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Cutaneous Angiocentric T-cell Lymphoma

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=국문초록=

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지 두 현 · 장 경 애 · 장 성 은 · 최 지 호 · 고 재 경

Angiocentric T-cell lymphoma는 혈관중심성의 부정형 림프구 침윤을 특징으로하는 매우 드문 피부 T세포 림프종의 한 아형으로서 전세계적으로 대만, 일본등 동아시아 국가에서 주로 보고되고 있다. 저자들은 62세 여자에서 발생한, 국소적으로 비교적 경계가 명확한 홍반성 판의 임상양상을 보였으며 전신 증상은 동반되지 않았고 전산화단층촬영, 골주사, 갈륨주사 등의 검사상 내부장기 침범소견은 보이지 않아서 피부에만 국한된 경우로 사료되는 Angiocentric T-cell lymphoma 1예를 보고하는 바이다.

Key Words : Angiocentric T-cell Lymphoma

Introduction

Angiocentric T-cell lymphoma(AL) is a rare subtype of cutaneous T-cell lymphoma characterized by angiocentric, angioinvasive, and angiodestructive infiltrate containing atypical lymphocytes[1-2]. Recently many cases have been reported from Taiwan and Japan in association with Epstein-Barr virus(EBV)[3-6], but limited data are available in Korea. We report a case of AL confined to the skin.

Report of a case

A 62-year-old female had asymptomatic plaques on her flank for 6 months. Examination revealed localized, relatively well-circumscribed, erythematous indurated plaques on the right flank area (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. A biopsy specimen showed angiocentric and angioinvasive infiltrate in the dermis (Fig. 2). Lymphocytes containing atypical lymphoid cells were visible around blood vessels, and angio-invasion was also observed (Fig. 3). But epidermotropism, hemophagocytosis and necrosis were absent in our case. The immunohistochemical studies demonstrated positive labeling of the infiltrating lymphoid cells for CD45RO(UCHL-1) whereas CD20(L-26), CD30(Ki-1) and CD56 showed no immunoreactivity. 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 AL and treatment was started with oral prednisone. 4 weeks later, there was much improvement of the skin lesions.

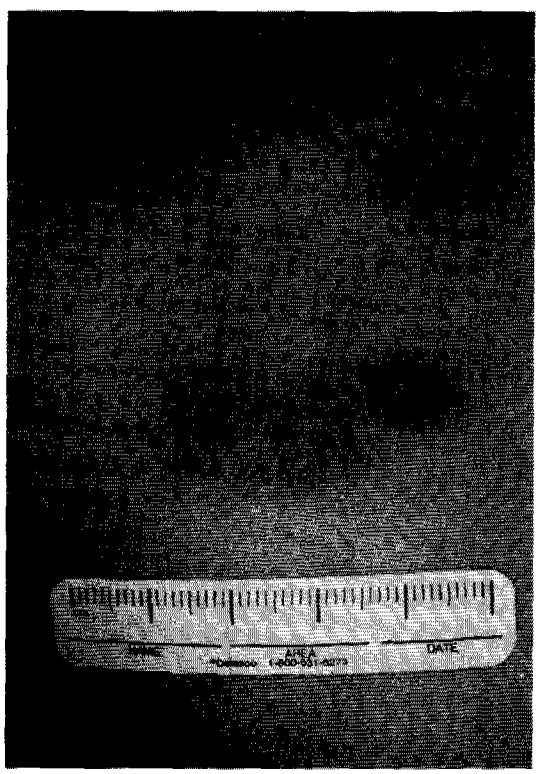

Fig. 1. Localized, relatively well-circumscribed, erythematous indurated plaques on the right flank area

Fig. 2. Low-power view demonstrates angio-centric and angioinvasive infiltrate in the dermis (H & E × 40).

Discussion

Angiocentric T-cell lymphoma is a rare subtype of cutaneous T-cell lymphoma characterized by angiocentric, angioinvasive, and angiodestructive infiltrate containing atypical lymphocytes[1-2]. The term "angiocentric immunoproliferative lesion(AIL)" was introduced by Jaffe et al[1]. to replace lymphomatoid granulomatosis and related disorders, including lymphocytic vasculitis and lethal midline granuloma and they graded cases into three histologic categories(grade Ⅰ-Ⅲ). GradeⅢ lesions represent overt malignant lymphoma, also termed "angiocentric lymphoma." But some authors[7] hypothesized that most cases of lymphomatoid granul-

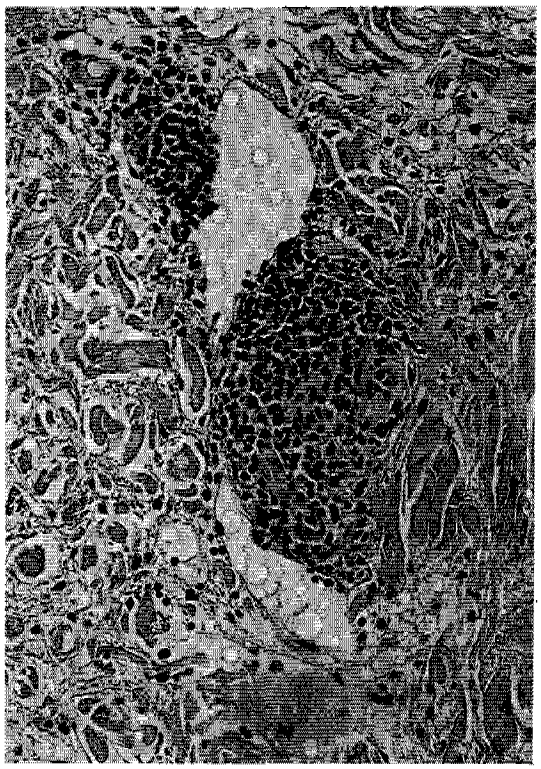

Fig. 3. Higher magnification showing angioinvasive lymphocytic infiltrate containing atypical lymphoid cells (H & E × 200).

omatosis involving the lung represent a proliferation of EBV infected B-cells with a prominent T-cell reaction(T-cell rich B-cell lympho-ma), distinguishing these cases from angiocentric T-cell lymphomas. Recently a designation of T/NK-cell lymphoma was adopted, emphasizing the expression of antigens associated with both of these cell types[8]. Because the nasal area is the most common site of presentation, nasal T/NK-cell lymphoma was recommended for midline facial lesions[8-9], and identical tumors presenting in another anatomic site would be considered nasal type T/NK-cell lymphoma[8,10]. The common histo-pathologic characteristic of this T/NK cell lymphoma is angiocentricity with coagulative necrosis, and the most common immunophenotype is CD56+, CD2+, cytoplasmic CD3+[8-10]. The infiltrates are negative for surface CD3 and lack of clonal rearrangements of the TCR genes[8]. Finally, EBV-encoded RNA (EBER) is identified in virtually 100% of cases by in situ hybridization. Our case differed from nasal type T/NK cell lymphoma in several aspects. Histologically, it did not show coagulative necrosis seen in nearly 100% of cases of T/NK cell lymphoma and lacked CD56 immunoreactivity, although angiocentricity is common to both conditions. Clinically, our case presented with localized, relatively well-circumscribed, erythematous indurated plaques, not showing manifestations of a hemophagocytic syndrome which is common to T/NK-cell lymphoma[8,11], but showed relatively good response to systemic steroid therapy. Therefore we concluded that this was angiocentric T-cell lymphoma confined to the skin, although we did not perform clonal rearrangements of the TCR genes and in situ hybridization for EBER.

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