中文題目：芽細胞漿細胞樣樹突細胞腫瘤 -- 一位骨髓增生性疾病患者的瘀傷樣病灶

英文題目：Blastic plasmacytoid dendritic cell neoplasm: Bruise-like lesions in a man with myeloproliferative neoplasm

作　　者：劉子瑜[1] 俞佑[1]

服務單位：[1]國泰綜合醫院皮膚科

Introduction: Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is an aggressive hematological malignancy featuring unique “bruise-like” cutaneous lesions. Involvement of bone marrow, lymph nodes, or blood may also present. Prior history of other myeloproliferative disorders could be identified in some patients, indicating possible leukemic-transformation-like events of somewhat unknown pathogenesis.

Case report: A 67-year-old man with underlying BCR-ABL1 negative unclassifiable myeloproliferative neoplasm with leukocytosis (31.63 K/uL) and thrombocytosis (1073 K/uL) took hydroxyurea for five months. He presented rapidly enlarging neck masses and “easy bruising” in the recent month. Physical examination revealed several purplish non-blanchable macules and patches on the scalp, chest, and upper back (Figure 1a & 1b). Multiple enlarged lymph nodes were also found at the neck (Figure 1c). Blood tests showed leukocytosis (52.18 K/uL) with a proportion of blasts up to 74%, anemia (6.0 g/dL), and thrombocytopenia (45 K/uL). Skin biopsy revealed dense infiltrate of monomorphic, poorly-differentiated, medium-sized tumor cells on histopathology investigation (Figure 2a & 2b). Subsequent biopsies of bone marrow and a neck lymph node all showed identical histology result. A malignant hematopoietic neoplasm was suspected. Differential diagnoses included CD56+ acute myeloid leukemia (AML), nasal-type extranodal NK/T cell lymphoma, cutaneous T cell lymphoma, and BPDCN. The immunohistochemical (IHC) stains showed positive for CD4, CD56, and CD123 (Figure 2c, 2d, 2g), and negative for MPO and EBER (Figure 2e & 2f). According to the IHC stains, CD56+ AMLs and nasal-type extranodal NK/T cell lymphoma were excluded due to negative results for MPO and EBER. Positive results for CD4, CD56, and CD123 disclosed the diagnosis of BPDCN. He was then received treatment with hyper-CVAD (cyclophosphamide, vincristine, doxorubicin, and dexamethasone). Regretfully, he was expired due to septic shock during the treatment.

Discussion: BPDCN is a rare and clinically aggressive hematologic malignancy, which arises from the precursors of myeloid-derived resting plasmacytoid dendritic cells. Clinical manifestations include cutaneous lesions (~90%), involvement of bone marrow, lymph nodes, or blood, and central nervous system (CNS) involvement (~30%).[1,2] The skin lesions

may present as isolated purplish nodules, bruise-like patches, or disseminated purplish plaques.[3] The different skin presentation is not currently a significant prognostic factor. For patients diagnosed with BPDCN, about 10% to 20 % of patients have an antecedent history of hematologic malignancies.[4] The treatment for the newly diagnosed patients is induction chemotherapy with acute lymphoblastic leukemia (ALL)-like regimen that includes CNS prophylaxis or therapy. If the patient achieved complete remission, allogeneic hematopoietic cell transplantation was suggested.[5]

Conclusion: BPDCN is a rare but aggressive hematologic malignancy, mostly manifested as cutaneous lesions. In patients with myeloproliferative disorders who develop bruise-like skin lesions, we should take BPDCN into the differential diagnosis.

