SUNTEXT REVIEWS

SunText Review of Case Reports & Images
ISSN: 2766-4589

Open Access
Research Article
Volume 4:7

Insights into Mammary Paget's disease: A Dermoscopic Description of 8 Cases

Joutei Hassani KT*, Soughi M, Bennouna Z, Baybay H, Elloudi S, Douhi Z and Mernissi FZ

Department of Dermatology, University Hospital Hassan II, Morocco

***Corresponding author:** Joutei Hassani KT, Department of Dermatology, University Hospital Hassan II, Fes, Morocco; Tel: +212611533565; E-mail: kenzatahri10@gmail.com

Received date: 31 August 2023; **Accepted date:** 04 September 2023; **Published date:** 10 September 2023

Citation: Joutei Hassani KT, Soughi M, Bennouna Z, Baybay H, Elloudi S, Douhi Z, et al. (2023) Insights into Mammary Paget's disease: A Dermoscopic Description of 8 Cases. SunText Rev Case Rep Image 4(7): 203.

DOI: https://doi.org/10.51737/2766-4589.2023.103

Abstract

Background: Mammary Paget's disease (MPD) is an infrequent condition affecting the nipple-areola region. It represents an intraepithelial adenocarcinoma and often mimics the clinical appearance of inflammatory or infectious skin disorders. This entity is typically associated with an underlying ductal carcinoma. Histological analysis reveals Paget cells infiltrating the epidermis without evidence of invasion. Objectives: Our aim is to describe the clinical and dermoscopic features of MPD.

Materials and Methods: This study presents a prospective and descriptive analysis of 8 patients, accumulated within our training program over a 4-year period (from March 2020 to June 2023). Each case included in the study was histologically verified as MPD.

Results: A total of 8 female patients, with an average age of 57,1 years, were included in our study. The prevailing clinical presentation of MPD was in the form of a plaque involving both the nipple and areola. Dermoscopically, white scales were the most commonly observed pattern, appearing in 6 out of 8 cases (75%). Shiny white lines and linear vessels were identified in 5 out of 8 cases(62,5%), followed by structureless pink areas, dotted vessels, and erosion/ulceration, each noted in 50% of the cases.

Conclusion: The prevailing characteristics of an individual with MPD typically involves an older woman who presents with a unilateral, asymptomatic, erythematous plaque on the nipple and areola. Dermoscopically, MPD commonly features white scales, shiny white lines, linear vessels, dotted vessels and pink structurless areas. In scenarios where pigmentation is evident, the presence pigmented dots/granules could also be noted.

Keywords: Mammary paget disease; Nipple-areola complex; Intraepithelial adenocarcinoma; Dermoscopy

Introduction

Mammary Paget's disease (MPD), constituting approximately 1% of all breast cancers, is an uncommon intraepidermal adenocarcinoma localized within the nipple-areola complex. In around 90% of MPD cases, an underlying breast adenocarcinoma can be identified [1,2]. Clinically, MPD often presents with a non-specific morphological appearance, typically appearing as an ulcerated, crusted, or scaly patch or plaque on the nipple, sometimes extending to the areolar region [1]. Distinguishing MPD based on clinical observations can be intricate due to its potential resemblance to various benign and malignant skin conditions [3]. Early detection is essential to facilitate comprehensive evaluation for potential underlying breast malignancy. Although mammography and B-mode ultrasonography are commonly favored diagnostic techniques for various types of breast cancer, their effectiveness in diagnosing MPD is constrained by the absence of distinct features [4]. Consequently, there arises a necessity for alternative diagnostic approaches to enhance the prompt identification of MPD and alleviate diagnostic delays. Dermoscopy, widely utilized for the early detection of both malignant and benign skin ailments, offers promising potential in assisting the identification of MPD [5]. Given the rarity of MPD, its dermoscopic characteristics have not been well-defined in existing literature.

Objectives

Our aim is to describe the clinical and dermoscopic features of MPD.

