenging instance of occult breast cancer associated with Paget's disease of the nipple. Occult breast cancers are often not detectable through standard imaging methods, such as mammograms or ultrasounds, and may only be identified after symptoms appear or through more invasive diagnostic techniques. Paget's disease, a form of breast cancer that affects the skin of the nipple and areola, can be particularly difficult to diagnose early, as its symptoms—such as itching, redness, or discharge—can often mimic benign dermatological conditions. In many cases, Paget's disease is associated with an underlying invasive breast cancer that is not immediately apparent, making early detection and intervention critical. This case highlights the diagnostic journey of a patient presenting with atypical nipple symptoms, ultimately revealing an occult breast cancer linked to Paget's disease. Through this case, we aim to emphasize the importance of a thorough clinical evaluation and multi-disciplinary approach to diagnosis, as well as the significance of early recognition in improving patient outcomes.

Keywords: Paget's disease of the nipple; Symptoms; Therapeutic aspects; Occult breast cancer

1. Introduction

Paget's disease of the nipple (PDN) is a rare and often misdiagnosed condition characterized by the infiltration of adenocarcinoma cells into the nipple epidermis. It poses substantial diagnostic and therapeutic challenges due to its similarity to benign dermatological conditions and its association with in situ or invasive carcinoma. Its association with an occult infiltration carcinoma discovered on lymph node biopsy with no evident breast lesion is even more rare.

PDN is commonly present with symptoms such as pruritus, erythema, scaling, and, occasionally, sanguineous nipple discharge [1] as well as unilateral eczemata or psoriatic lesions with strong margins, that do not respond to topical treatment. Its diagnosis is made by cytological scraping of the nipple or punch biopsies . Because this tumor is associated with an underlying cancer, systemic assessment (e.g., patient history and clinical examination, age-appropriate cancer screening, imaging) is required.

This report details a PDN case in a female patient with concomitant occult invasive breast cancer.

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2. Case presentation

A 50-year-old G5P5 menopausal woman presented at our outpatient department with a 5-year history of persistent pruritus and a rash on her left nipple and areola. She also reported sporadic bloody nipple discharge and mild erythema. The patient had no family history of breast cancer or significant medical history. Upon physical examination, we observed a slightly inverted nipple with erythema, scaling, and minor excoriation localized to the left nipple and areola. No palpable breast masses, or axillary lymphadenopathy, or skin alterations were detected elsewhere on the breast. The contralateral breast appeared unremarkable.

Figure 1 Clinical presentation of patient presenting pagets disease

A **diagnostic mammogram** revealed type 2 tissue breast density with Increased density of the SEQ of the left breast classified ACR 3. Bilateral breast ultrasound revealed Thickening of the skin covering at the left areolar level. Retro-areolar ductal ectasia in the left breast, with finely echogenic content, and a Left axillary lymph node formation with thickened cortex.

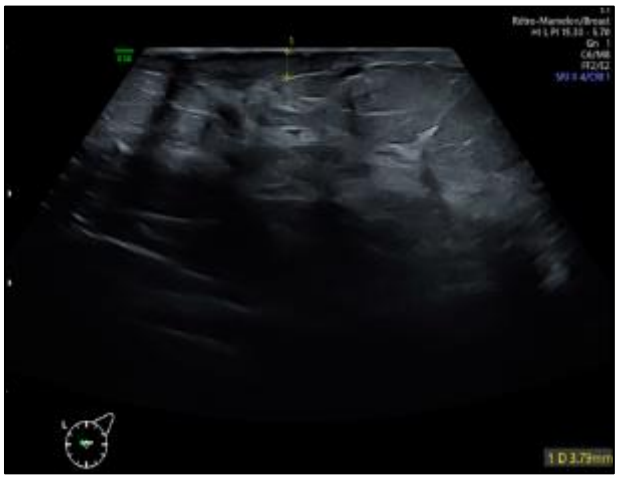

Figure 2 Thickening of a skin covering NAC

Given the persistence of the symptoms and the clinical suspicion of PDN, **punch biopsy** of the left nipple-areola complex and **a fine needle aspiration** of the axillary lymph node was performed. Histopathological examination and Immunohistochemical staining confirmed Paget's disease of the nipple and invasive ductal carcinoma (IDC) on the lymph node. In addition, immunohistochemistry revealed negative for estrogen (ER) and progesterone receptors with HER 2 receptor score at 3.

