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Paget's Disease of the Breast: A Tertiary Hospital Experience

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Abstract

Background: Paget's disease of the breast is rare. It is one of the foremost subtypes of carcinoma of the breast, accounting for only 1-3% of all breast tumors. It is often misdiagnosed because of its unusual presentation and absence of a breast lump in nearly half of the patients. This study aims at evaluating the epidemiology and outcome of surgical treatment of Paget's disease of the breast in Port Harcourt, Nigeria.

Methods: This is a retrospective study of cases of Paget's disease of the breast seen and managed in the University of Port Harcourt Teaching Hospital, Port Harcourt, Nigeria over a 20 year period. The patients were analyzed for age, sex, clinical presentation and treatment given.

Results: A total of 24 patients out of 2,105 cases of carcinoma of the breast were seen to have Paget's disease. All were females aged 30 to 55 years and all had unilateral disease. Only 10% of the patients had a palpable mass on the breast. All the patients had biopsies either of the skin or both skin and breast lump.

Conclusion: Paget's disease of the breast is uncommon and easily misdiagnosed. This calls for wedge resection and/or histology of any mass seen for any patient presenting with eczematous lesions on the nipples.

Because of this, many patients in our part of the world present very late. This study, therefore, aims at evaluating and highlighting the epidemiology of this condition and treatment outcomes in our environment in view of the late presentations.

Research Methodology

This is a retrospective study carried out at the University of Port Harcourt Teaching Hospital. All patients with suspected excoriations/ bleeding from the nipples and/or areola, with or without a mass seen between June 1999 and May 2019 were included in the study. Those who did not undergo confirmatory investigations were excluded from the study. Ethical approval was obtained from the hospital. The 24 cases confirmed out of a total of 2,105 patients with cancer of the breast were analyzed for age, sex, and clinical characteristics. All the patients were further investigated with ultrasonography and mammography. They also had a biopsy of an ellipse of skin of the nipple-areola complex and all palpable breast masses. Magnetic Resonance Imaging was not functional in our center during the study period and was therefore not done for any of them. Those found to have invasive carcinoma underwent Modified Radical Mastectomy, while those with ductal carcinoma *in-situ* had simple mastectomy.

Results

The prevalence of Paget's disease of the breast was found to be 1.14% of all cancers of the breast in our center. All the patients were women, with ages ranging between 30 and 55 years (mean age 41.9 years). They presented with clinical findings suggestive of Paget's disease of the breast. These included eczematous lesions or excoriations at the nipple, bleeding from the nipple-areola complex, and a palpable mass. All the patients presented with eczematous lesions or excoriations at the nipple-areola complex in addition to other symptoms (**Figure 1**). Seven (29.2%) of these patients had excoriation of the nipple-areola complex with bleeding from the area (**Figures 2 and 3**). Nine (37.5%) patients had eczematous lesions only and there was no associated bleeding. Pictures of two of our patients with eczematous lesions and excoriation of the nipple with associated bleeding are shown below.

Keywords

Breast; Nipple-areola complex; Paget's disease; Management

Introduction

Sir James Paget described the eczematous lesions of the nipple associated with an underlying breast carcinoma in his original report of 1874 [1]. It is an uncommon disease of the nipple-areola complex with a prevalence of 1-4.3% of all breast cancers [2]. The diagnosis is often missed because only about 50% of patients present with a palpable lump [3,4]. Also, the eczematous manifestation of this condition may respond to topical agents, making the diagnosis even more confusing [3].

