Intravascular relapse of an extra-nodal NK/T-cell lymphoma, nasal-type, presenting as diffuse and eruptive telangiectasia

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Introduction: **Intravascular (IV) lymphomas** are characterized by the presence of **neoplastic lymphocytes** (mostly of the B-cell lineage) **restricted to the lumen of small vessels**. We report an **exceptional case of IV NK/T-cell lymphoma** (IVNKTL) occurring in a patient with **a history of extranodal NK/T-cell lymphoma, nasal-type** (ENKTL).

Case report: A 64-year-old woman consulted for a recent history of asymptomatic and diffuse eruption, without B symptoms. **Four years ago**, **she was treated** for a **stage I nasal ENKTL with an ongoing complete response**. Clinical examination showed **diffuse telangiectasia of the trunk** with **erythematous and telangiectatic patches/plaques of the trunk and limbs** (figure 1). **Epstein-Barr virus** (EBV) **viremia was present** (5 log copies/ml). The histological examination of a pre-thoracic skin biopsy showed a **dermal IV proliferation of large cells** with a **CD2+ CD3+ CD4+ CD56+/- Granzyme B+ phenotype. EBER in situ hybridization was positive,** reflecting EBV infection of the neoplastic cells (figure 2). **These findings supported the diagnosis of IVNKTL.** Staging investigations revealed bone involvement **without nasal lesions.** A treatment with **multiple drug chemotherapy** was started. **However she died 1 month after.**

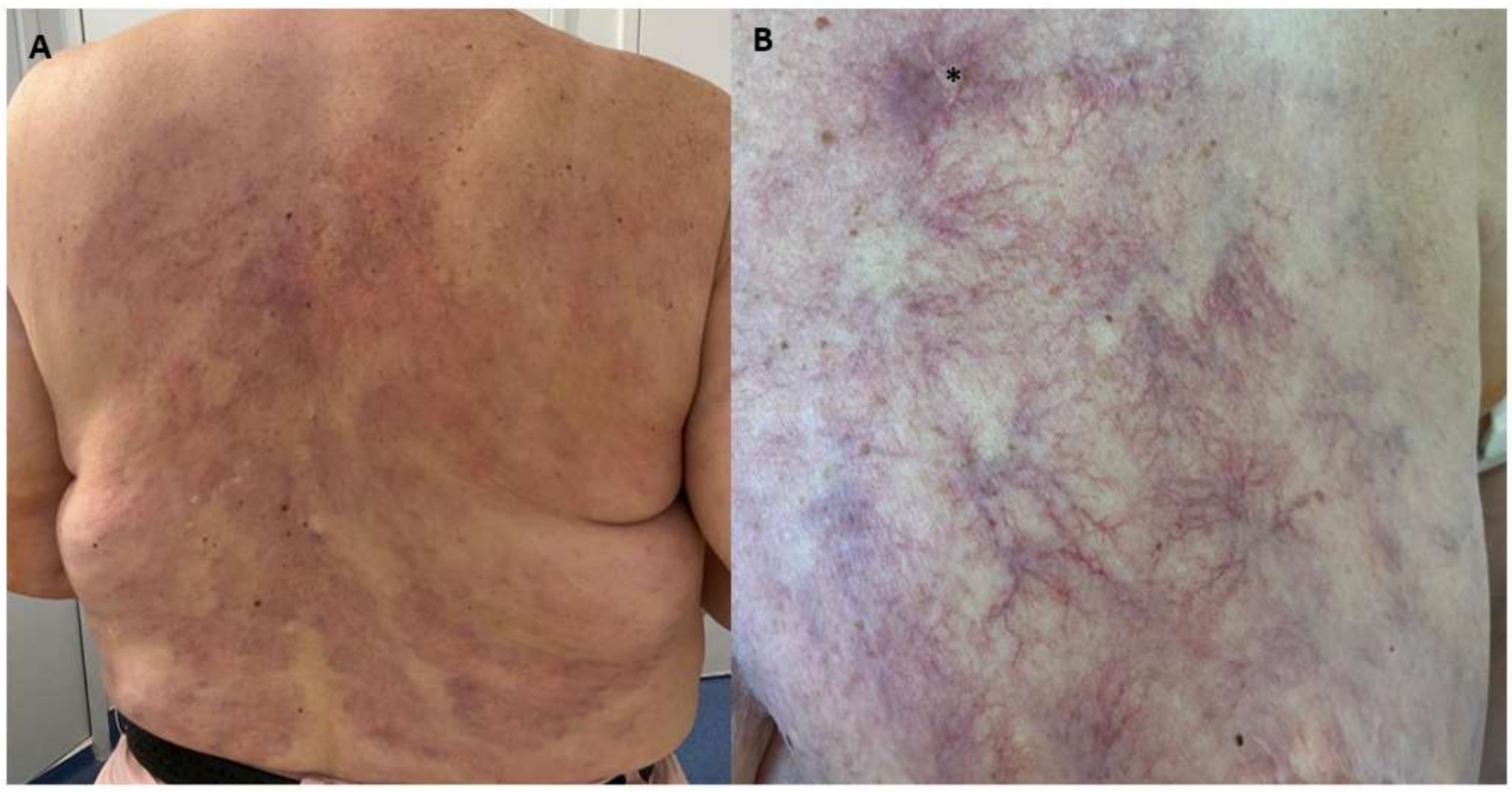

Figure 1. Clinical presentation. Diffuse telangiectasia distributed on the back (A), and the chest (B). Area of the skin biopsy (*)

