Portuguese Journal of Dermatology and Venereology

PERMANYER

Check for updates

Port J Dermatol and Venereol.

CASE REPORT

Dermatoscopic features of blastic plasmacytoid dendritic cell neoplasm: a case report and review of the literature

Características dermatoscópicas de neoplasia blástica de células dendríticas plasmocitoides: relato de caso e revisão da literatura

Daniel Argüello-Ruiz[1], A. Camila Martín-Zamora[1], and Betzabé Rojas-Mena[2]*

[1]Department of Dermatology; [2]Department of Pathology. Hospital Calderón Guardia, San José, Costa Rica

Abstract

Blastic plasmacytoid dendritic cell neoplasm is a malignant, aggressive, and rare tumor arising from plasmacytoid dendritic cell precursors. Because of its infrequency, information on clinical features and best treatment options is still lacking. Dermatoscopic features on this entity have only been reported in a few cases so far. Therefore, we describe a new case and review published data on this topic.

Keywords: Blastic. Plasmacytoid. Dendritic. Neoplasm. Dermatoscopy. Tumor.

Resumo

Neoplasia de células dendríticas plasmocitoides blásticas (BPDCN) é um tumor maligno, agressivo e raro que surge de precursores de células dendríticas plasmocitoides. Devido à sua infrequência, ainda faltam informações sobre características clínicas e melhores opções de tratamento. Características dermatoscópicas nesta entidade foram relatadas apenas em alguns casos até agora. Portanto, descrevemos um novo caso e revisamos dados publicados sobre este tópico.

Palavras-chave: Blastico. Plasmocitóide. Dendrítico. Neoplasia. Dermatoscopia. Tumor.

Correspondence:
A. Camila Martín-Zamora
E-mail: acamilamartin@gmail.com

Received: 27-04-2024
Accepted: 02-07-2024
DOI: 10.24875/PJDV.24000040

Available online: 15-07-2024
Port J Dermatol and Venereol. 2024;82(4):278-281
www.portuguesejournalofdermatology.com

Introduction

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is an uncommon hematologic neoplasm, arising from plasmacytoid dendritic cell precursors. It tends to have an aggressive course and a poor prognosis. Presentation most commonly includes cutaneous lesions, with or without bone marrow involvement and leukemic spread[1]. Skin involvement may manifest as a solitary plaque/nodule with a brown to violaceous bruise-like tonality or as multiple nodules[2]. Dermatoscopic characteristics in this entity have seldom been published.

We present a case report of this infrequent neoplasm, describing dermatoscopic findings in our patient.

Clinical case

A 78-year-old male patient presented with a 4-month history of a painful indurated nodule on his scalp. Physical examination was notable for an erythematous-violaceous nodule of 3 × 3 cm on the vertex of the scalp (parieto-occipital region) (Fig. 1). Cervical lymphadenopaties were present.

Dermatoscopy revealed rosettes, perifollicular erythema, and nonblanchable red-violaceous structures. Some orange structureless areas were also evidenced (Fig. 2).

Histopathology showed infiltration of the dermis and hypodermis by cells with a blastic appearance, with positive TDT, focal CD43, Bcl-2, CD123, TCL-1, CD4, CD56, and Ki67 index of 30%. The diagnosis was compatible with BPDCN (Fig. 3).

Flow cytometry of the skin biopsy documented a cell population with an immunophenotype compatible with the same diagnosis.

Bone marrow aspirates evidenced a monomorphic hypercellular bone marrow. Flow cytometry reported 90% of cells with the same pathological immunophenotype. The leukemia spread was also noted, with a total of 59.120 leukocytes/μL, 77% of which were described as large cells of a lymphocytic appearance.

Treatment was started with chemotherapy, including 1 cycle of cyclophosphamide, doxorubicin, vincristine, prednisone followed by 5 cycles of cyclophosphamide, vincristine, dacarbazine. The patient presented full remission which lasted for roughly 2 months, subsequently undergoing relapse and death.

Discussion

BPDCN is a rare hematologic malignancy with variable forms of presentation. Cutaneous involvement is found in most cases[1].

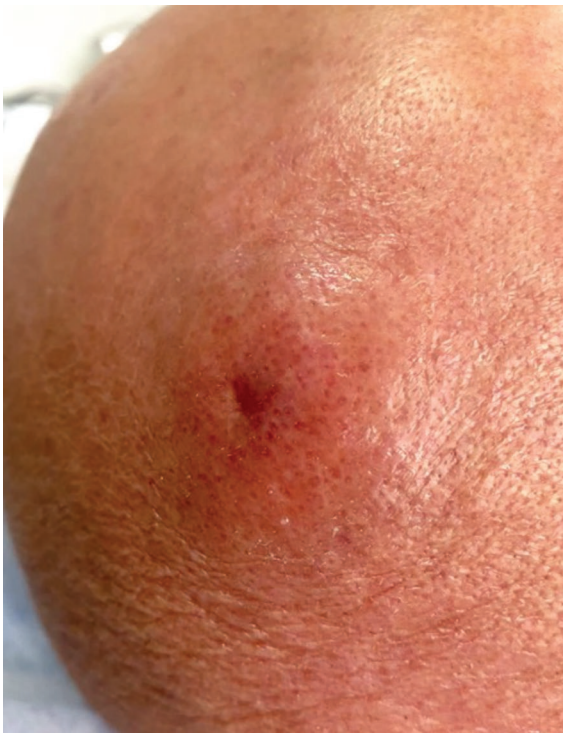

Figure 1. Erythemato-violaceous indurated nodule on the vertex of the scalp.

