

혈액식작용 증후군을 동반한 구인두의 T/NK세포 림프종

가
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A Case of Oropharyngeal T/NK-Cell Lymphoma with Hemophagocytic Syndrome

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- ABSTRACT -

T/NK-cell lymphoma (TNKL) is a subtype of angiocentric T-cell lymphoma showing the characteristics of biphenotype of T-cell and NK-cell, frequent association with Epstein-Barr virus (EBV), and poor clinical course. It usually develops in nasal cavity, paranasal sinus, and nasopharynx. Hemophagocytotic syndrome is a histiocytic proliferation associated with phagocytosis of the hemopoietic elements resulting in fever, profound pancytopenia, hepatosplenomegaly, lymphadenopathy, and coagulopathy. It is considered as a negative prognostic factor for hematologic neoplasms. Epstein-Barr virus infection is considered as pathogenic factor for hemophagocytotic syndrome. We experienced a case of EBV-infected oropharyngeal lymphoma of T/NK-cell lymphoma associated with hemophagocytotic syndrome, which was confirmed by immunohistochemical study, in situ hybridization, and bone marrow biopsy. We report this case with brief review of literatures. (**J Clinical Otolaryngol 2001;12:83-88**)

KEY WORDS : Lymphoma · T-cell lymphoma · EB virus infection · Hemophagocytic syndrome.

서 론 (angiocentric immunoprolifera-
tive lesion, AIL) 3 T -
(angiocentric T - cell lymphoma, ATL)
T/NK - (T/NK - cell lymphoma, TNKL) T - NK -
T - (peripheral . TNKL
T - cell lymphoma, PTL) , ,
TNKL(nasal TNKL)
: 2000 12 8 , TNKL(nasal type
: 2001 5 27 ,
: , 403 - 720 6 665 TNKL) .¹⁾
가 (Hemophagocytotic syndrome,
: (032) 510 - 5526 . : (032) 510 - 5821
E - mail : parkent@olmh.cuk.ac.kr HS) - - (monocyte - macr -

phage - histiocyte) (he - 10 13 .
 mophagocytic cell) 가 2
 39.5
 , TNKL
 Epstein - Barr virus 가
 (EBV)가 (2-4) (Fig. 1)
 , (aryepig -
 lottic fold), (arytenoid process)가
 T/NK -
 가 Wegener ,
 EBV X -
 T/NK - 1 (Wa -
 ters and Caldwell view)
 증 례
 37 가 1998 9 8 2 가
 1 EBV IgM IgG
 , C - ANCA (cytoplasmic antineutr -
 ophilic cytoplasmic antibody)

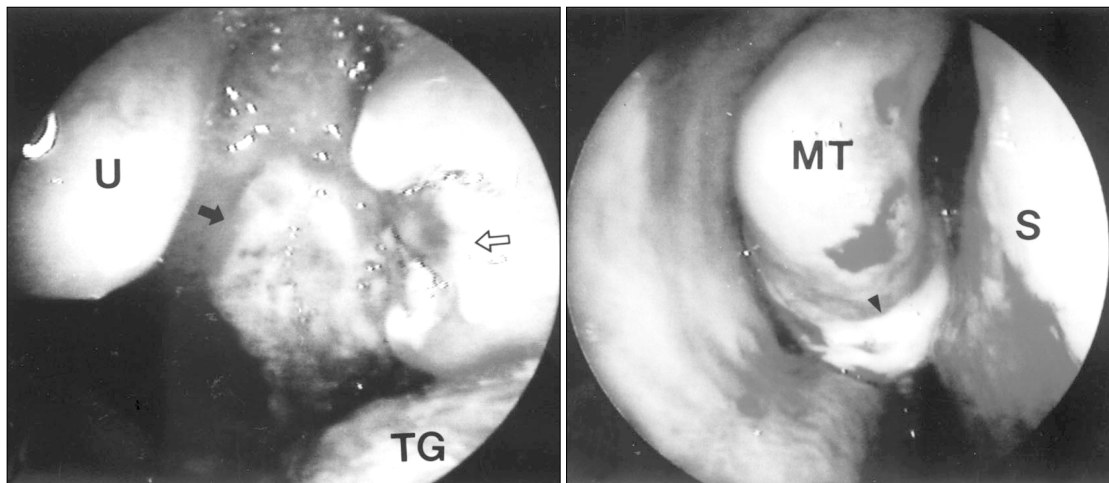


Fig. 1. Initial photograph of oropharynx and nasal cavity. A : Deep ulcerative lesion in the posterior wall of oropharynx (black arrow) and left tonsil (white arrow) (U : uvula, TG : tongue). B : Posterior end of right middle turbinate was covered with whitish patch (arrowhead). Removing this patch revealed erosive, friable middle turbinate (MT : middle turbinate, S : septum).

