Unusual association of diseases/symptoms

Isolated adenocarcinoma of the nipple

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Correspondence to Muneer Ahmed, muneer_ahmed@hotmail.co.uk**Summary**

A 47-year-old female presented with a 1-year history of 'eczematous change' to the right nipple. Bilateral mammography and ultrasound were entirely normal. Free hand biopsy demonstrated invasive adenocarcinoma of the nipple. The patient underwent a right-sided central segmentectomy and sentinel node biopsy. Histology demonstrated that the nipple was almost completely replaced by an invasive ductal carcinoma with a maximum diameter of 13 mm. Invasion of the underlying breast tissue was to a depth of 3 mm. A single sentinel lymph node demonstrated metastatic carcinoma. Her oestrogen receptor status was positive while HER-2 status was negative. The patient subsequently underwent right-sided axillary node clearance to level three nodes. All 17 nodes in the specimen were found to be within normal limits. She is scheduled to undergo radiotherapy, chemotherapy and hormonal treatment.

BACKGROUND

This case is important because it demonstrates that patients can present with malignancy in the absence of radiological findings. It highlights that any persistent skin changes require histological diagnosis and this should not be delayed.

To find an adenocarcinoma of the nipple without any underlying breast mass is rare. The fact that this is such a rare case with the reinforcement of the need for triple assessment of breast disease is the reason we decided to write this up.

CASE PRESENTATION

A 47-year-old female presented with a 1-year history of 'eczematous change' to the right nipple. This had been unresponsive to topical steroids prescribed by her general practitioner in the community. She denied any nipple inversion, discharge or self-palpating a breast lump. She had no family history of breast cancer, was premenopausal with two children. She had no other medical issues and took no other medications. She was an active smoker.

On examination there were confluent crusted papules and plaques consistent with eczematous change confined

Figure 1 Histopathological specimen demonstrating invasive adenocarcinoma replacing the connective tissue of the nipple.

to the right nipple-areolar complex. There were no palpable masses or axillary lymphadenopathy present.

INVESTIGATIONS

Bilateral mammography and ultrasound scanning were performed with no abnormality identified. Due to the high suspicion of Paget's disease an open free-hand biopsy (FHB) of the right nipple was performed.

DIFFERENTIAL DIAGNOSIS

- Paget's disease of the nipple
- Adenocarcinoma of the nipple
- Breast adenocarcinoma with nipple invasion.

TREATMENT

The FHB demonstrated that the connective tissue of the nipple was infiltrated by invasive adenocarcinoma.

The patient was discussed at the breast multidisciplinary team meeting with the conclusion that the patient should undergo a right-sided central segmentectomy and sentinel node biopsy. The above procedure was performed uneventfully with the histology acquired (figure 1) demonstrating that the nipple was almost completely replaced by an invasive ductal carcinoma of no special type, grade 2, with a maximum diameter of 13 mm. It extended up to the epidermis without invasion. Invasion of the underlying breast tissue was to a depth of 3 mm. Adequate clearance margins were attained with the closest margin being the inferior margin at 15 mm. High grade ductal carcinoma in-situ was present within the lactiferous ducts of the nipple with the nearest clearance margin of 4 mm laterally. A single hot and blue sentinel lymph node demonstrated metastatic carcinoma. Her oestrogen receptor status was positive while HER-2 status was negative.

The patient subsequently underwent right-sided axillary node clearance (ANC) to level 3 nodes. All 17 nodes in the specimen were found to be within normal limits.

OUTCOME AND FOLLOW-UP

She is scheduled to undergo chest wall and axillary radiotherapy, chemotherapy and commence hormonal treatment.

DISCUSSION

Malignant changes of the nipple commonly occur when there is an underlying breast malignancy, as in the case of 95% of cases of Paget's disease.¹ It is also commonly the case when an underlying breast adenocarcinoma involves the nipple through invasion. In their pathological study, Lagios *et al* demonstrated carcinomatous involvement of the nipple-areola complex in 30% (45/149) of patients. A large proportion of patients with nipple involvement by invasive lesions were those whose tumours were located within 2.5 cm of the nipple border.² Morimoto *et al*³ in their pathological specimen study concluded that non-infiltrating carcinomatous tumours within 5 cm of the nipple border and, possibly, multiple tumours, must be considered to have a high risk of nipple-areola involvement. However, interestingly in our case there were no changes in the underlying breast identified on clinical or

radiological investigation. On histological examination of the central segmentectomy specimen there was only 3 mm of inferior margin invasion of underlying breast tissue. Such a case is very rare, with only one other case found in the literature. Ohsumi *et al* demonstrated a case of a invasive adenocarcinoma originating from the duct of the nipple. This patient underwent mastectomy as opposed to breast conserving surgery and axillary clearance. Metastatic foci were found in one of nine axillary nodes.⁴ Interestingly the nipple lesion was visualised on mammographic and ultrasound imaging as opposed to our case which was not.

The sensitivity of mammography in detecting breast cancers in women over 60 years of age approaches 95%,⁵ while ultrasound has a high sensitivity for breast pathology and also a high negative predictive value particularly in younger cohorts.⁶ These two modalities used in conjunction with highly suspicious clinical presentations are the standard techniques in breast departments.

Learning points

- Skin changes involving the nipple in the absence of an underlying breast lump can be a sign of malignancy.
- Our case highlights the issue that malignant presentations of the nipple do occur in the absence of any clinical or radiological findings of underlying malignancy.
- Our patient was treated with topical steroids for 1 year by her general practitioner before being referred to our breast unit for triple assessment. This delay in her referral prevented her prompt diagnosis and treatment.
- Our case, therefore, highlights to the general medical community that any patient presenting with skin changes to the breast should be urgently referred for triple assessment under the 2-week wait cancer pathway to a specialist breast unit. This management plan would ensure that there is no delay in diagnosing such malignant lesions.

Acknowledgements The authors would like to thank Dr Mark Rogerson for kindly producing the histopathological slides used in this article.

Competing interests None.

Patient consent Obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Ahmed M, Basit A. Isolated adenocarcinoma of the nipple. *BMJ Case Reports* 2011;10.1136/bcr.02.2011.3825, date of publication

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