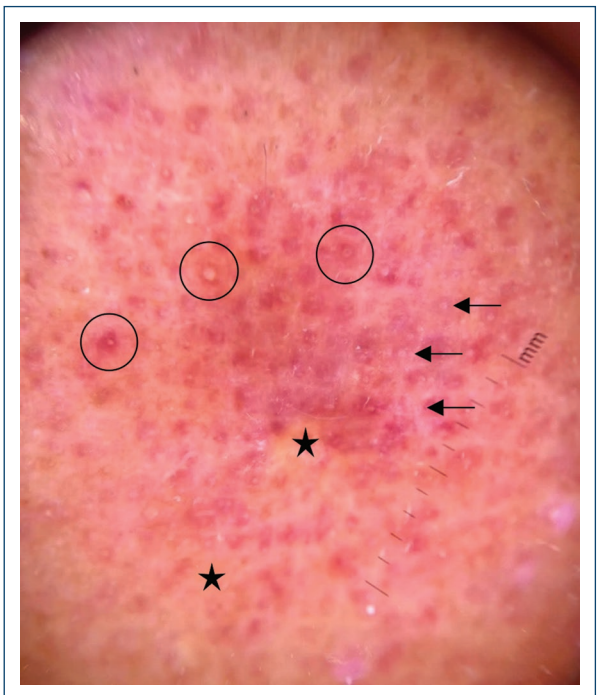

Figure 2. Dermatoscopy evidenced rosettes (arrows), perifollicular erythema (circles), nonblanchable red-violaceous structures, and orange structureless areas (stars).

Table 1. Clinical and dermoscopic characteristics documented in blastic plasmacytoid dendritic cell neoplasm

Age, sex	Location	Clinical morphology	Dermatoscopic features	Author, year of publication
75, male	Scalp (frontal and parietal region)	Large purplish, bruise-like lesion with nodular formation	Areas varying from red to purple with yellowish perifollicular structures	Martín-Carrasco et al., 2019[3]
14, male	Cheeks and left gluteus	Cheeks: gray-cyanotic dark spots resembling bruises. Left gluteus: bluish-purple nodule	Polymorphic large bluish-purple spots with a tendency to merge	Valiev et al., 2019[4]
65, male	Upper trunk	Asymptomatic nodules and plaques	Reddish and purplish nonblanchable structureless areas, surrounded by white halos. Homogeneous bluish-white areas	Nicklas et al.[5]
61, female	Upper trunk	Violaceous nodules and plaques	Pink-to-purple structureless areas with a white halo	Nicklas et al.[5]
78, male	Scalp (parieto-occipital region)	Erythemato-violaceous nodule	Rosettes, perifollicular erythema, and nonblanchable red-violaceous structures. Orange structureless areas	Case report in the present article

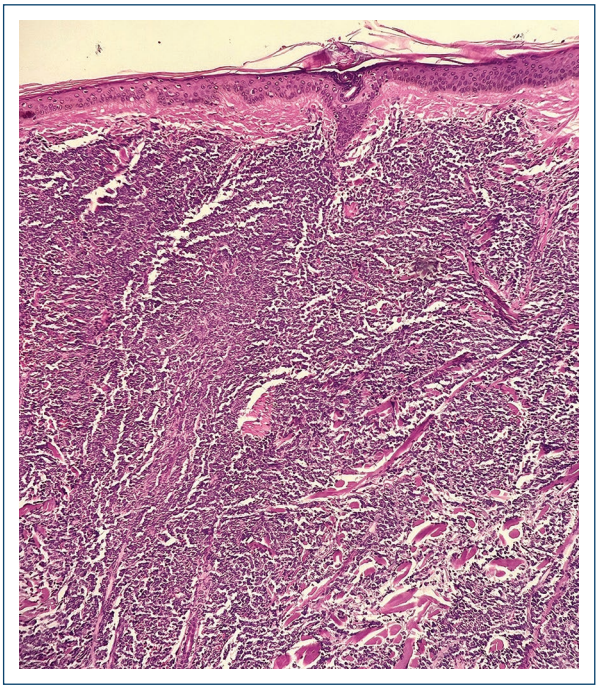

Figure 3. Histopathology evidenced a dense lymphocytic infiltrate forming cellular sheets, dissecting between collagen fibers, enveloping adnexal structures, and projecting deeply to the subcutaneous tissue. Red blood cell extravasation was also present.