Materials and Methods

This is a prospective and descriptive study that involves the analysis of 8 cases of MPD. These cases were collected from the dermatology department of Hassan II University Hospital over a period of 4 years, from March 2020 to June 2023. Each case underwent a thorough clinical and dermoscopic examination during routine dermatological consultations, conducted by the same examiner and subsequently evaluated by two examiners. Dermoscopic images were captured using a dermlite 4 dermoscope, employing both immersion and non-immersion techniques. The diagnosis of MPD was confirmed through histological examination for all cases.

Results

Table 1 resumes the epidemiological and lesional characteristics of the patients. A total of 8 female patients, with an average age of 57,1 years (range: 42–69), were included in our study (Table 1).

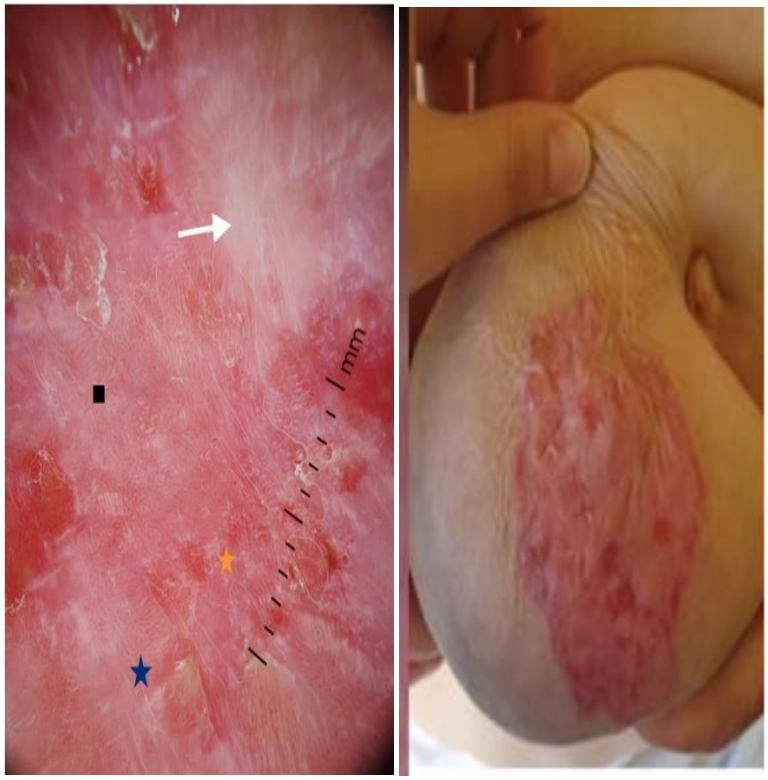

Figure 1: *Dermoscopy of mammary paget's disease: Black Square: dotted vessels, blue star: linear vessels, orange star: shiny white lines, white arrow: white structurless area.*

Figure 2: *Dermoscopy of MPD: Blue star: Dotted vessels, Black star: Erosion.*

Figure 3: *Dermoscopy of pigmented MPD: Blue star: Brown dots/Globules, Black star: White scales.*

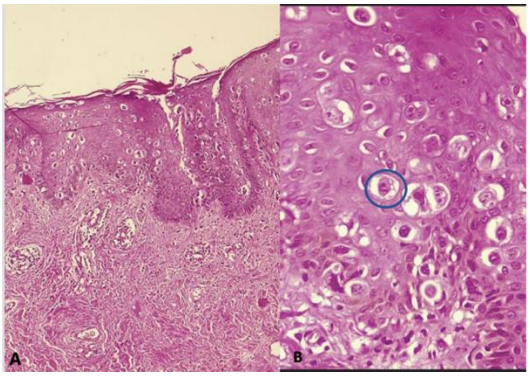

Figure 4: *A - intradermal tumor proliferation composed of small clusters or isolated large cells with abundant clear cytoplasm and vesicular nuclei featuring prominent nucleoli(x40) B- Blue cercle: Paget's cell (x100).*

