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Deceitful clustered papules on the scalp of a middle-aged woman

Massimo Mascolo[1], Francesca Pagliuca[1], Claudia Costa[2], Massimiliano Scalvenzi[2]

1 Department of Advanced Biomedical Sciences, Pathology Section, University of Naples Federico II, Naples, Italy
2 Department of Dermatology, University of Naples Federico II, Naples, Italy

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Corresponding author: Massimo Mascolo, MD, PhD, Department of Advanced Biomedical Sciences, Pathology Section, University of Naples Federico II, Via Pansini, 5, 80131, Naples, Italy Naples, Italy. Tel &Fax. +390817464532/3414. Email: massimo.mascolo@unina.it

The patient

A 67-year-old woman came to dermatologic examination for the presence of an inconspicuous lesion on the scalp; time of onset could not be established. The lesion was located on the parietal region and consisted of a small erythematous nodule (Figure 1a). Nevertheless, clustered reddish papules on the right temporal region caught the dermatologists' attention (Figure 1b).

Dermoscopic evaluation of the temporal lesion showed a red-to-pink homogeneous area with polymorphous vascular pattern and one peripheral arborizing vessel (Figure 2).

Biopsy of the parietal lesion revealed a nodular proliferation of basaloid cells with peripheral palisading and areas of dedifferentiation surrounded by nodular lymphoid infiltrates (Figure 3a). Similarly, the temporally located lesion was characterized by a dense small-sized lymphoid infiltrate within the dermis (Figure 3c). Staining for CD20, CD79a, Pax-5, Bcl-6, CD10 and Bcl-2 revealed strong positivity of all lymphoid cells at both sites (Figure 3b,d); CD3, CD5, CD138 and MUM-1 resulted negative. The proliferation rate was low.

What is your diagnosis?

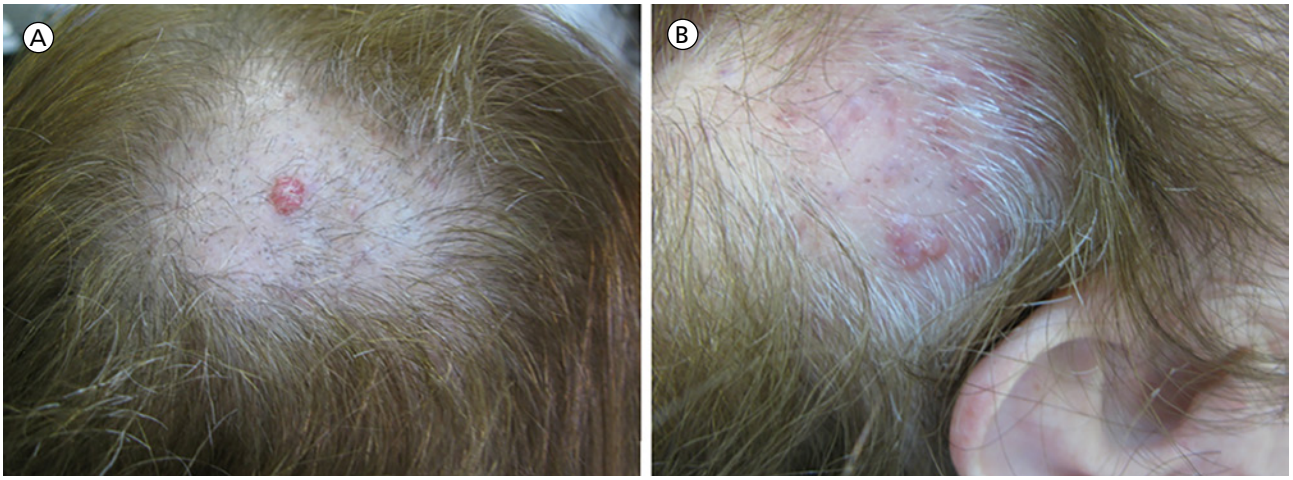

Figure 1. Clinical appearance of the erythematous nodule on the parietal region (a) and of the clustered reddish papules on the right temporal region (b). [Copyright: ©2017 Mascolo et al.]

