SHORT COMMUNICATION

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Primary Cutaneous Adult T-cell Leukaemia/Lymphoma Presenting as Generalized Purpura

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Adult T-cell leukaemia/lymphoma (ATLL) is a peripheral T-cell neoplasm caused by human T-cell leukaemia virus type 1 (HTLV-1). ATLL is classically divided into subtypes: acute, lymphoma, chronic and smouldering (1); however, in cases without peripheral blood, nodal or visceral involvement, the disease is classified as primary cutaneous ATLL (pcATLL) (2). Here, we introduce a patient with pcATLL presenting as generalized purpura.

CASE PRESENTATION

A 71-year-old female had initially noticed punctate purpura on her lower legs 2 years earlier. The lesions gradually increased and spread to the trunk and upper limbs. She had been treated with topical clobetasol propionate with the diagnosis of pigmented purpuric dermatosis, with only a limited response. Her medical history was unremarkable, and there was no family history. Physical examination revealed multiple palpable purpura less than 3 mm in diameter scattered on the trunk and extremities (**Fig. 1**a–c). Laboratory investigations showed a normal white blood cell count without atypical lymphocytes, lactate dehydrogenase was 190 U/L, soluble interleukin-2 receptor was high at 943 U/mL, and serum calcium was 9.4 mg/dL. Immunological findings demonstrated that the HTLV-1 antibody exceeded the upper limit of detection. Positron emission tomography-computed tomography failed to detect any organ or nodal involvement. Histopathological examination of a skin biopsy revealed infiltration of medium-sized atypical lymphocytes with extravasation of red blood cells in the superficial dermis (Fig. 1d). Immunohistochemistry showed that the infiltrating cells were positive for CD8 and CD25 (Fig. 1e and f) with loss of CD7; CD4 and CD20 were negative, and granzyme B was not overexpressed. Southern blot analysis confirmed monoclonal integration of the HTLV-1 provirus in the skin sample. The diagnosis of pcATLL was made. Narrowband ultraviolet B phototherapy was initiated with topical corticosteroids, leading to a partial improvement.

DISCUSSION

Skin manifestations of ATLL are categorized into patch, plaque, multipapular, tumoral, erythrodermic and purpuric types (3). Among these, the purpuric type is relatively rare, and only 5 cases have been reported in the English literature to date (**Table I**). The purpuric type is further divided into 2 subtypes: punctate and macular (3). Our case presented numerous purplish-red papules and therefore corresponds to the purpuric punctate subtype. To the best of our knowledge, this is the first reported case of purpuric punctate pcATLL occurring in the absence of blood, nodal or visceral involvement, thereby expanding the clinicopathological spectrum of ATLL (Table I) (4–8).

Pathophysiology of purpura development in ATLL has been attributed to tumour cell-mediated vascular injury rather than thrombocytopenia or leukocytoclastic vasculitis (8). The expression of CCR4 by ATLL cells and the presence of its ligand CCL17 in dermal blood vessels suggest that the CCL17/CCR4 axis likely facilitates skin homing of ATLL tumour cells to the skin (9). Granzyme B produced by ATLL tumour cells may damage blood vessels, leading to erythrocyte extravasation around tumour infiltrates on histopathology (8). However, this scenario is inconsistent with our case, in which no granzyme B overexpression was observed.

The cutaneous subtype is an independent prognostic indicator in ATLL (3). The punctate purpuric subtype has a better prognosis than the macular purpuric subtype (mean survival 73.4 months vs 2.1 months; Table I) (3), which is consistent with the clinical course of our patient. CD8 expression, as seen in our case and generally

Table I. Previous reports of adult T-cell leukaemia (ATLL)/lymphoma with purpuric lesions.