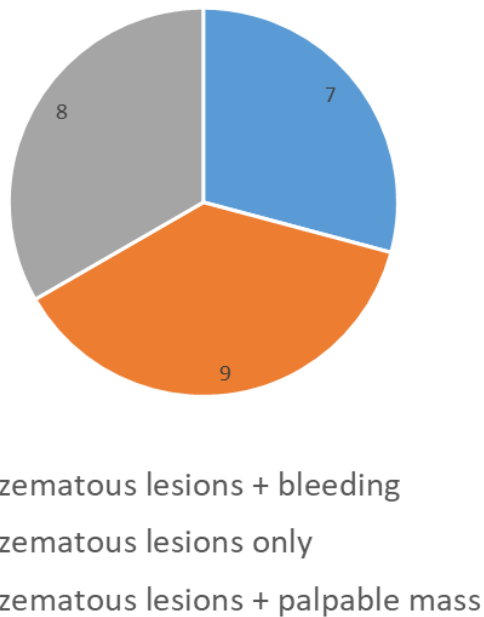


Figure 1: Pathological findings/treatment given.

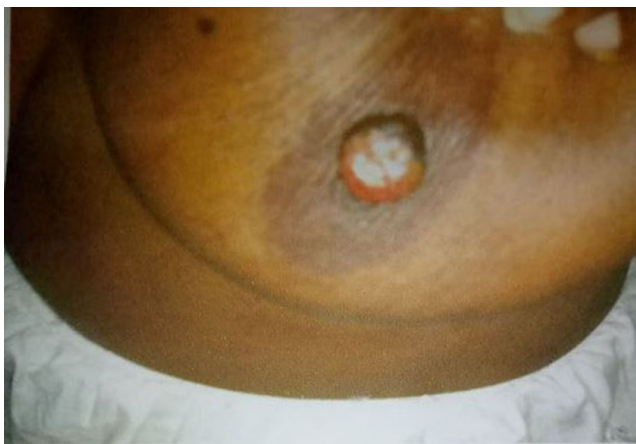


Figure 2: Paget's disease of the nipple with excoriation.

Palpable breast masses were noticed in 8 patients (33.3%). All the 24 patients were further investigated with mammography and Ultrasound Scan (USS). The USS was able to detect multifocality and/or multicentricity in seven patients. The mammography picked micro-calcifications in all those with palpable masses and in half of those without palpable masses. All the patients were subjected to a biopsy of an ellipse of skin of the nipple-areola complex and any palpable masses. Microscopically, the typical pagetoid cells which are characterized by the presence of large, ovoid cells with abundant, clear, pale-staining cytoplasm were found in the skin biopsies and the excised masses were positive for malignancy. Pathological findings were ductal carcinoma *in situ* (DCIS) (n=3), invasive ductal carcinoma (IDC) (n=19), and

invasive lobular carcinoma (ILC) (n=2). Modified radical mastectomy was performed for 21 patients and simple mastectomy for the remaining three.



Figure 3: Paget's disease of the nipple with excoriation and bleeding.

Discussion

Paget's disease of the breast is not a very common disease. The prevalence of 1.4% of all breast cancers seen in our center is in line with the 1– 4.3% reported by Tavassoli et al. in 1999 [2].

All our patients are women obviously because of the predominance of breast cancer in women [4]. However, men can also be affected [5,6].

The mean age of 41.9 years in our center is significantly younger than the 55 years earlier reported [7]. This is not surprising because Paget's disease has now been reported in younger women and even adolescents [8].

The clinical presentation patterns in our study comprising mainly eczematous nipple lesions, nipple excoriations/ulceration/bleeding and a palpable mass are in line with studies in the literature (**Table 1**) [3,4,9].

Table 1: Demographic and clinical characteristics of the patients.

Age (Years)	Sex	Unilateral or Bilateral	Presentation
30	F	Unilateral	Eczematous lesions/excoriations
42	F	Unilateral	Excoriation + mass
38	F	Unilateral	Excoriation + bleeding
46	F	Unilateral	Excoriation + bleeding
42	F	Unilateral	Excoriation + mass
41	F	Unilateral	Excoriation + mass
47	F	Unilateral	Eczematous lesions/excoriations
39	F	Unilateral	Excoriation + bleeding

