

Case Series

Paget's disease of the breast: a case series

Solomon N. Elenwo*, Rollings S. Jamabo

Department of Surgery, University of Port Harcourt Teaching Hospital, Port Harcourt, Nigeria

Received: 29 April 2020

Revised: 05 July 2020

Accepted: 06 July 2020

***Correspondence:**

Dr. Solomon N. Elenwo,

E-mail: snelenwo@yahoo.co.uk

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ABSTRACT

Paget's disease of the nipple is a rare disease with a rather bizarre presentation which makes the diagnosis easily missed. Three cases are presented here with different modes of presentation and histological findings. A biopsy provided the definitive diagnosis in the three patients and the treatments offered was based on the histological findings - modified radical mastectomy and simple mastectomy for invasive carcinoma and carcinoma in-situ respectively.

Keywords: Paget's disease, Breast, Nipple-areola complex, Case series

INTRODUCTION

Paget's disease of the nipple was first described by Sir Paget in 1874.¹

It is a very common disease and many of the patients present with only eczematous lesions involving the nipple.^{2,3}

Some patients may present with lump while others do not. Diagnosis of this condition, therefore, is often missed.

This study presents three patients with different manifestations of the disease with a view to highlighting a few of the different manifestation.

CASE SERIES

Case 1

A 30-year old female trader was referred to the breast clinic of our hospital in April 2017 with an 8-month history of an eczematous lesion on her left nipple.

After self-medication for a while, she was seen by a general practitioner who prescribed anti-fungal creams for her.

Initial response was followed by recurrence and lack of any further response.



Figure 1: Paget's disease of the nipple with excoriation.

Examination of the breast revealed excoriation of the nipple of her left breast. No palpable breast or axillary mass (Figure 1).

Ultrasonography and mammography were unrewarding. Biopsy of an ellipse of skin of the involved area of the nipple showed the typical pagetoid cells and a ductal carcinoma in-situ.

The patient underwent a simple mastectomy. She is still without recurrence as she is still being followed up.

Case 2

A 46-year-old mother of four who was referred to the breast clinic by a dermatologist in August 2018 with complaints of a “bleeding excoriation” on the left nipple of 10 months duration.

A painless eczematous lesion was first noticed and was followed 8 months later by recurrent bleeding which made her seek medical attention.



Figure 2: Paget's disease of the nipple with excoriation and bleeding.

On breast examination, the left nipple was almost completely eroded with profound contact bleeding (Figure 2).

There was no palpable mass on the breast and the ipsilateral axilla. Both breast ultrasonography and mammography revealed no abnormalities.

A wedge biopsy of the involved nipple showed invasive ductal carcinoma. A modified radical mastectomy (MRM) was done and she is without recurrence till date.

Case 3

A 47-year-old teacher who was referred to the breast clinic in March 2019 on account of a mass on the right breast of 2 months duration.

She had for over 4 months, an excoriation on the nipple of that breast which she had ignored.

On examination, she was found to have an eczematous lesion on the right breast, a palpable 3 cm wide, hard and non-tender sub-areolar lump with no palpable ipsilateral axillary nodes.

A mammogram revealed the lump with a few microcalcifications. A biopsy of the lump showed invasive ductal carcinoma. She underwent an MRM and is still being followed up for any recurrence.

DISCUSSION

Paget's disease of the nipple is a relatively uncommon disease with a reported prevalence of 1-4.3%.⁴ Like the regular breast cancer, it is a disease of women.²

The mean age of presentation is reported to be around the age of 55 years but is now being seen among younger women.^{5,6} Our case 1 is an example, being only 30 years of age.

The hallmark of presentation is an eczematous lesion on the nipple which was seen in the three patients presented. This may partially respond to anti-fungal therapy and this contributes to late presentation and misdiagnosis.³

Our 3 patients had unilateral disease as this is said to be much more common, but bilateral disease has also been reported.⁷

Bleeding from the excoriated nipple is often seen particularly in long neglected cases as in our case 2.

Only our third case presented with a palpable mass. While this is not an uncommon finding, it is less likely to be seen in patients who present early.¹⁻³

All the three patients had mammography done but only case 3, with a palpable mass had a significant mammographic finding.

This is because mammography is said to be most rewarding in the presence of a palpable mass.^{8,9}

A definitive diagnosis of Paget's disease of the nipple is only possible via a biopsy, either of the involved nipple skin and/or a palpable lump.

In over 90% of cases of Paget's disease of the nipple, an underlying cancer is identified by a biopsy.¹⁰ All our patients had a biopsy done for them and they all showed a malignancy.

The possible histologies range from a carcinoma in-situ to an invasive carcinoma.

Our 3 cases showed this range. Ductal carcinoma in-situ is said to be more likely in patients without a palpable mass as seen in our case 1.¹⁰

Our second and third patients had MRM. This is in line with the thinking that cases of Paget's disease of the nipple with invasive ductal carcinoma have a high risk of axillary node involvement.¹¹

We considered a simple mastectomy as the minimum surgery for our first case with ductal carcinoma in-situ because studies have shown a high incidence of recurrence following treatment with a local excision and radiotherapy.^{12,13}

CONCLUSION

Paget's disease of the nipple is an uncommon disease which requires a high index of suspicion and a good knowledge of the symptomatology for prompt diagnosis. The cases presented clearly demonstrate this and highlight the need for adequate enlightenment of both patients and medical practitioners.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Elenwo SN, Jamabo RS. Paget's disease of the breast: a case series. Int Surg J 2020;7:2706-8.