Breast MRI, which was performed to rule out any underlying breast lesion which could have been missed on mammogram, it revealed Paget disease of the left areolar-nipple plate extended to the retro-areolar glandular tissue over a distance of approximately 8mm. Asymmetric glandular enhancement classified ACR 3 with no pathological translation. Unilateral enlarged axillary lymph nodes with thickened cortex (malignant in appearance). Based on the clinical presentation and pathological findings, a diagnosis of both Pagets disease of the nipple and Infiltrating ductal carcinoma was established.

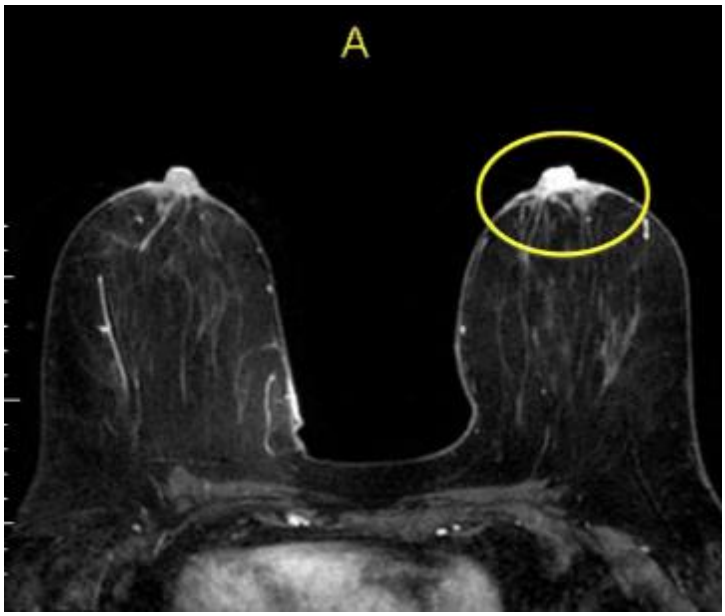

Figure 3 MRI imaging showing thickening of NAC with no underlying lesions

Due to the presence of the axillary lymph node a **TAP CT** was done to search for secondary lesions. It revealed thickening of the areolar-nipple complex of 11mm with an axillary lymph node suspicious of malignancy.

