Dermatology Practical & Conceptual

Salmon-Colored and White Areas on Dermoscopy as Supportive Findings in the Diagnosis of Primary Cutaneous Marginal Zone Lymphoma

Giovanni Biondo[1,2], Simona Sola[3], Carlotta Pastorino[2], Cesare Massone[2]

1 Dermatology and Sexually Transmitted Disease Unit, P. Giaccone Hospital, University of Palermo, Palermo, Italy
2 Dermatology Unit, Galliera Hospital, Genoa, Italy
3 Surgical Pathology, Galliera Hospital, Genoa, Italy

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Corresponding author: Cesare Massone, MD, Dermatology Unit, Ospedali Galliera, Via Volta 6, 16128, Genova, Italy. Email: cesare.massone@galliera.it

Introduction

Dermoscopy can be a useful tool to help clinicians in the diagnosis of cutaneous B-cell lymphomas.

Case Presentation

A 61-year-old man presented with an irregularly growing erythematous plaque measuring 28 × 35 mm, which had appeared on the left thigh a few months before (Figure 1A). He was otherwise healthy and could not recall an arthropod bite or a trauma in the same area. He was not taking any drugs and did not report weight loss, fever, or night sweating. The physical examination was normal. Upon polarized non-contact dermoscopy, we observed a diffuse salmon-colored area with focally white areas (Figure 1B).

The lesion was totally excised with differential diagnosis of granulomatous dermatosis, amelanotic melanoma, cutaneous lymphoma, sarcoma, or metastasis. Histopathology showed a dense nodular dermal lymphocytic infiltrate extend-

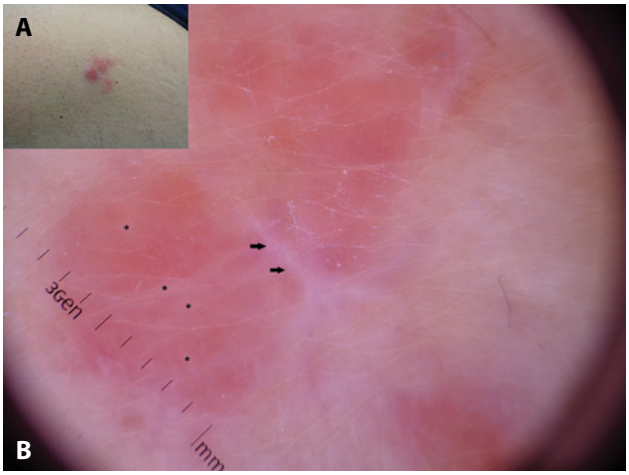

Figure 1. (A) Irregular erythematous plaque on the left thigh. (B) Polarized noncontact dermoscopy: A diffuse salmon-colored area on the background, together with focally white areas (marked with black arrows) and white circles (marked with asterisks). [Copyright: ©2019 Biondo et al.]

