

Polarized dermoscopy of mammary Paget disease*

Dermatoscopia da doença de Paget mamária

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Abstract: Mammary Paget's disease is a rare intraepithelial adenocarcinoma, located on the nipple/areola complex, highly associated with breast cancer. Although the international literature emphasizes the dermatoscopic pattern of mammary Paget's disease pigmented variant, the authors describe the dermoscopic findings of classical Paget's disease and demonstrate the presence of chrysalis-like structures, criteria recently described in the literature and not yet reported in Paget's disease.

Keywords: Dermoscopy; Microscopy, polarization; Paget's disease, mammary

Resumo: Doença de Paget mamária é considerada um adenocarcinoma intra-epitelial raro, localizado no complexo mamilo-aréola,com alta associação ao câncer de mama. Apesar da literatura mundial realçar o padrão dermatoscópico da doença de Paget mamária variante pigmentada os autores descrevem os achados dermatoscópicos da doença de Paget clássica realçando a presença das estruturas crisálida-símiles, critério recentemente descrito na literatura mundial e ainda não relatado na Doença de Paget.

Palavras-chave: Dermoscopia; Doença de Paget mamária; Microscopia de polarização

CASE REPORT

A 77- year- old woman was referred to our department by a gynecologist, regarding a 5- year history of progressively enlarged erythematous plaque in her left breast, with occasional burning. She underwent a radical mastectomy and lymph node dissection of the right breast due to adenocarcinoma diagnosed 15 years before the onset of this skin lesion (Figure 1). Clinical appearance of the lesion shows an eczematous-like plaque with a thin, asymmetric and scaly surface, brownish-pink pigmentation in the left areola and nipple with centrifugal growth associated

with nipple retraction (Figure 1). General physical examination revealed no abnormalities. There were no palpable masses in the breast or axillary lymphadenopathy. Dermoscopic examination of the central portion of the lesion showed a whitish-pink area with irregular linear vessels and chrysalis-like structures (Figure 2), whereas the darker portion was characterized by dark brown diffuse pigmentation with irregular blue-gray dots (Figure 2). The clinical and dermoscopic features, although not specific, suggestmammary Paget's disease or amelanotic

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melanoma. Incisional biopsy was performed. Histopathological sections stained with hematoxylineosin showed large atypical cells with hyperchromatic eccentric nuclei and abundant cytoplasm throughout all levels of the epidermis (Figure 1). The tumor cells contained melanin granules within the cytoplasm and were positive for periodic acid-Schiff (PAS) staining - compatible with Paget's disease of breast (Figure 1). The patient was referred to surgical excision and radical mastectomy and hormonal blockade was carried out.

COMMENTS

Mammary Paget's disease, first described in 1874, is an intraepidermal adenocarcinoma of the areola and/or nipple, generally associated with carcinoma of the underlying lactipherous ducts. ^{1,2} It is an uncommon form of breast cancer with prevalence in the sixth decade of life, developing in 70% of cases in postmenopausal women. ¹

Clinically, Paget's disease mimics inflammatory and infectious diseases; in dermoscopy, however, the diagnosis of their pigmented variant is difficult, since

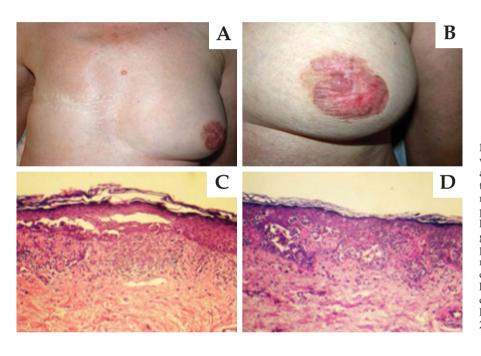


FIGURE 1: A. Clinical appearance of lesion with radical mastectomy scar on the right and change in morphology and pigmentation of the left nipple and areola; B. In more detail, lesion shows erythematous plaque on the left areolar region, with pinkish-brown color associated with centrifugal growth and retraction of the left nipple; C. Histopathological sections of mammary Paget's disease: aggregates of atypical cells in the epidermis, with large hyperchromatic nuclei and abundant cytoplasm (HE-200x); D. Melanin granules in the cytoplasm of tumor cells (PAS-200x)

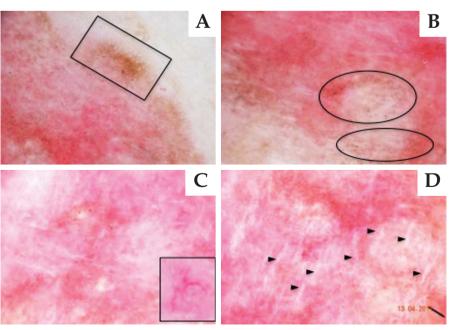


FIGURE 2: Dermatoscopic appearance of lesion. On dermoscopy the lighter portion corresponded to a whitish-pink area of the left nipple and areola, whereas the darker portion was characterized as follows: A. Light brown diffuse pigmentation and B. Irregular black dots and small blue-gray structures (peppering) irregularly distributed; C. Irregular linear vessels and D. Under polarized light, the presence of bright white streaks structures called chrysalis-like structures; note the parallel arrangement of these structures

it displays a nonspecific pattern, with diffuse irregular pigmentation and regression-like structures, as observed in melanoma.² So far, there are no descriptions in the literature of the dermoscopic criteria of the classic form of Paget's disease.^{3,4}

The presence of irregular linear vessels represents the vascular pattern commonly found in melanoma.⁵ In addition, multiple blue-gray dots (peppering) correspond to melanophages in the papillary dermis, while the white scar areas to fibrosis. Both structures can be found in regression areas of melanocytic lesions. The shiny white streaks, originally termed chrysalis-like structures, reveal shiny, bright white and linear streaks that can only be visualized with polarized light dermoscopy. The lines are generally oriented parallel or orthogonally to each other. These structures represent new or remodeled collagen bundles. Collagen bundles are visible and more evident under polarized dermoscopy due to their birrefringent properties that cause rapid randomization of polarized light. Similar criteria have been described in many conditions like dermatofibroma, scars, basal

cell carcinoma (including Pinkus' fibroepithelioma variant) and piogenic granuloma.^{6,7} When present in melanocytic lesions, they are suggestive of Spitz nevus or melanoma.⁶

The diagnosis of Paget's disease should be based not only on clinical and dermoscopic observation but also associated with histopathological and immunohistochemical analysis.^{8,9}

Surgical excision is the recommended treatment (modified radical mastectomy with lymphadenectomy), but recently studies have shown that breast-conserving surgery found no significant differences in terms of overall or disease- free survival.

Polarized dermoscopy is a versatile optical tool, easy to use, practical and that can help in early diagnosis of mammary Paget's disease and their clinical and histopathologic correlation. Until now, no description of the chrysalis-like structures in Paget's disease has been reported. Therefore, shiny white streaks (or chrysalis-like structures) are an additional dermoscopic feature to be considered in mammary Paget's disease. \square

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