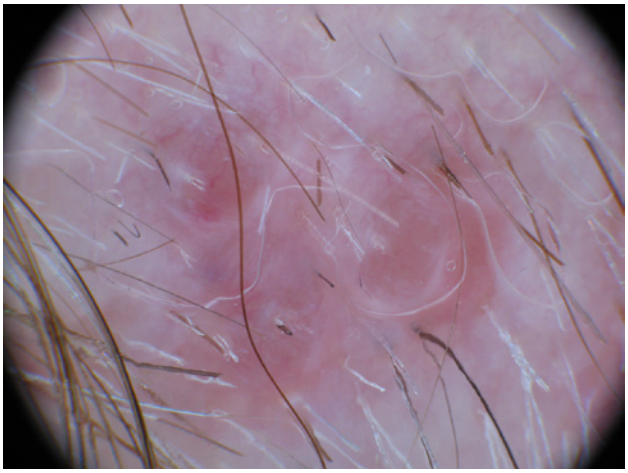

Figure 2. Dermoscopic appearance of the temporal papules. [Copyright: ©2017 Mascolo et al.]

Answer

Cutaneous follicle center B-cell lymphoma coexisting with nodular basal cell carcinoma on the parietal region.

Discussion

The case presented above of cutaneous collision tumors shows an unusual association of a very common and easily diagnosed skin malignancy (basal cell carcinoma) and a rare one (primary cutaneous follicle B-cell lymphoma).

Primary cutaneous follicle B-cell lymphomas (PCFBCLs) are lymphoproliferative proliferation of germinal center B-cells that affect the skin in the absence of extracutaneous involvement at time of the diagnosis. PCFBCL represents a low-grade tumor with a good prognosis, which predominantly affects middle-aged patients with no sex predilection.

Clinical features are manifold and aspecific: it usually presents as solitary or multiple, regionally clustered erythematous or violaceous plaques or nodules located in the head and neck region or on the trunk [1]. Confusion with other malignant or non-malignant conditions may easily occur. Among these, basal cell carcinoma (BCC) deserves particular attention. It is a far more common neoplasm affecting sun-exposed skin, which shares with PCFBCL site predilection (head and neck and trunk) and clinical manifestation, such as erythematous nodule or papule. Infrequently, BCC may collide with other epithelial or non-epithelial tumors: the most reported combination is with melanocytic proliferations. In the given case, coexistence at the same site of one much simpler to recognize lesion (basal cell carcinoma) with a quite rare one (PCFBCL), could be misleading and result in missing the right diagnosis.