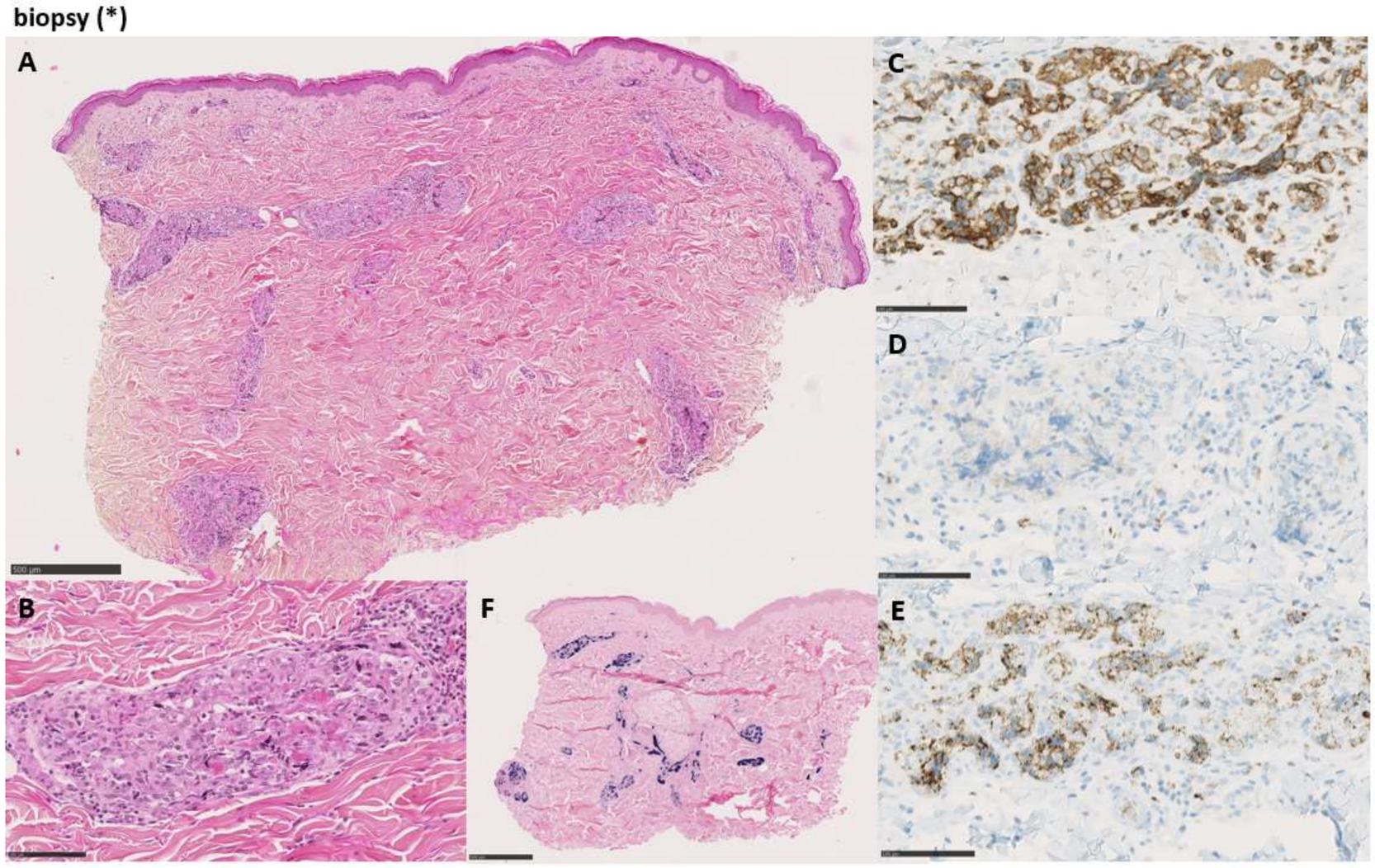

Figure 2. Histological (A et B) and immunohistochemical (C : CD2 ; D : CD56 ; E : Granzyme B) faetures ; positivity of EBER in situ hybridation (F)

Discussion: Acquired, diffuse, eruptive and non-ascending telangiectasia can reveal viral infection, mastocytosis, monoclonal gammopathy (POEMS, TEMPI syndromes), IV B-cell lymphoma.
We reported an original case of diffuse telangiectasia leading to the diagnosis of IVNKTL.

To date, **36 IVNKTL** have been reported (24 men, mean age of 50 years):

- Similarly to ENKTL, the diagnosis of IVNKTL is based on **(1) a neoplastic cell proliferation, (2) of NK/T-cell phenotype (CD2+ CD56+)**, **(3) expressing cytotoxic proteins (TIA-1, granzyme B, perforine)** with **(4) EBV infection**. However, this proliferation is **confined to the lumina of vessels in IVNKTL whereas it takes place in the extravascular tissue in case of ENKTL**,
- IVNKTL involved mostly **the skin (26/36)**, followed by the central nervous system (8/36), lung (4/36), liver and bone (2/36), heart, spleen, testicles, ileum and kidney (1/36),
- The skin involvement presented as **erythematous patches/plaques (n=26)** sometimes **hyperpigmented (n=4)** or **purpuric (n=2)**; **nodules (n=4)** or **ulceration (n=1). The lesions were mostly multiple (n=24) and were strictly localized on the trunk (n=7), lower limbs (n=10) or were diffuse (n=9). Presence of telangiectasia on the plaques was noted in 2 cases,**
- The prognosis was **poor** with a **1-year survival rate of 32%.**

Only **2/ 36 IVNKTL** were associated with ENKTL. **We reported the third case**. The occurrence of these **two rare** and **closely related entities** in the same patient **suggests a common origin** (IV relapse of the ENTKL). However, **in the absence of available clonality analysis, the hypothesis of 2 different EBV-related lymphoproliferations remains possible.**