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Epstein-Barr virus(EBV)와 연관된 Nasal type T/Natural Killer(NK)-cell lymphoma 1예

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=Abstract=

Nasal type T/Natural Killer(NK)-cell lymphoma associated with Epstein-Barr virus(EBV)

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Nasal type T/NK-cell lymphoma는 병리조직학적으로 응고성 괴사가 동반된 혈관중심성 침윤을 특징으로하며 T-cell과 NK-cell 항원을 모두 표현하는 면역조직화학염색 소견을 보이는 매우 드문 피부 림프종으로서 EBV와의 연관성이 흔히 보고되고있다. 저자들은 42세 여자에서 발생한, 전신적인 홍반성 구진, 판, 결절의 임상양상을 보였으며 피부 병변에서의 EBV-encoded RNA(EBER)에 대한 in situ hybridization상 양성소견을 보였고 전산화단층촬영, 골주사, 갈륨주사 등의 검사상 비강, 위장관 및 다른 내부장기 침범소견은 보이지 않아서 피부에만 국한된 경우로 사료되는 nasal type T/NK-cell lymphoma 1예를 보고하는 바이다.

Key Words : T/NK-cell Lymphoma, EBV

Introduction

A designation of T/NK-cell lymphoma was recently adopted, emphasizing the expression of antigens associated with both of T-and NK-cell types[1]. Because the nasal area is the most common site of presentation, "nasal T/NK-cell lymphoma" was favored as the primary term of choice for midline facial lesions.[1,2] Tumors with an identical morphology, phenotype, and genotype but presenting in another anatomic site would be considered as nasal type T/NK-cell lymphomas.[1,3] The following case describes a cutaneous T/NK-cell lymphoma associated with Epstein-Barr virus (EBV).

Report of a case

A 42-year-old female had asymptomatic papules, plaques, and nodules on her face, trunk and extremities for 8 months. Examination revealed multiple, erythematous, purpuric, crusted papules and indurated plaques on her extremities (Fig. 1). Physical examination was normal except the skin lesions. The family history, the past medical history and the review of systems were all unremarkable. The results of the following studies were within normal limits or negative: complete blood cell count, urinalysis, liver function test. Roentgenogram of the chest was also normal. Histo-pathologic examination of a plaque from the left lower leg showed infiltrating cells involving mainly deeper dermis and subcutaneous tissue. Examination of the reticular dermis revealed polymorphous angiocentric lymphohistiocytic infiltrate (Fig. 2). Atypical lymphoid cells, histiocytes and some plasma cells were visible around blood vessels, and necrosis was observed. The immunohistochemical studies demonstrated positive labeling of the infiltrating lymphoid cells for CD2, cytoplasmic CD3, CD45RO(UCHL-1), and CD56 whereas CD20(L-26) and CD30(Ki-1) showed no immuno-reactivity. But the infiltrates were lack of clonal rearrangements of the TCR genes. Gallium scan, bone scan and chest-abdomen-pelvis CT revealed no evidence of internal organ involvement. These clinical and histopathologic findings were consistent with a diagnosis of cutaneous T/NK-cell lymphoma and EBV studies were done in serum and lesional tissue. Antibodies to viral capsid antigen (VCA), early antigen (EA) and Epstein-Barr nuclear antigen (EBNA) were all positive except IgM-anti-VCA in serologic studies and the titers

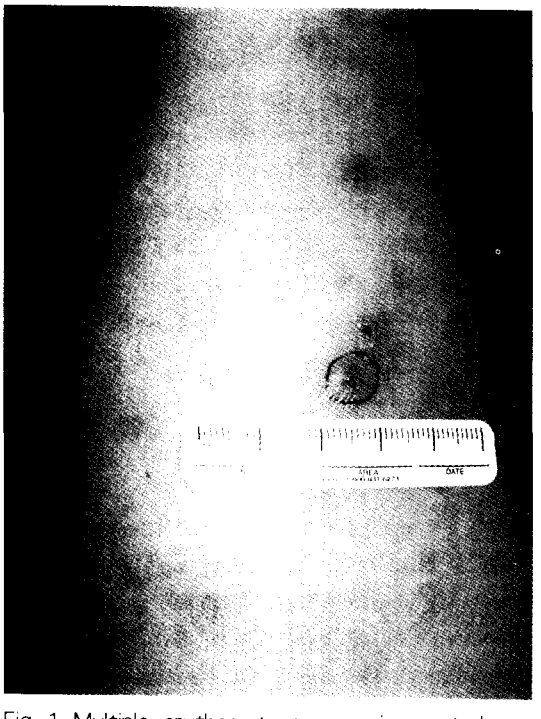

Fig. 1. Multiple, erythematous, purpuric, crusted papules and indurated plaques on the extensor surface of the lower leg

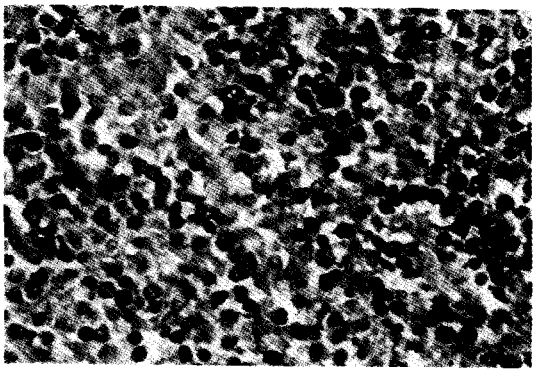

Fig. 2. Biopsy specimen showing polymorphous angiocentric infiltrate containing atypical lymphoid cells (H & E ×200).

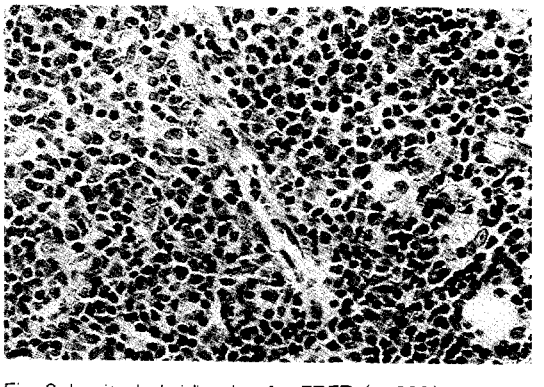

Fig. 3. In situ hybridization for EBER (×200).

were regarded as significantly elevated based on background data in general population. In addition, EBV encoded RNA(EBER) was demonstrated in lesional skin by in situ hybridization (Fig. 3). Treatment was started with oral cyclophosphamide, 100 mg daily and prednisone, 50 mg daily. Four weeks later, there was some improvement of the skin lesions.

Discussion

Nasal type T/NK-cell lymphoma is a rare subtype of cutaneous lymphoma characterized by angiocentricity with coagulative necrosis.[1] The most common immunophenotype is CD56+, CD2+, cytoplasmic CD3+ but the infiltrates are negative for surface CD3 and lack of clonal rearrangements of the TCR genes.[1-3] In addition, EBV-encoded RNA (EBER) is identified in virtually all of the cases by in situ hybridization.[1] Therefore, it has many similarities with classical lymphomatoid granulomatosis[4-9] which was recently hypothesized by some authors[10] as a proliferation of EBV infected B-cells with a prominent T-cell reaction(T-cell rich B-cell lymphoma). Clinically, our case presented with multiple, erythematous, purpuric, crusted papules and indurated plaques, not showing manifestations of a hemophagocytic syndrome which is common to T/NK-cell lymphoma[1,11], but showed relatively good response to systemic steroid therapy. More observations are required to evaluate the prognosis.

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