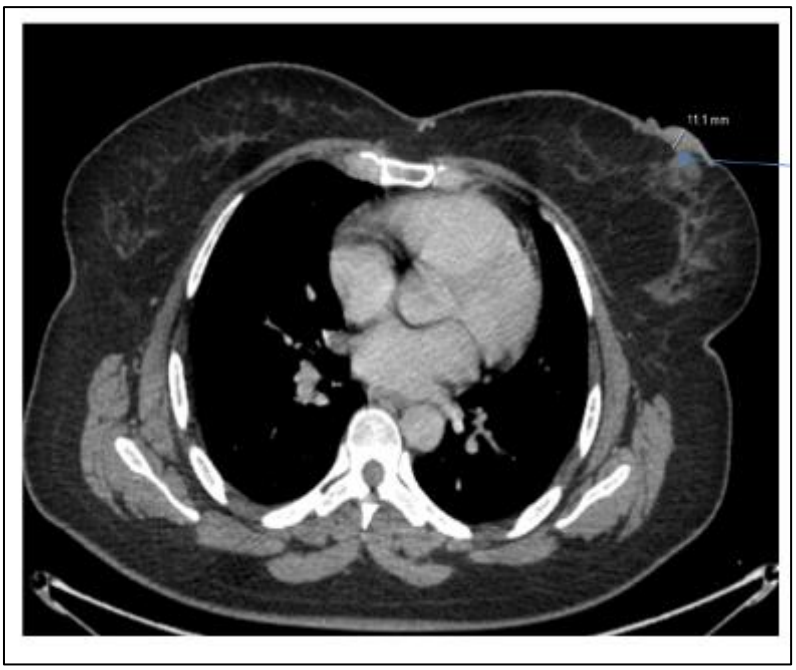

Figure 4 TAP CT revealing thickening of the areolar-nipple complex of 11mm

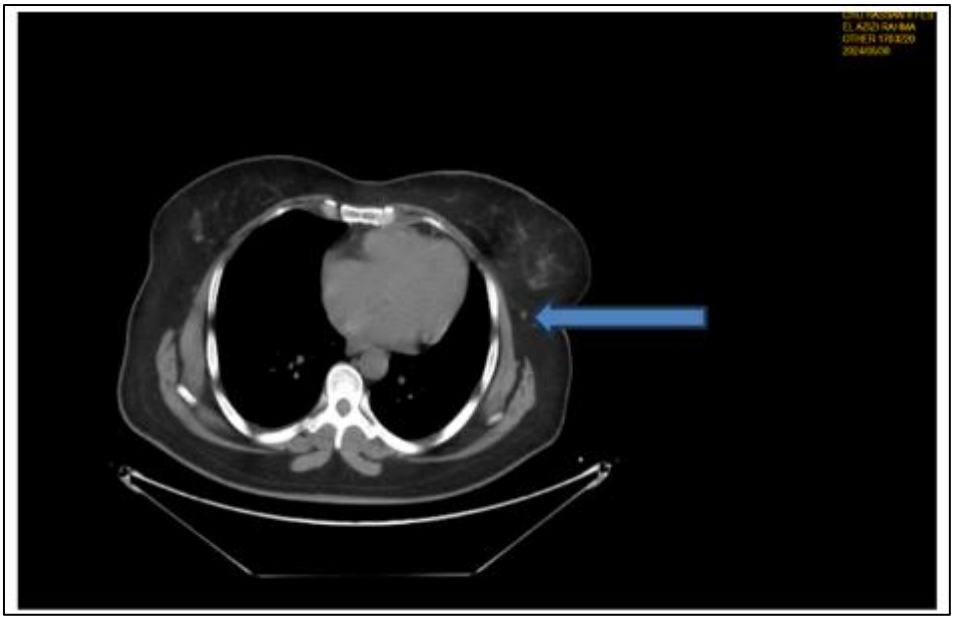

Figure 5 An axillary lymph node suspicious of malignancy (blue arrow)

The patient's case was discussed in a **multi-disciplinary team meeting** with the decision to start her on chemotherapy first with anti HER2, then proceed with a surgical approach. surgical intervention that was recommended was a conservative approach with resection just the NAC but the patient opted for a radical mastectomy which was done with lymphadenectomy fig 6-7 .

