

Rosettes: An Additional Clue for the Identification of the Primary Cutaneous Marginal Zone Lymphoma

Maria Eugenia Gil¹, Rosario Peralta², Maria Cecilia Laporta¹, Jimena Lorenzo¹,
María Mercedes Nussold¹

¹ School of Medicine, University of Buenos Aires, Argentina

² Dermatology Department, Instituto de Investigaciones Médicas "A. Lanari", University of Buenos Aires, Argentina

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Corresponding author: Maria Eugenia Gil, MD, Paraguay 2155, Ciudad Autónoma de Buenos Aires, Argentina.
Tel: (+54) 011-61562798, E-mail: meugenia@fmed.uba.ar

Introduction

Primary cutaneous marginal zone lymphoma (PCMZL) is a B-cell lymphoma arising in the skin and accounts for a low incidence of all primary cutaneous lymphomas (2-7%) [1]. Dermoscopic features that have been described are salmon-colored area/background, serpentine vessels, scales and white circles [2]. Rosettes are a specific form of a white shiny structure seen with polarized dermoscopy [3]. We report a case showing multiple rosettes on PCMZL.

Case Presentation

A 36-year-old male patient presented an 8-month history of an enlarging asymptomatic lesion on his lower back. Physical examination showed a well-defined, indurated, reddish plaque measuring 1 x 2 cm. Dermoscopy revealed

salmon-colored area/background, serpiginous vessels, and multiple rosettes (Figure 1).

Neither adenopathies nor other lesions were palpated in the rest of the tegument. The histopathological examination presented preserved epidermis and a dense dermal lymphoid infiltration, which was arranged in a nodular pattern. The immunohistochemistry was characterized by the expansion of the marginal zone cellular population BCL 2 + and CD 20 +. Nodal lymphocytes were CD3 - and peripheral lymphoid population was CD3+ (Figure 2).

The diagnosis was PCMZL. A blood test was performed, with normal values: hemogram, hepatogram, urine and creatinine, beta two microglobulin assay, lactate dehydrogenase (LDH) and serological test for Epstein Barr Virus (EBV) and Cytomegalovirus (CMV). The computed tomography (CT) evaluation displayed no other lesions and the surgical excision with 2 cm of diameter was performed.

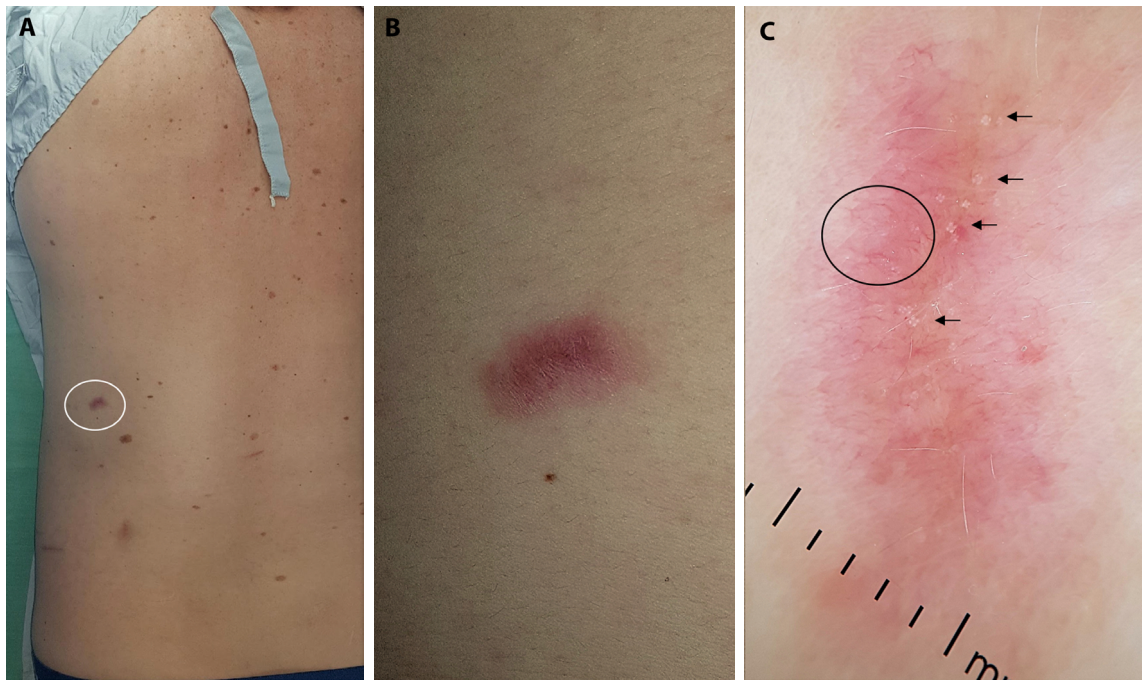


Figure 1. (A-B) Clinical presentation of a well-defined, indurated, reddish plaque measuring 1 x 2 cm on lower back (white circle). (C) Polarized dermoscopy shows a salmon-colored background with serpiginous vessels (black circle) and multiple rosettes (arrows) of different sizes.