Table 1: *Epidemiological and lesional characteristics of patients.*

Sex	
Males	0
Females	8(100%)
Mean age (years)	57,1
Mean evolution time (months)	22
Anatomic distribution	
Nipple	3 (37,5%)
Areola	0
Nipple and areola	5 (62,5%)
Clinically pigmented	
Yes	2 (25%)
No	6 (75%)
Clinical aspect	
Patch	1 (12,5%)
Plaque	4 (50%)
Papule/nodule	0
Ulceration	3 (37,5%)

Table 2: *Frequency of dermoscopic features in mammary Paget's disease.*

Dermoscopic Features	
Structureless pink areas Structureless white areas	4 (50%) 1 (12,5%)
Granules/dots/globules brown Granules/dots/globules blue/grey	1 (12,5%) 1 (12,5%)
Dotted vessels Linear vessels Erosion/ulceration	4 (50%) 5 (62,5%) 4 (50%)
White scales Yellow scales White shiny lines	6 (75%) 2 (25%) 5 (62,5%)

The median duration of progression before the first consultation was 22 months (range: 8–36). The prevailing clinical presentation of MPD was in the form of a plaque (50%) involving both the nipple and areola (62,5%).The majority of patients exhibited the non-pigmented form of MPD (75%). The most frequent dermoscopic pattern of MPD are resumed in (Table 2) and were white scales (75%), shiny white lines and linear vessels (62,5%) (Figure 1), followed by structure less pink areas (50%), dotted vessels (50%) (Figure 2), erosion/ulceration (50%), Yellow scales (25%) and structurless white areas (12,5%). In the cases of pigmented MPD, we noted the presence of brown/grey globules and dots (12,5%) (Figure 3). In all our cases, the diagnosis of MPD was confirmed by histology showing an intradermal tumor proliferation composed of small clusters or isolated large cells with abundant clear cytoplasm and vesicular nuclei featuring prominent nucleoli (Figure 4).

Discussion

In 1874, Sir James Paget introduced the concept of Paget's disease (PD) of the breast, identifying this condition in 15 women exhibiting chronic eczematous skin changes on the nipple and areola [6]. Remarkably uncommon, PD constitutes a mere 1-4% of all breast cancer cases [7]. The zenith of incidence for mammary Paget's disease rests between 50 and 60 years of age, with its occurrence predominantly confined to the female population, rendering it exceedingly rare among males. In around 90% of MPD cases, an underlying breast adenocarcinoma can be identified [1,2]. MPD typically exhibits a gradual onset that unfolds over a period of months to years, frequently manifesting unilaterally within a single breast. The ailment commonly initiates within the nipple region and subsequently progresses towards the areolar region, occasionally extending to involve surrounding skin in advanced stages. The lesions manifest as eczematoid, erythematous, thickened, moist, or crusted areas, characterized by irregular borders. These may encompass fine scaling, induration, infiltration, secretion, bleeding, ulceration, and even the occurrence of nipple invagination [8]. The most prevalent clinical presentation of MPD in our serie was an erythematous plaque involving the nipple and areola complex. While the condition is more commonly associated with post-menopausal individuals or those in their sixth decade [8], it is pertinent to note that 2 of our patients were in their forties and weren't in a menopausal state.

Given the multifaceted range of clinical presentations, individuals displaying these features often encounter diagnostic challenges, initially being managed under the presumption of various other conditions such as eczema, psoriasis, melanoma, infections, or traumatic events [8-10]. Dermoscopy emerges as a highly promising instrument in discerning MPD from alternative diagnoses. In the literature, there are 34 dermoscopic descriptions in case reports of MPD [11] and only one case-control retrospective study including 22 cases [12]. Z. Appala et al. discovered that the prevailing dermoscopic aspects in MPD consisted primarily of white scales (86.4%) and structureless pink areas (81.8%). These were succeeded by the presence of dotted vessels (72.7%), erosion/ulceration (68.2%), and white shiny lines (63.6%). In our study, white scales were also the most prevalent dermoscopic pattern. However, in contrast to Appala's findings, linear vessels exhibited a higher frequency than dotted ones which can be explained by the fact that the epidermis in MPD may undergo squamous or papillomatous hyperplasia, resulting in distinct orientations of the underlying vessels [13]. Regarding PMPD, dermoscopic observations encompass atypical pigment networks, structureless areas in shades of blue-grey, brown or blue dots, and globules, alongside additional non-pigmented manifestations such as structureless pink and/or white areas [12].