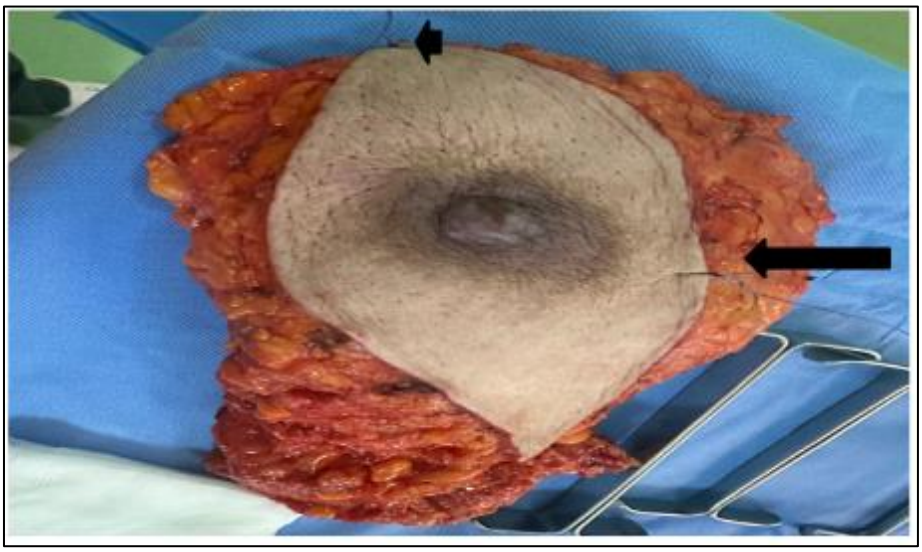

Figure 6 Gross photograph of the specimen post mastectomy : two long threads (arrow) indicates the superior margin, and a short thread (arrowhead) denotes the medial margin

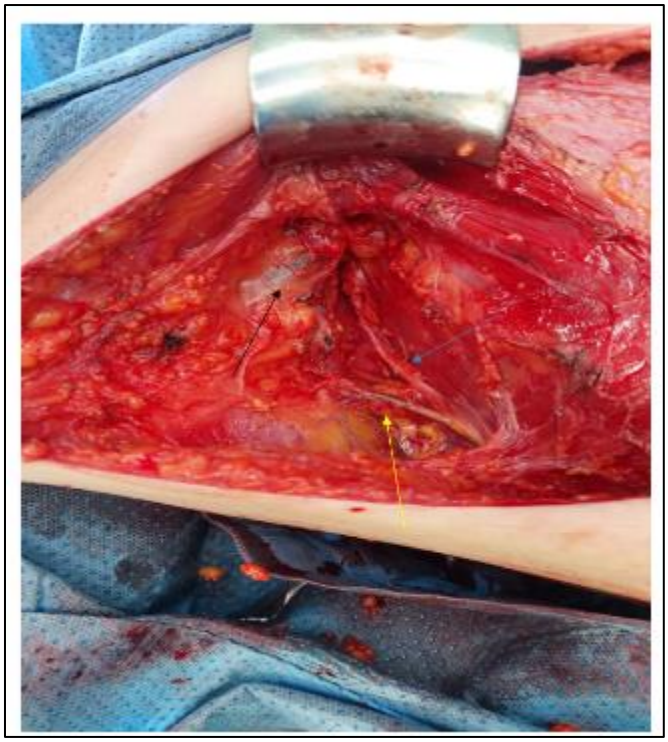

Figure 7 Photograph of lymphadenectomy with anatomical margins

- -Axillary vein at le superior limit (black arrow)
- -Latissimus dorsi flap at the external limit (yellow arrow)
- -And the long thoracic nerve (blue arrow)

The postoperative pathological report indicated:

- Complete therapeutic response, without tumor residue.- Chevallier Grade 1 , TANB grade of Sataloff.- Resection limits are correct. -Presence of Paget disease. - Lymph Node dissection: 23N-/23N.- Stage: ypT0N0

Subsequently, the patient received postoperative radiotherapy and hormonal therapy to minimize the risk of disease recurrence. No adverse events were reported during the follow-up period.

3. Discussion

Paget's disease of the breast, a disorder of the nipple–areola complex, was first described by Sir James Paget in 1874.[1] It is a rare, often insidious form of intraepithelial carcinoma of the breast, typically presenting with eczema-like changes of the nipple and areola. While most frequently associated with underlying invasive breast carcinoma, Paget's disease can occasionally present in isolation or with occult (hidden) breast cancer. The clinical diagnosis is often challenging, as the symptoms such as erythema, scaling, and pruritus of the nipple are non-specific and may mimic benign dermatological conditions.[2]