Reference	Sex	Age	ATLL subtype	Organ involved	Prognosis	Punctate or macular
Present case	F	71	Primary cutaneous	None	Indolent at 24 months follow-up	Punctate
(4)	M	70	Acute	Blood, bone marrow, stomach, central nervous system	Died 5 months after diagnosis	Punctate
(5)	M	87	Acute	Peripheral blood, inguinal lymph nodes	Died ≈2.5 months from onset of purpura	Macular
(6)	F	62	Acute	Peripheral blood, bone marrow	Complete remission	Punctate
(7)	M	76	Lymphoma	Lymph nodes, stomach	Died 9 months after first admission	Punctate
(8)	F	78	Acute	Peripheral blood, bone marrow	Died 3 months after onset of purpura	Macular

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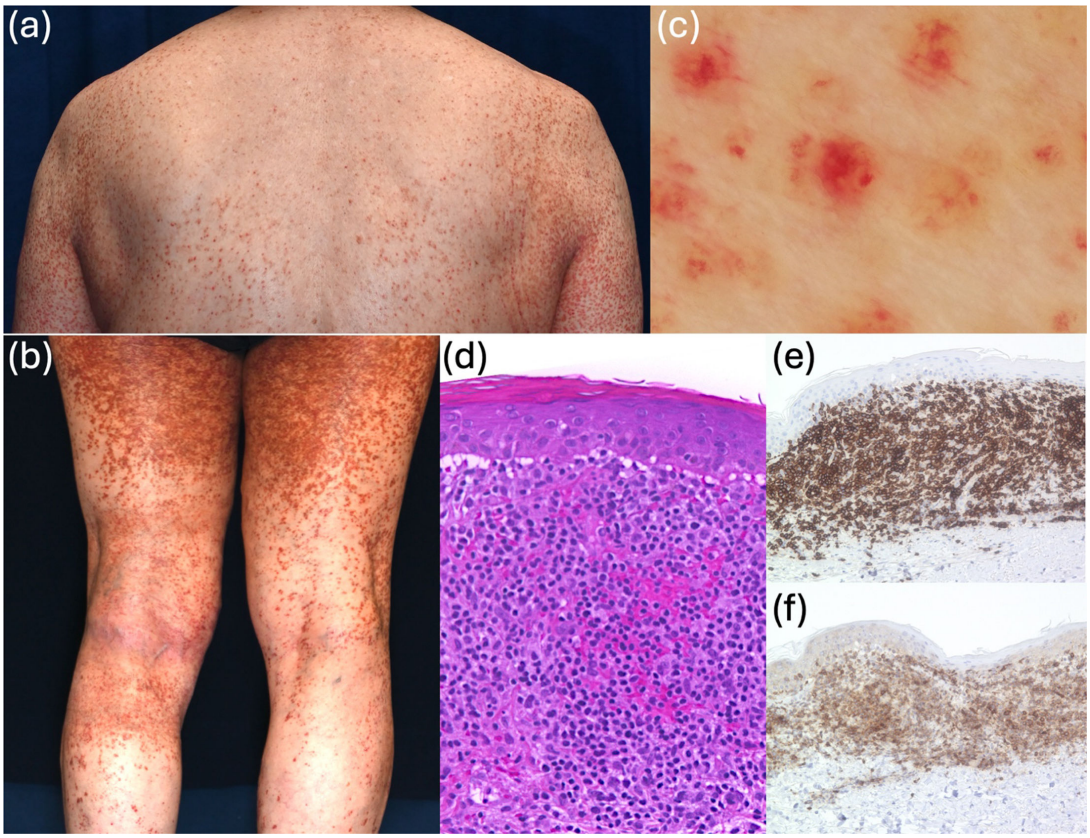

Fig. 1. Clinical and histopathological findings. (a, b) Disseminated punctate purpura on the back (a) and proximal thigh (b). (c) Multiple purplish-red areas with indistinct borders on polarized dermoscopy. (d) Infiltration of medium-sized atypical lymphocytes with erythrocyte extravasation in the superficial dermis (haematoxylin-eosin stain, ×20). (e, f) Immunohistochemistry showing CD8 (e) and CD25 (f) expression in the atypical lymphocytes (×20).

considered a cytotoxic phenotype, is relatively common in ATLL and is not associated with poor prognosis (10). Our case illustrates that ATLL should be included in the differential diagnosis of generalized purpura.

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Data availability statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

The authors have no conflicts of interest to declare.

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