Case Report

Blastic plasmacytoid dendritic cell neoplasm mimicking both clinical and dermatoscopic angiosarcoma of the scalp

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Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare variant of cutaneous lymphoma and mainly occurs in elderly patients, mostly presenting as multiple purplish tumours or nodules. This case shows an unusual presentation of BPDCN, clinical and dermatoscopically simulating an angiosarcoma of the scalp.

Keywords: Angiosarcoma, blastic plasmacytoid dendritic cell neoplasm, cutaneous angiosarcoma of head and neck, cutaneous lymphoma, dermoscopy

Introduction

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare variant of cutaneous lymphoma (0.7%) which originates from plasmacytoid dendritic cells. Clinically, it usually presents as multiple cutaneous lesions of highly variable morphology such as purplish tumours or nodules, bruise-like spots, or as an isolated tumour. Leukaemic dissemination is infrequent in the initial stages of the disease, but may progress rapidly to a fulminant leukaemia with an average survival...
of 8.7 months. On the other hand, cutaneous angiosarcoma is a tumour that originates from endothelial cells with an aggressive course that often presents as a bruise-like lesion with erythematous-purplish nodules. Cutaneous angiosarcoma of the head and neck of the elderly is the most common type of cutaneous angiosarcoma. Dermatoscopic findings of cutaneous angiosarcoma have been reported as areas of different colour gradient (light red-purple). We present a case of BPDCN simulating an angiosarcoma of the scalp illustrating the considerable clinical and dermatoscopic similarities between these two diseases.

**Case report**

A 75-year-old male was referred to our dermatology unit for evaluation of asymptomatic lesions that had started two months previously in the frontal region and had expanded to cover both the left and right parietal regions. On examination, there was a large purplish, bruise-like lesion with nodular formation (Figure 1). Dermoscopy revealed areas varying from red to purple with yellowish perifollicular structures (Figures 2a and 2b). Histopathological examination showed a dense diffuse infiltrate of monomorphic blastic cells that did not involve the Grenz zone (Figures 3a and 3b), and the neoplasm was positive for CD4, CD56, CD123, CD303 and TCL1 markers (Figure 4). EBV test was negative thus supporting a diagnosis of blastic plasmacytoid dendritic cell neoplasm. Whole body CT showed the presence of lymphadenopathy suggestive of malignancy in both inguinofemoral regions (Figure 5). Bone marrow biopsy and blood smear were negative. The patient underwent CHOP therapy (6 cycles),

**Figure 1.** Purplish, bruise-like lesion with nodular formation in the frontal and both parietal regions.

**Figure 2.** Dermoscopy image: Areas varying from red (red arrow) to purple (purple arrow) with yellowish perifollicular structures (yellow arrow).
resulting in partial remission one year after starting treatment.

**Discussion**

BPDCN is a rare variant of cutaneous lymphoma (0.7%) that originates from plasmacytoid dendritic cells. It tends to present in elderly patients and more frequently in women (male to female ratio 2.2:1-3:1).\(^1,2\) The initial presentation usually consists of disseminated cutaneous lesions with high morphological variability that can appear as purplish tumours or nodules, bruise-like spots, or, less frequently, an isolated tumour.\(^1,3\) Apart from the cutaneous clinical symptoms, these patients may present with lymph node, central nervous system, or bone marrow involvement.\(^1,2\) Leukaemic dissemination is infrequent in the initial stages of the disease, but it progresses rapidly to a fulminant leukaemia with average survival time of 8.7 months.\(^3\) Nevertheless, if the lesions are scanty and localised, with early treatment (chemotherapy and radiotherapy for isolated

![Figure 3.](image1) (a) Dense infiltrate of monomorphic blastic cells (black arrows) not affecting the Grenz zone (H&E x 20). (b) Neoplastic cells were positive for CD56 (x 20).

![Figure 4.](image2) Positive immunostaining for CD303 in neoplastic cells (x 20).

![Figure 5.](image3) Whole body CT: Lymphadenopathies suggestive of malignancy in both inguinofemoral regions.
lesions) the outcome can be favourable.\textsuperscript{3,5} Since clinically and histologically it is very difficult to distinguish from leukaemia cutis or myeloid leukaemia, the diagnosis is established by immunohistochemistry. In BPDCN, the most characteristic markers are CD4, CD56, CD123, CD303 (Figure 4), and TCL1,\textsuperscript{1,2,5} whereas the expression of myeloid markers such as CD33 favours a diagnosis of acute myeloid leukaemia. On the other hand, the absence of CD4 and EBV positive test are characteristic of an extranodal NK/T-cell lymphoma.\textsuperscript{5}

Cutaneous angiosarcoma, or malignant angioendothelioma, is a tumour originating from endothelial cells with an aggressive course that often presents as a bruise-like lesion with erythematos-purplish nodules.\textsuperscript{4} Cutaneous angiosarcoma of the head and neck in the elderly is the most common of the three types described with the other two being post-radiotherapy and secondary to chronic lymphoedema.\textsuperscript{4}

The dermatoscopic appearance of cutaneous angiosarcoma has been reported as areas of different colour varying from light red to purple with whitish-yellowish perifollicular structures.\textsuperscript{5,7}

It is worth noting the great clinical similarity of our case with scalp angiosarcoma, which can present at the same location as a purplish bruise-like formation with nodular structures. However, it also has a striking similarity on a dermatoscopic level, with areas of colouration that vary from dark red to purple with skin-coloured areas and whitish perifollicular structure formation very similar to those described in the angiosarcoma. The dermatoscopic image of the BPDCN has never been described before.

This case illustrates the great variability in clinical presentation of BPDCN, which simulated an angiosarcoma of the scalp in this case. As there are also similarities in dermatoscopic appearances between the two entities, histological study is required to establish the diagnosis.

References