The presence of Paget's disease without evident masses in breast imaging should raise suspicion for occult malignancy, as it is often associated with ductal carcinoma in situ (DCIS) or invasive carcinoma, though these can be difficult to detect on routine imaging.[3] Paget's disease of the nipple is isolated in 1.4 to 13.3% of cases and associated with ipsilateral breast cancer in 82 to 100% of cases. Of these, 13.3 to 52% are carcinomas in situ and 30 to 60% are invasive carcinomas.[4]

Imaging is critical in diagnosing underlying breast cancer. Mammography has a sensitivity of 97% when a palpable mass is present but drops to around 50% when no mass is palpable.[5] MRI has demonstrated greater sensitivity in detecting associated malignancies, especially when mammograms and ultrasounds are inconclusive, and is recommended for surgical planning in Paget's disease.[6]

The pathophysiology of PDN suggests that tumour cells originating in the breast ducts invade the nipple's epithelium and epidermis, leading to the skin changes observed clinically.[7] In cases of occult PDN, the underlying breast tumour may remain undetectable, either because it is too small, deeply located, or obscured by dense breast tissue. Occult breast cancer poses significant diagnostic challenges. It is often only when cancer cells are detected in regional lymph nodes, through biopsy, that the underlying malignancy is uncovered.[8]

Several case reports have illustrated instances of occult breast cancer discovered only after lymph node metastasis, with Paget's disease being the only outward sign. A retrospective study reviewed by Jones et al. showed that biopsy of suspicious axillary lymph nodes in patients with nipple changes suggestive of Paget's disease, even in the absence of a detectable primary tumour, can reveal malignancy.[9]

3.1. Diagnostic Challenges

The diagnosis of PDN associated with occult breast cancer is complicated by the lack of definitive imaging findings. Mammography and ultrasound may fail to identify small or deeply located tumours, particularly in patients with dense breasts. Moreover, the dermatological presentation often mimics benign conditions such as eczema or dermatitis, making clinical differentiation difficult. Therefore, biopsy of nipple lesions and suspicious lymph nodes remains essential in establishing the diagnosis.[10]