Especially when dealing with patients with skin of color, there is a pressing need for more comprehensive dermoscopic characterizations, given that even benign lesions (such as lentigo, seborrheic keratosis, nevi, and nipple-areola melanosis) can bear resemblance to PMPD. Hence, any newly appearing or changing pigmented lesions within the nipple-areola complex should be subjected to biopsy [14].

Conclusion

Ours is the first Moroccan study describing the dermoscopic patterns of MPD although its limitation by a modest sample size and the lack of a control group. The prevailing characteristic of an individual affected by MPD typically corresponds to an elderly woman exhibiting a unilateral, asymptomatic, erythematous patch or plaque affecting the nipple. Dermoscopically, this condition often manifests as pink structureless areas, scattered by white scales, linear and dotted vessels. There is the potential to observe areas with brownish pigmentation, as well as pigmented dots or granules in the pigmented form of MPD.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

Conflicts of Interest

The authors do not declare any conflict of interest

References

1. Waldman RA, Finch J, Grant-Kels JM, Stevenson C, Whitaker-Worth D. Skin diseases of the breast and nipple: benign and malignant tumors. J Am Acad Dermatol. 2019; 80: 1467-1481.
2. Gaurav A, Gupta V, Koul R, Dabas S, Sareen R, Geeta K, et al. Practical consensus recommendations for Paget's disease in breast cancer. South Asian J Cancer. 2018; 7: 83-86.
3. Errichetti E, Avellini C, Pegolo E, De Francesco V. Dermoscopy as a supportive instrument in the early recognition of erosive adenomatosis of the nipple and mammary Paget's disease. Ann Dermatol. 2017; 29: 365-367.
4. Gunhan-Bilgen I, Oktay A. Paget's disease of the breast: clinical, mammographic, sonographic and pathologic findings in 52 cases. Eur J Radiol. 2006; 60: 256-263.
5. Cinotti E, Galluccio D, Ardigo M, Spataro G, Perrot JL, Rubegni P, et al. Nipple and areola lesions: dermoscopy and reflectance confocal microscopy features. J Am Acad Dermatol. 2018.
6. Paget J. On disease of the mammary areola preceding cancer of the mammary gland. St Bartholomew's Hosp Rep. 1874; 10: 87-89.
7. Valdes EK, Feldman SM. Paget's disease of the breast. Breast J. 2006; 12: 83.
8. Lopes Filho LL, Lopes IM, Lopes LR, Enokihara MM, Michalany AO, Matsunaga N. Mammary and extramammary Paget's disease. An Bras Dermatol. 2015; 90: 225-231.
9. Brugues A, Iranzo P, Diaz A, Pena A, Estrach MT, Carrera C. Pigmented mammary Paget disease mimicking cutaneous malignant melanoma. J Am Acad Dermatol. 2015; 72: 97-98.
10. Liu J, Zhao Z, Zhou J, Zhang J. An unusual presentation of Paget's dis- ease of the nipple in a young woman: a case report. Int J Clin Exp Med. 2015; 8: 4694-4696.
11. Cinotti E, Galluccio D, Tognetti L, Habougit C, Manganoni AM, Venturini M, et al. Nipple and areola lesions: review of dermoscopy and reflectance confocal microscopy features. J Eur Acad Dermatol Venereol. 2019; 33: 1837-1846.
12. Apalla Z, Errichetti E, Kyrgidis A, Stolz W, Puig S, Malvehy J, et al. Dermoscopic features of mammary Paget's disease: a retrospective case-control study by the International Dermoscopy Society. J Eur Acad Dermatol Venereol. 2019; 33: 1892-1898.
13. Weedon D. Tumors of cutaneous of appendages, extramammary Paget's disease. In Weedon D, ed. Skin Pathology, 3rd edn. Elsevier, China. 2010; 33: 790.
14. Robles-Tenorio A, Rendon A. Dermoscopic features of pigmented mammary Paget disease. Dermatol Pract Concept. 2023; 13.