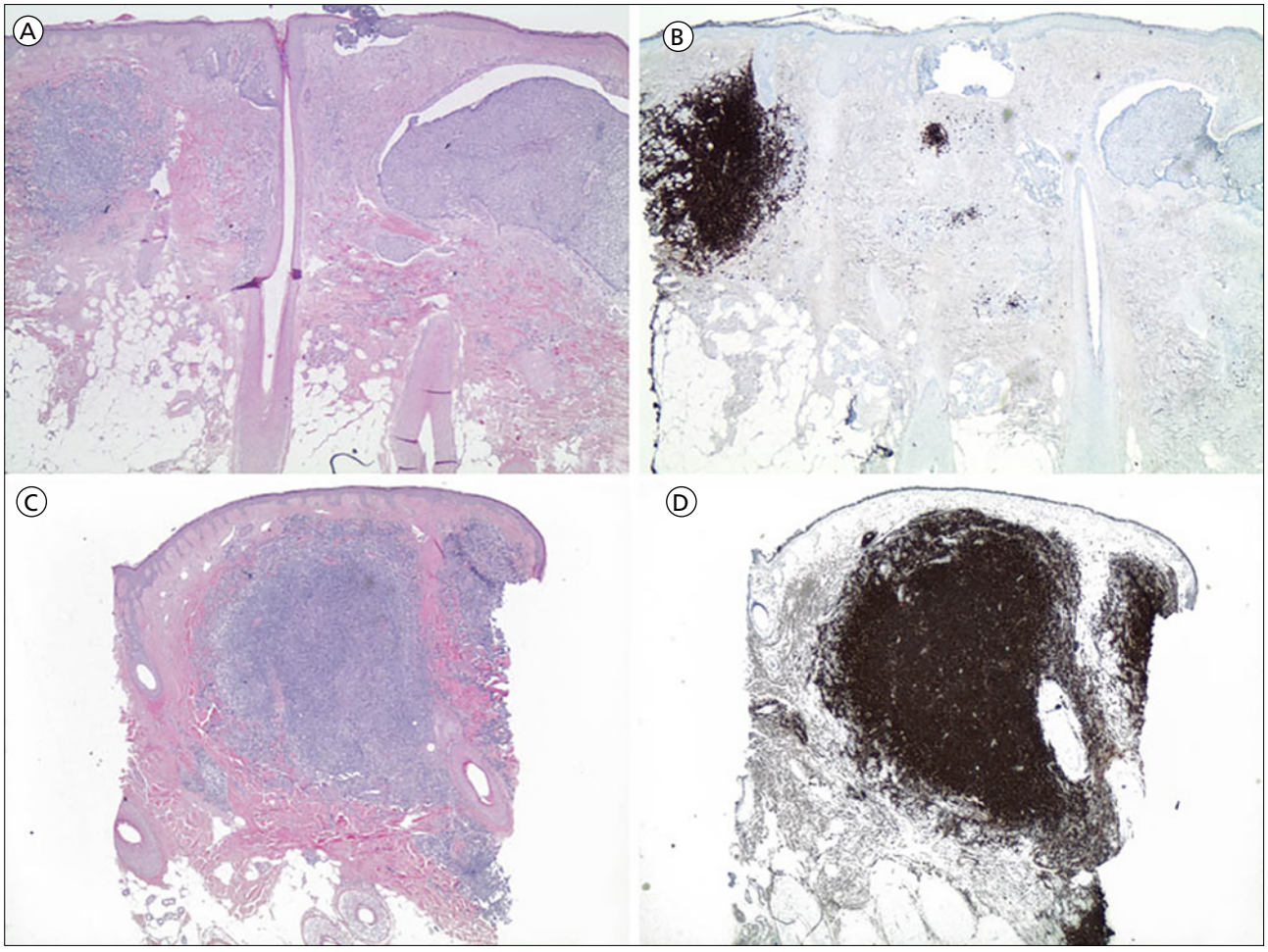

Figure 3. Histopathology. Microscopic appearance of the parietal nodule (a) and of the temporal lesion (c); immunostaining for CD20 at both sites (b, d). [Copyright: ©2017 Mascolo et al.

Although a dermoscopic pattern for CBCLs has recently been described [2,3], consisting of the presence of thin arborizing vessels, scales and white circles/areas in a salmon-colored background, it could be unspecific; therefore, histopathology remains crucial for their identification.

Histopathological appearance of PCFBCL consists in a dense, follicular or diffuse lymphoid infiltrate comprised of small-to-intermediate sized centrocytes admixed with centroblasts, with interspersed reactive T-lymphocytes. It involves the dermis and occasionally the hypodermis with sparing of the epidermis. Neoplastic cells are immunoreactive for CD20, CD79a, Pax-5, CD10 and Bcl-6 whereas Bcl-2 is usually negative. The Ki-67 labeling index demonstrates a reduced and non-polarized proliferation rate in contrast with reactive germinal centers; thus it can be useful in the differential diagnosis with cutaneous lymphoid hyperplasia.

BCC is usually a straightforward diagnosis on histological slides. Proliferation of elongated basaloid cells with recognizable mitotic activity, forming nests or growing in a nodular or, uncommonly, infiltrative fashion, usually accompanied by solar elastosis or other dermal degenerative changes is the distinctive microscopic appearance of BCC. No ancillary studies are routinely needed.

While narrow local excision with negative margins is almost always to be considered curative for BCCs, treatment strategies for CBCLs include surgical excision, radiotherapy, chemotherapy, anti-CD20 agents (rituximab) and immunotherapy. Combined radio-chemotherapy is needed only when disseminated disease occurs. Prognosis is very good, with 5-year survival rates, which range from 95% to 100%; however, recurrences are common even in those cases with complete clinical response to therapy, so that long-term follow up is required.

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