

SHORT COMMUNICATION

Disseminated Primary Cutaneous CD8⁺ Small/Medium-sized Pleomorphic T-cell Lymphoma Responding to HydroxychloroquineValerie C. Amann^{1#}, Jil Dreier^{1#}, Desislava Ignatova¹, Jivko Kamarashev¹, Katrin Kerl¹, Werner Kempf², Steve Pascolo¹, Reinhard Dummer¹, Antonio Cozzio¹ and Emmanuella Guenova^{1*}¹Department of Dermatology, University Hospital Zurich, Gloriastr. 31, CH-8091 Zurich, and ²Kempf und Pfaltz, Histologische Diagnostik, Zürich, Switzerland. *E-mail: emmanuella.guenova@usz.ch[#]These authors contributed equally to this work and should be considered as first authors.

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Primary cutaneous CD8⁺ small/medium-sized pleomorphic T-cell lymphoma (CD8⁺ SMP TL) is a rare entity with most probably indolent clinical behaviour. Several cases with facial localisation, mostly localised to the ear, have previously been described (1). Reports of extrafacial involvement are limited, and mostly involve acral sites such as the hands and feet. The classification concerning this rare cutaneous lymphoma remains to be defined. To date, no treatment modalities have been systematically evaluated.

CASE REPORT

We report a case of a 54-year-old man with disseminated CD8⁺ SMP TL and excellent treatment response to hydroxychloroquine. The disease initially presented in 2010 with several erythematous papules and small nodules localised on the left foot which were treated successfully with low dose soft X-ray irradiation (32 Gy using 100 kV) (2). Staging was negative for extracutaneous involvement. About 5 months after the initial presentation, the disease progressed slowly with

the development of multiple brownish-reddish nodules disseminated on the whole body surface area, most accentuated on the trunk. Routine PCR diagnostic procedure revealed monoclonal rearrangement of the T-cell receptor, and immunohistochemical analysis revealed positivity for CD8 and TIA1 (Fig. 1). Interestingly, the bystander inflammatory infiltrate was dominated by CD68⁺ histiocytes, which accounted for an unusual granulomatous-like histological appearance of the lesions (Fig. 1). This fact, as well as 3 relapses after low-dose X-ray radiotherapy and intralesional steroid application, motivated us for a therapeutic approach with low dose systemic corticosteroids (7.5 mg prednisolone daily) and hydroxychloroquine (400 mg/day), which resulted in immediate clinical improvement of the lesions (Fig. 2).

DISCUSSION

The patient's history was positive for X-linked agammaglobulinemia (XLA; i.e. Bruton agammaglobulinemia), a primary inherited immunodeficiency syndrome

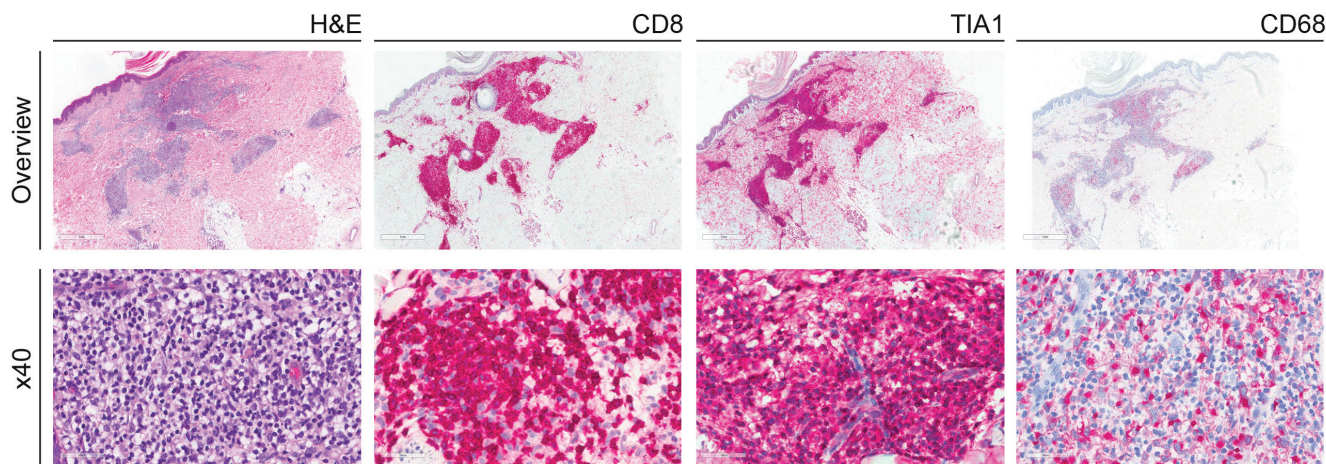


Fig. 1. Primary cutaneous CD8⁺ small/medium-sized pleomorphic T-cell lymphoma.