40	F	Unilateral	Ecematous excoriations	lesions/
41	F	Unilateral	Excoriation + mass	
55	F	Unilateral	Excoriation + mass	
44	F	Unilateral	Excoriation + bleeding	
47	F	Unilateral	Excoriation + mass	
34	F	Unilateral	Ecematous excoriations	lesions/
41	F	Unilateral	Ecematous excoriations	lesions/
48	F	Unilateral	Excoriation + mass	
37	F	Unilateral	Ecematous excoriations	lesions/
39	F	Unilateral	Excoriation + mass	
43	F	Unilateral	Excoriation + bleeding	
44	F	Unilateral	Ecematous excoriations	lesions/
41	F	Unilateral	Excoriation + bleeding	
42	F	Unilateral	Ecematous excoriations	lesions/
38	F	Unilateral	Ecematous excoriations	lesions/
46	F	Unilateral	Excoriation + bleeding	
n=24				

All our patients had unilateral disease but bilateral cases have been reported [10]. It may even be seen in ectopic breasts or accessory nipples [8,11]. A palpable mass as reported in our study is frequently found [3,4].

Table 2: Pathological findings/treatments given.

Pathological Findings	Number (n=24)	Treatment Given	Number With 5-Years Survival
Ductal Carcinoma <i>in-situ</i>	3	SM	2
Invasive Ductal Carcinoma	19	MRM	4
Invasive Lobular Carcinoma	2	MRM	1

MRM= Modified Radical Mastectomy; SM= Simple Mastectomy

In our study, all those with invasive cancer underwent Modified Radical Mastectomy (MRM), while the two with DCIS had simple mastectomy. There does not appear to be any unanimity as to the appropriate treatment of this disease. Most of those with invasive cancer have a high risk of axillary node involvement, accounting for why many surgeons consider MRM for them [22]. Even those with *in-situ* carcinoma, even when limited in extent, have a high incidence of recurrence when treated with local excision and radiotherapy [23,24]. However, some workers have reported good outcomes following breast conservation surgeries with or without radiotherapy for Paget's disease of any histology confined to the central portions of the breast only [22,25,26]. They suggest that mastectomy should be reserved for cases

Mammography, breast ultrasonography and a biopsy were the major investigative tools used in our study. Multifocality is not uncommon [12,13]. The sensitivity of mammography is, however, not very encouraging as up to 65% of these patients will have a negative mammography in the presence of an underlying cancer [14]. Mammography seems to be more reliable in the presence of a palpable mass [15,16]. Microcalcifications found in our study are among the findings on mammography suggesting malignancy [15,17].

Breast ultrasonography is also useful; especially if mammography is not helpful [18]. Magnetic Resonance Imaging (MRI) was not functional in our center during the study period. It is said to be more sensitive than both mammography and ultrasonography [17,19] and may pick up unsuspected tumours [20,21].

A biopsy is always advised because no other investigative tool can give conclusive evidence of the absence of an underlying carcinoma. It reveals the presence of a cancer in over 90% of patients [13]. An underlying malignancy is very common in Paget's disease, with a prevalence of 92-100% being reported in the literature [12,22]. A wedge biopsy, instead of a superficial shave biopsy or punch biopsy, is said to be the best method for histological diagnosis [23]. If this is not useful or is inconclusive, a second biopsy or even an excision of the nipple is recommended for a full thickness biopsy [10]. An invasive ductal carcinoma is the most frequent finding in those with a palpable mass but a Ductal carcinoma *in situ* (DCIS) is the most likely finding in cases without a palpable mass [13]. Immunohistochemistry was not done for any of our patients but Paget's disease mostly shows negativity to both estrogen and progesterone receptors because of poor differentiation of the underlying cancer (**Table 2**) [9].

that relapse [25]. This will, however, require adequate follow-up with mammography [27].

Conclusion

Paget's disease is an uncommon condition found predominantly in women. The prevalence in our center follows the pattern in the literature. Most of our patients were seen in late stages due to poor awareness even amongst medical personnel. Diagnosis is by clinical suspicion and biopsy, although imaging plays a significant role. The high incidence of late presentation and the difficulty of adequate follow-up

make it difficult for conservative breast surgeries to be advised in our centre.

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