Dermatoscopic features in this entity have been described only in a few cases, with pink/red to violaceous homogeneous areas, bluish areas or spots, and white halos as the most frequently described findings[3-5]. Nicklas et al. have hypothesized that the violaceous structureless areas may be related to the dermal infiltration of neoplastic cells and the presence of hemorrhage within the tumor and the white and the homogeneous bluish areas may be associated with fibrosis[5].

Our patient presented with some of these previously described features, such as nonblanchable red-violaceous structures that could correspond with the lymphocytic infiltrate and red blood cell extravasation. We also describe characteristics not previously documented, such as rosettes and perifollicular erythema that we believe correspond to perifollicular infiltration of neoplastic cells, and rosettes may represent the follicular occlusion due to this perifollicular infiltration and the presence of fibrosis.

Dermatoscopic features described in distinct types of lymphomas are very polymorphic. In our case, findings were different from those described in other lymphoproliferative tumors presenting as nodules or plaques. Nodular/plaque-type primary cutaneous T- and B-cell lymphomas more often present unfocused vessels (linear, dotted, and linear-curved), focal white and orange structureless areas and white lines[6,7]. Orange structureless areas appear to be the most strongly associated feature with these lymphomas, believed to be correlated with nodular lymphocytic infiltrates in the dermis[7]. Interestingly, although not evidenced in previous cases,

our patient presented this feature. Rosettes have also been reported in T-cell pseudolymphoma, classic mycosis fungoides, and recently in a case of primary cutaneous marginal zone lymphoma[8-10].

BPDCN is an uncommon tumor that prompts early diagnosis due to its aggressive behavior. Because of its infrequency, there are still no well-defined dermatoscopic criteria that may aid in its diagnosis. Furthermore, different clinical and histopathological presentations will also lead to variable features, as described in the different reported cases. Dermatoscopic features such as red/pink to violaceous structureless and white halos may be a clue to this diagnosis, but we observed new characteristics such as rosettes, perifollicular erythema, and orange structureless areas. These findings in association with a compatible clinical presentation must lead to early biopsy and additional studies.

Funding

None.

Conflicts of interest

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript or for the creation of images, graphics, tables, or their corresponding captions.

References

1. Pagano L, Valentini CG, Grammatico S, Pulsoni A. Blastic plasmacytoid dendritic cell neoplasm: diagnostic criteria and therapeutical approaches. Br J Haematol. 2016;174:188-202.
2. Julia F, Petrella T, Beylot-Barry M, Bagot M, Lipsker D, Machet L, et al. Blastic plasmacytoid dendritic cell neoplasm: clinical features in 90 patients. Br J Dermatol. 2013;169:579-86.
3. Martín-Carrasco P, Domínguez-Cruz J, Sendin-Martin M, Zulueta-Dorado T, Conejo-Mir J. Blastic plasmacytoid dendritic cell neoplasm mimicking both clinical and dermatoscopic angiosarcoma of the scalp. Hong Kong J Dermatol Venereol. 2019;27:27-30.
4. Valiev TT, Seregin GZ, Serebryakova IN, Chernyshova OA, Kupryshina NA, Palladina AD, et al. Blastic plasmacytoid dendritic cell neoplasm. *Pediatr Hematol Oncol Immunopathol*. 2019;18:79-89.
5. Nicklas C, Bellolio E, Lopez E, Goldberg I, Geller S, Navarrete-Dechent C. Dermoscopy of blastic plasmacytoid dendritic cell neoplasm in two patients. Clin Exp Dermatol. 2021;46:950-2.
6. Errichetti E, Geller S, Zalaudek I, Longo C, Kyrgidis A, Akay BN, et al. Dermatoscopy of nodular/plaque-type primary cutaneous T- and B-cell lymphomas: a retrospective comparative study with pseudolymphomas and tumoral/inflammatory mimickers by the International Dermoscopy Society. J Am Acad Dermatol. 2022;86:774-81.
7. Sławińska M, Sokołowska-Wojdyło M, Olszewska B, Nowicki RJ, Sobjanek M, Zalaudek I. Dermoscopic and trichoscopic features of primary cutaneous lymphomas - systematic review. J Eur Acad Dermatol Venereol. 2021;35:1470-84.
8. Alves RG, Ogawa PM, Enokihara MM, Hirata SH. Rosettes in T-cell pseudolymphoma: a new dermoscopic finding. An Bras Dermatol. 2021;96:68-71.
9. Nakamura M, Huerta T, Williams K, Hristov AC, Tejasvi T. Dermoscopic features of mycosis fungoides and its variants in patients with skin of color: a retrospective analysis. Dermatol Pract Concept. 2021;11:e2021048.
10. Gil ME, Peralta R, Laporta MC, Lorenzo J, Nusshold MM. Rosettes: an additional clue for the identification of the primary cutaneous marginal zone lymphoma. Dermatol Pract Concept. 2023;13:e2023176.