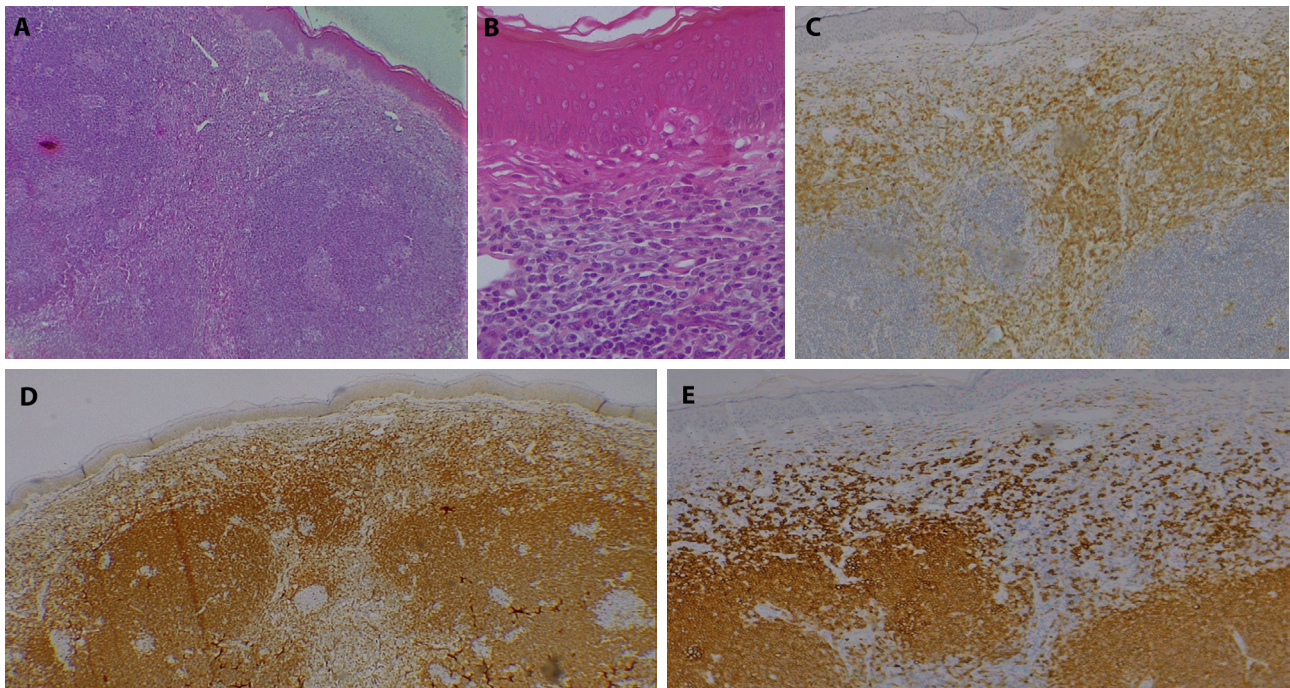


Figure 2. Histopathological analysis. (A) Dense dermal lymphoid infiltrate which was arranged in a nodular pattern (H&E, $\times 10$). (B) Preserved epidermis (H&E, $\times 40$). (C) CD3 - nodal lymphocytes/CD3 + peripheral lymphoid population. (D) BCL2 +. (E) CD 20+.

Conclusions

Rosettes are defined as 4 bright white circles arranged as a square resembling a 4-leaf clover, mainly localized in the follicular openings. Despite being well-described dermoscopically, the precise histology correlation and optical

significance are not known. There are different types that would be caused by the interaction of keratin-filled adnexal openings with the polarized light: smaller ones (0.1-0.2 mm), usually oriented at the same angle, and larger ones that would be the result of concentric fibrosis around the follicles (0.2-0.5 mm) [3].

Table 1. Dermoscopy findings of primary cutaneous B-cell lymphoma.

Dermoscopy findings of primary cutaneous B-cell lymphoma
Salmon-colored background area
Arborizing /Serpentine vessels
Scales
White circles
Rosettes

They have been characteristically described in actinic keratosis and squamous cell carcinoma. However, they were commonly seen in non-lesional actinic damaged skin, scars, many tumoral and inflammatory skin lesions and hence are not lesion specific.

Related to cutaneous lymphoproliferative disorders there are isolated reports of their presence in T-cell pseudolymphoma and classic Mycosis Fungoides in patients with skin of color [4-5].

The dermoscopic findings of primary cutaneous B-cell lymphomas are not specific, characterized by salmon-colored background/area, serpentine vessels, scales and white circles [6]. Considering that the size of the rosettes can vary, we noticed in our case multiple small rosettes like targeted follicles instead of white circles. Both signs could be a progression of the same feature [3].

Differential diagnosis of PCMZL is wide, including benign and malignant diseases. They are frequently misdiagnosed as inflammatory and infectious lesions or as other cutaneous neoplasms such as basal cell carcinoma or amelanotic melanoma.

Diagnosis of PCMZL may be challenging and histopathological examination is mandatory for a definite diagnosis. Dermoscopy could play an adjuvant role in the achievement of the diagnosis. This case provides an additional clue for its identification.

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