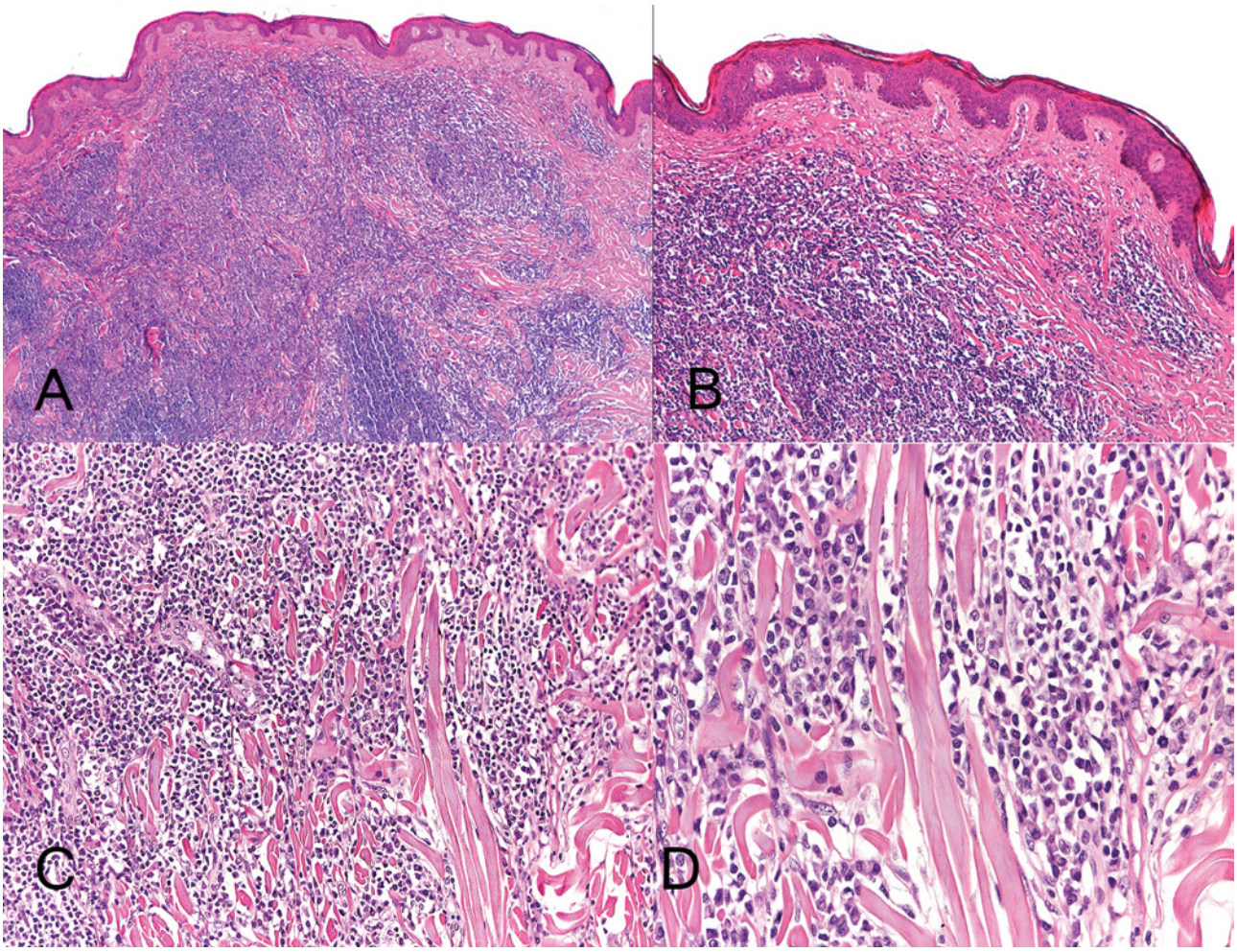

Figure 2. (A) Below a clear-cut grenz zone in the papillary dermis, there is a dense nodular dermal lymphocytic infiltrate composed of reactive lymphoid follicles surrounded by small to medium-sized cells (marginal zone cells). No dilated vessels in the papillary dermis are seen; areas of reactive fibrosis in the dermis are present (hematoxylin and eosin [H&E], ×20). (B) Focally, the grenz zone is reduced due to patchy, nodular, more superficial infiltrate in the papillary dermis (H&E, ×100). (C) Marginal zone cells, plasma cells, lymphoplasmacytoid cells, and slight increase of vessels within the infiltrate (H&E, ×200). (D) Plasma cells and lymphoplasmacytoid cells (H&E, ×400). [Copyright: ©2019 Biondo et al.]

ing to the subcutaneous fat composed of reactive lymphoid follicles surrounded by a diffuse infiltrate composed by small to medium sized cells (marginal zone cells). In addition, plasma cells (at the margins of the infiltrate), lymphoplasmacytoid cells, small reactive lymphocytes, and occasional eosinophils were observed.

A clear-cut grenz zone between the infiltrate and the epidermis was present (Figure 2). Neoplastic cells stained positive for CD20 and CD79a; cells stained negative for Bcl-6, Bcl-2, CD5, and CD10; reactive T lymphocytes were CD3+. A monoclonal expression of kappa chain was found. Further complete staging investigations were negative for nodal or systemic involvement. Following WHO criteria [1], primary cutaneous marginal zone lymphoma (PCMZL) was diagnosed.

Discussion

PCMZL is a low-grade malignant primary cutaneous B-cell lymphoma (PCBCL). Clinically, patients present with solitary or multiple, asymptomatic, rarely ulcerative pink-violet to red-brown papules, plaques, and nodules localized preferentially on the extremities or trunk. Mascolo et al. described the dermoscopic features of 10 PCBCL cases, observing white circles with a salmon-colored area in 9/10 cases, scales in 7/10 cases, and arborizing vessels or a polymorphous vascular pattern in 5 and 2 cases, respectively [2]. Piccolo et al. also described the same features in 2 additional patients [3]. Geller et al. expanded the study, examining 58 dermoscopic images of PCBCLs, and reported a salmon-colored area (79.3%) and prominent blood vessels (77.6%), mostly of serpentine (linear-irregular) morphology (67.2%) [4]. Ghahramani observed

Table 1. Dermoscopic, Clinical, and Histological Features of PCBCLs Reported in the Literature