A well demarcated, non epidermotropic infiltrate composed of small- to medium-sized pleomorphic lymphocytes. The neoplastic cells coexpress CD8 and the cytotoxic granule-associated RNA binding protein TIA1. Note the marked predominance of CD68 positive histiocytes in the infiltrate. H&E: hematoxylin and eosin staining; CD: cluster of differentiation.

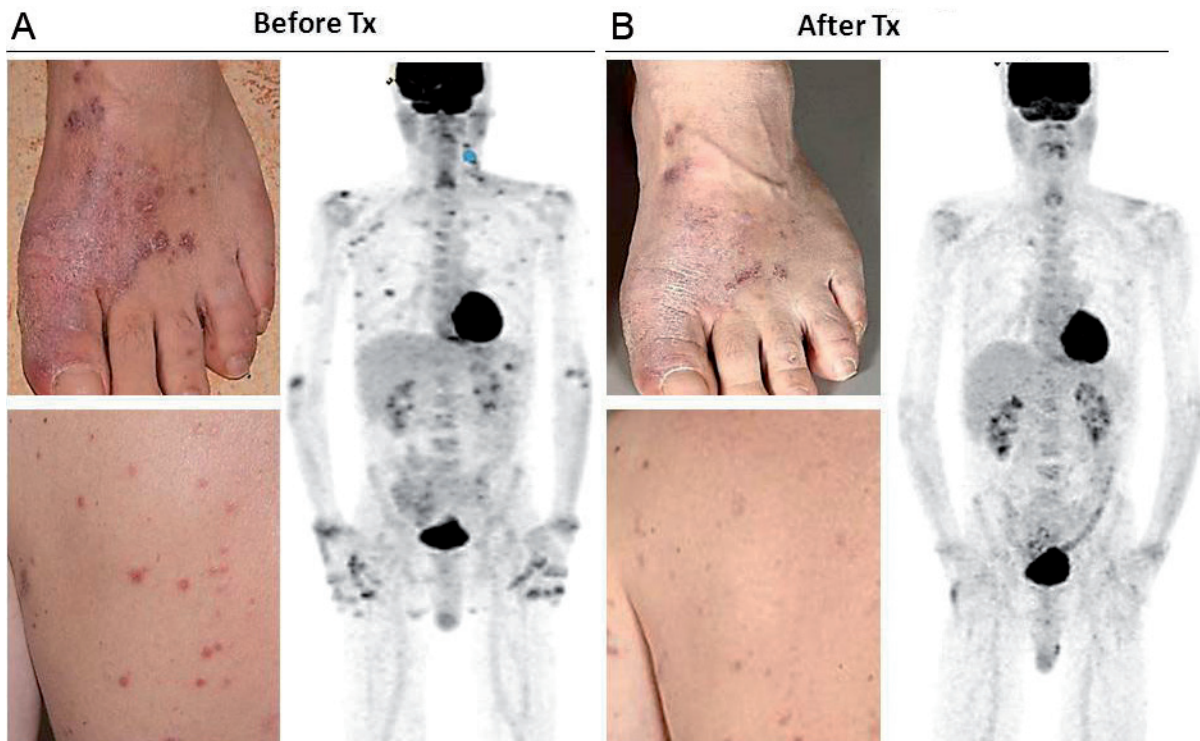


Fig. 2. Clinical and positron emission tomography findings before start of treatment (A) and after 9 months of treatment with low dose systemic corticosteroids and hydroxychloroquine (Tx) (B).

with insufficiency of B-cell antibody production caused by mutations in the gene coding for Bruton tyrosine kinase (BTK). He had been on intravenous immunoglobulin replacement since XLA was first diagnosed at age 17, and had a history of chronic sinusitis as well as chronic obstructive pneumopathy. Furthermore, he was on low dose methotrexate for treatment of seronegative polyarthritis.

In general, patients with antibody deficiencies seem at higher risk to develop haematological malignancies, including cutaneous lymphoma. However, these malignancies are predominantly of B-cell origin (3). The correlation of primary cutaneous peripheral T-cell lymphoma, especially CD8⁺ SMPTL, and XLA is still elusive. One recent case report from Park et al. (4) describes a primary cutaneous T-cell lymphoma, not otherwise specified in a patient with XLA. In 2001, Kanavaros et al. (5) reported of an extranodal cytotoxic T-cell lymphoma in another patient with XLA. There are no previously documented cases of CD8⁺ SMPTL in XLA. Assumptions of pathogenesis include, but are not limited to, antigen overstimulation through chronic infections in an immunodeficient setting. This may explain, to a certain extent, the granulomatous type of the bystander tumour inflammation, as seen in our patient.

There is no standard treatment for CD8⁺ SMPTL, and current treatment options are limited. Hydroxychloroquine is an unusual treatment for cutaneous lymphoma, which proved extremely effective in this patient.

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The authors declare no conflict of interest.

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