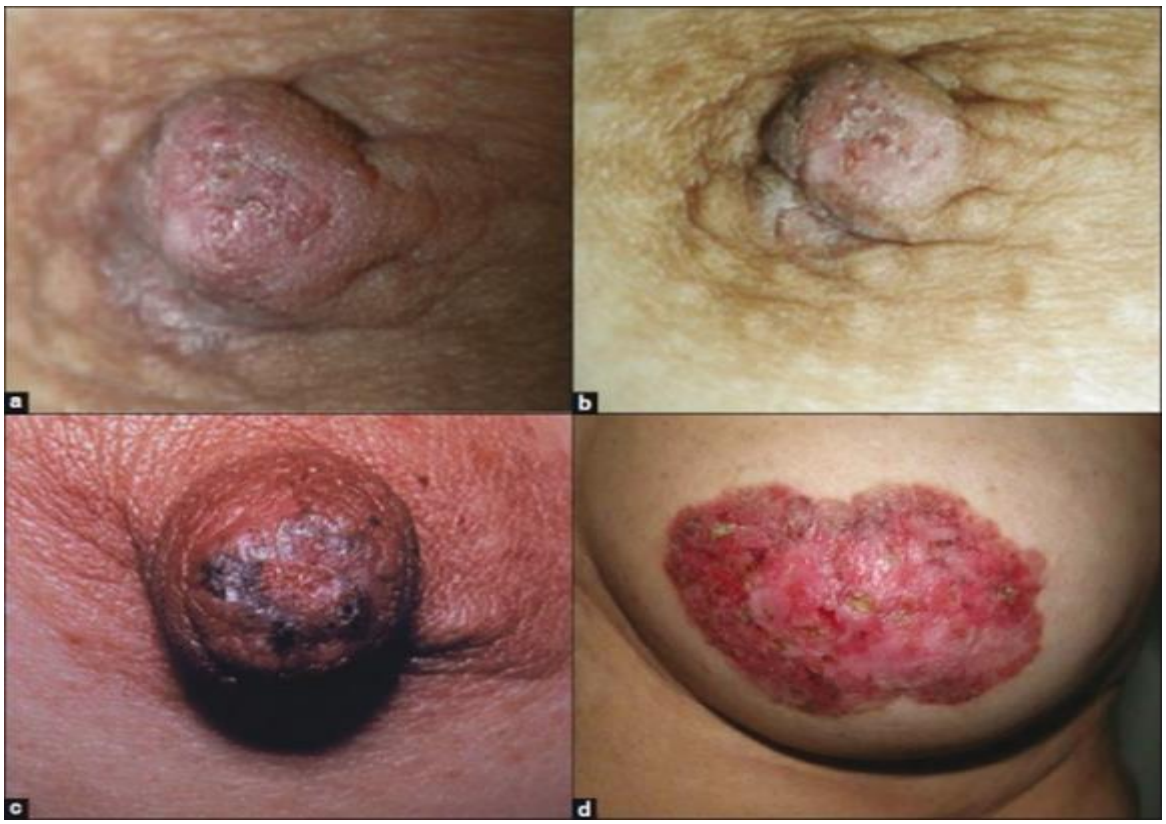

Figure 8 ***Google images***: (a and b) Paget's disease of the nipple. The clinical appearance is usually a thickened, eczematoid crusted lesion with irregular borders. (c) Scaly, erythematous, crusty pigmentation and thickened plaques on the nipple, spreading to the surrounding areolar areas. (d) Advanced lesions show skin thickening, redness, erythema, erosion of the nipple and scaling around the nipple–areola

Lymph node biopsy is increasingly recognized as a valuable diagnostic tool in identifying occult breast cancer. Studies have shown that sentinel lymph node biopsy can reveal metastatic disease in patients with no detectable primary tumour on imaging.[11] This procedure aids in staging and therapeutic planning and is particularly useful when PDN is the only visible sign.

3.2. Management and Treatment

Treatment typically involves surgical excision of the nipple-areola complex and any underlying tumour. In the presence of occult malignancy, mastectomy may be performed depending on tumour extent and lymph node involvement. MRI findings can guide the surgical approach and extent of resection.[6]

For hormone receptor-positive tumours, adjuvant hormonal therapy is indicated, while HER2-positive tumours may benefit from trastuzumab-based regimens. Invasive cases may require chemotherapy and/or radiotherapy, particularly when nodal involvement is confirmed.[12]

Recent research supports earlier and aggressive treatment strategies. Houssami et al. found that timely intervention improves survival, particularly when occult disease is detected and managed promptly.[13]

3.3. Prognosis

The prognosis of PDN with occult breast cancer varies depending on tumour stage and nodal involvement. Early-stage, localized disease generally has a favourable outcome with appropriate treatment. However, advanced disease involving lymph nodes or distant metastasis is associated with poorer prognosis. Nonetheless, recent findings suggest that with adequate surgical and adjuvant therapy, patients with imaging-negative tumours can achieve outcomes comparable to those with more overt disease presentations.[14]

4. Conclusion

This case highlights the diagnostic and therapeutic challenges in managing PDN associated with occult breast cancer. The condition often presents subtly, mimicking benign dermatologic disorders, and may evade detection on routine imaging. Lymph node biopsy serves as a crucial diagnostic adjunct in such scenarios. Early biopsy, MRI evaluation, and multidisciplinary treatment are essential to improve outcomes. Ongoing research is necessary to refine diagnostic algorithms and enhance early detection strategies for occult breast malignancies in patients with PDN.

Compliance with ethical standards

Disclosure of Conflict of Interest

The authors declare that they have no conflict of interest.

Statement of Ethical Approval

The present research work does not contain any studies performed on human or animal subjects by any of the authors.

Statement of Informed Consent

Informed consent was obtained from the patient for publication of this case report and any accompanying images.

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