Dermoscopic, Clinical and Histological Features of PCBCLs Reported in the Literature							
Authors	**Number of Cases**	**Sex**	**Age**	**Localization**	**Clinical Features**	**Dermoscopic Features**	**Histologic Features**
Mascolo M et al 2016 [2]	6 PCMZL 2 PCFCL 2 PCLBCL	7 M 3 F	51.9 (20-73)	3 Trunk 2 Arm 1 Forearm 1 Thigh 1 Retroauricular 1 Neck 1 Axillary	Solitary red/pinkish nodules	(9/10) White circles (6/10) Salmon-colored area (7/10) Scales (5/10) Arborizing vessels (2/10) Polymorphous vascular pattern	6 Patchy, nodular or diffuse infiltrate of small centrocyte-like cells CD20+, CD79a+, Bcl2+, Bcl6-, CD10- 2 Diffuse infiltrate of small-medium cleaved cells CD10+, Bcl6+, Bcl2- 2 Dense, diffuse infiltrate of large round cells, frequent mitoses, elevated Ki-67, Bcl2+, MUM1+, CD10-
Geller S et al 2018 [4]	31 PCMZL 14 PCFCL 10 Indolent PCBCL 3 PCLBCL	NR	NR	32 Trunk 18 Extremities 8 Head and neck	Red/violaceous papules, plaques, nodules, solitary, trunk/extremities (PCMZL) Pink/violaceous papules, nodules head (PCFCL) Red/bluish infiltrated plaques, nodules or tumor lower extremities (PCLBCL)	(46/58) Salmon-colored area (45/58) Blood vessels (6/58) Scale (4/58) Ulceration	NR
Piccolo V et al 2016 [3]	2 PCMZL	1 M 1 F	75.5 (74-77)	Breast Back	Patchy infiltrated oval erythematous lesion Multiple reddish, variably sized, firm nodule	(2/2) Arborizing vessels (2/2) Salmon-colored area (2/2) White areas/circles	Dense, diffuse, lymphoid infiltrate within the dermis and the subcutaneous fat, consisting of small-cleaved lymphocytes and lymphoplasmacytic cells admixed with plasma cells, mainly located at the periphery of the infiltrate CD20+, CD79a+, Bcl2+, CD5-, CD10, Bcl-6-
Ghahramani et al 2018 [5]	1 PCFCL 1 PCMZL	NR	NR	Helix Eyebrows	NR	Spermatozoa-like structures (PCFCL) Orange-yellowish patchy areas (PCFCL) Crystalline structures (PCFCL) Pseudopod-like vessels (PCFCL) Dull red background Structureless patches	Pseudopod-like vessels reflect dilated ectatic vessels surrounding the neoplastic follicles

NR = not reported; PCBCL= primary cutaneous B-cell lymphoma; PCFCL = primary cutaneous follicle center lymphoma; PCLBCL = primary cutaneous large B-cell lymphoma; PCMZL = primary cutaneous marginal zone lymphoma.

spermatozoa-like structures, orange-yellowish patchy areas, crystalline structures, and pseudopod-like vessels in primary cutaneous follicle center lymphoma, but not in PCMZL [5]. These findings are summarized in Table 1.

Our observation confirms that a salmon-colored area upon dermoscopy is typical even if not exclusive of PCBCL. In fact, it brings to mind the orangish-yellowish areas observed in granulomatous dermatoses such as sarcoidosis, lupus vulgaris, and granulomatous rosacea, where also linear branching vessels are seen [6,7], while granuloma annulare shows peripheral structureless reddish-yellowish-orange areas with variable blurry vessels [8,9]. It is difficult to correlate the salmon color of PCBCL with pathology; an explanation could reside in the increased vascularization inside the dense nodular neoplastic lymphoid infiltrate in the mid and deep dermis (Figure 2C). White areas probably correlate with areas of reactive fibrosis in the dermis or might correlate with focally reduced grenz zone due to patchy, nodular, more superficial infiltrate in the papillary dermis (Figure 2B). Recently, pseudopod-like vessels have been correlated with dilated ectatic vessels surrounding the neoplastic follicles [5].

As further dermoscopic differential diagnosis, amelanotic melanoma shows dotted vessels,

linear-irregular vessels, hairpin-irregular vessels, serpentine vessels, or a combination of them (polymorphic vessels); also milky-red areas can be frequently visualized [10]. Contrary to previous experiences [2,3], in our patient prominent vessels with serpentine morphology were not observed. Also on dermoscopic-pathologic correlation, no dilated vessels were observed in the superficial dermis; prominent blood vessels have been correlated with neoangiogenesis, a phenomenon that in our case probably did not occur yet in the superficial dermis because it might be correlated with a different stage of the disease.

Conclusions

In conclusion, a salmon-colored area and white areas on dermoscopy might be suggestive, even if not unique, of PCBCL, supporting clinicians in recognizing primary lesions and recurrences [11], moreover identifying correct site of biopsy.

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