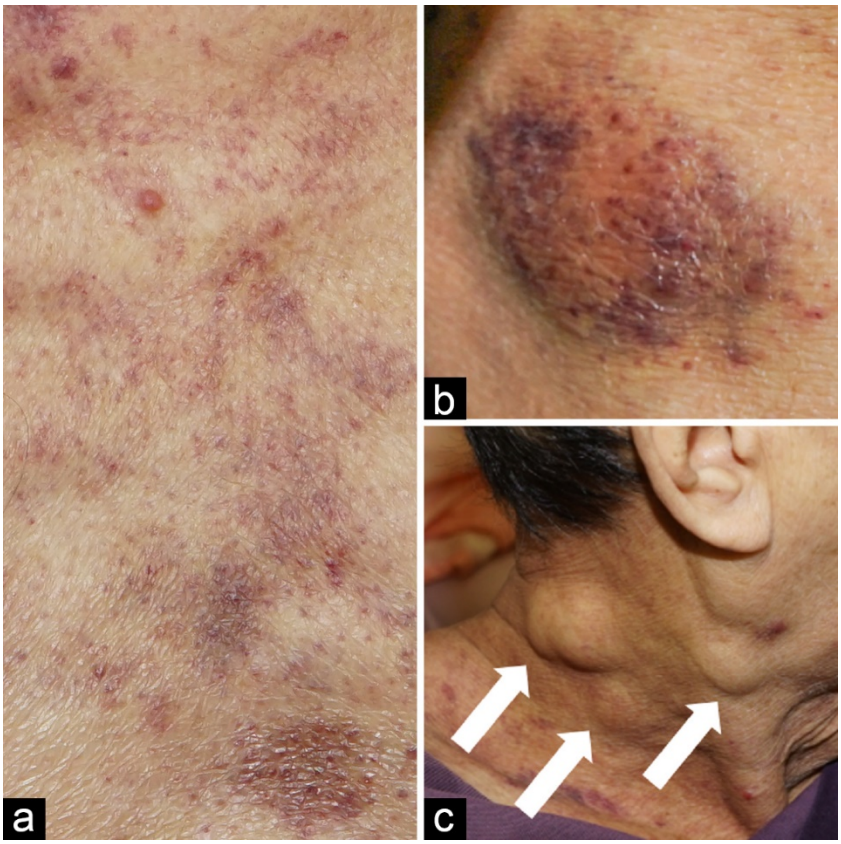

Figure 1. Clinical presentation: (a) Bruise-like macules (violaceous macules) at the chest; **(b)** a purplish nodule at the right chest; **(c)** multiple palpable neck lymph nodes (white arrows).

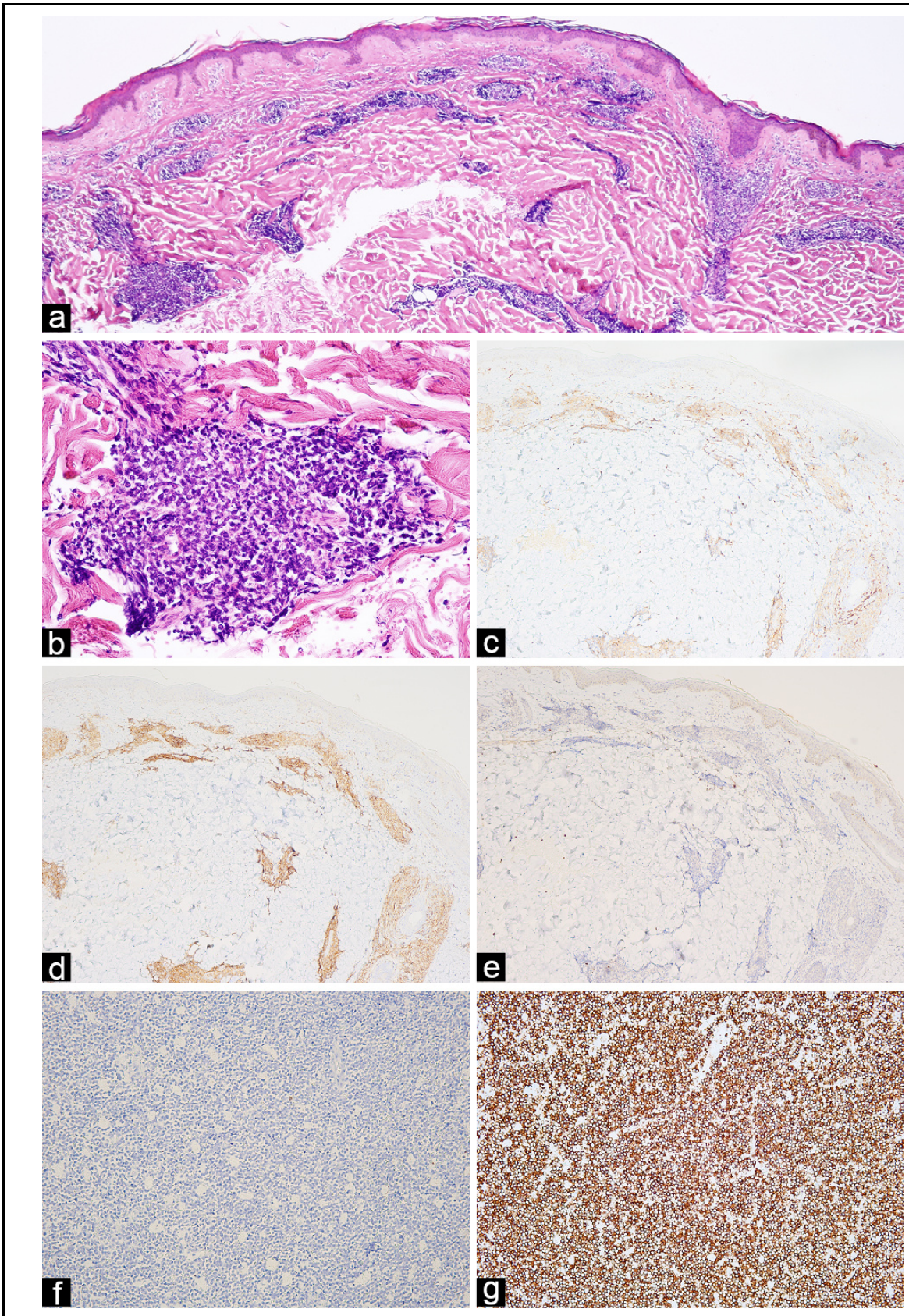

Figure 2. Histopathological investigation: (a) Histopathology stain of the skin biopsy revealed tumor cell infiltration to dermis (H and E, x100); **(b)** monomorphic, poorly-

differentiated, medium-sized tumor cells (H and E, x400); immunohistochemical (IHC) staining of skin biopsy revealed **(c)** positive for CD4 (x100), **(d)** positive for CD56 (x100), and **(e)** negative for MPO (x100); **(f)** IHC stain of neck lymph node biopsy revealed negative for EBER (x200), but **(g)** positve for CD123 (x200).

References

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