: Hemophagocytosis T/NK

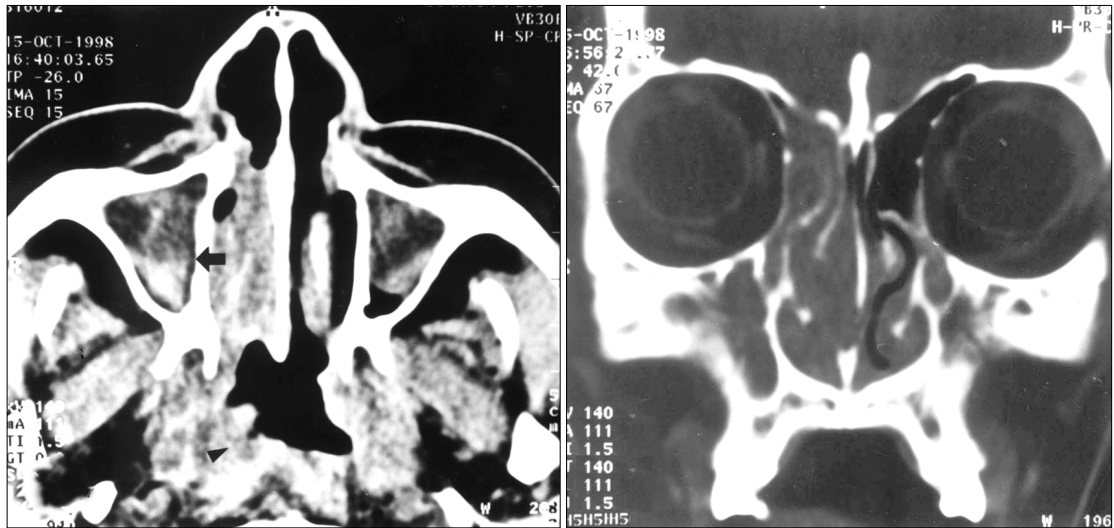


Fig. 2. PNS computerized tomography (CT). A : Axial view through the Rosenmeller fossa showed soft tissue densities in both maxillary sinuses and heterogeneity especially in right maxillary sinus (arrow). Right nasal cavity was filled with soft tissue density which was extending into the nasopharynx and obliterating Rosenmeller fossa (arrowhead). B : In coronal view, whole nasal cavity and sinuses of right side were filled with soft tissue density.

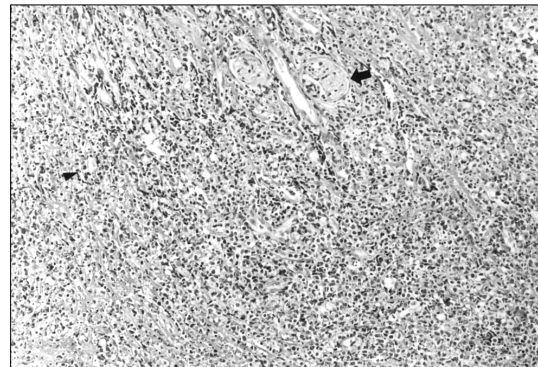


Fig. 3. Photomicrograph of oropharyngeal lesion. Atypical lymphoid cells infiltrated around vessel walls (arrowhead) and perineural spaces (arrow) (H-E stain, $\times 100$).

pr -
ednisolone 60 mg , 11 5 6
EBV T/NK -
(T/NK - cell lymphoma)

(Fig. 3). UCHL1 (pan T - cell marker) NK
CD56 (in situ hybridization) EBV EBV - encoded RNA (EBER)
(Fig. 4).

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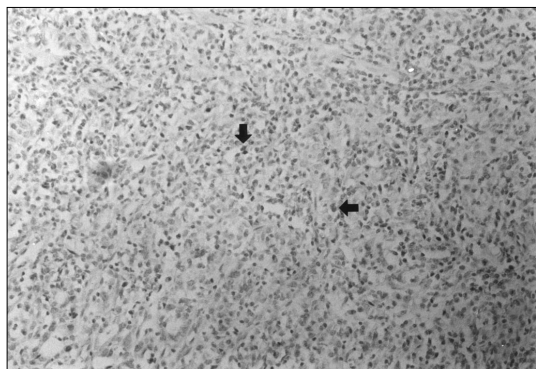


Fig. 4. Photomicrograph of oropharyngeal lesion via in situ hybridization for EBERs (x 100). Positive high signals in the nuclei of lymphoid tumor cells (arrow) are noted.

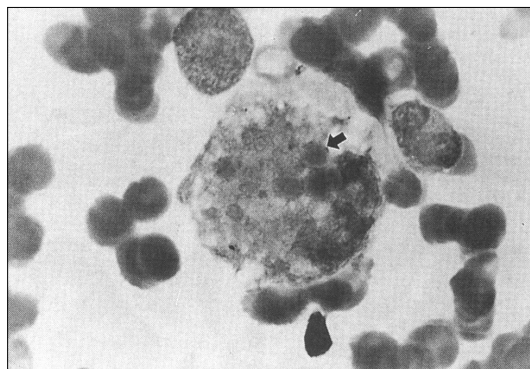


Fig. 5. Oil power examination of bone marrow smear. Macrophage engulfs some erythrocytes (arrow) and necrotic nuclear debris (Wright stain, x 1000).

(hemophagocytosis)
 11 11 5 cyclophosphamide,
 vincristine, prednisolone(CVP)
 (Fig. 5). 11 25 5 cycl -
 ophosphamide, adriamycin, vincristine, prednisolone
 (CHOP) 12 3

고 찰

T - (peripheral T - cell lymphoma,
 PTL) T -
 (post - thymic lymphoma)
 가 T -
 (thymic lymphoma) T -
 5)
 (A revised European - Am -
 erican classification of lymphoid neoplasms, REAL)
 PTL 15% 가
 가
 (angiocentric lymphoma)
 6)
 T/NK -

T - NK - (biphenotype)
 TNKL
 가 1) TNKL
 NK -
 CD56 NK -
 CD16 CD57 T -
 CD2 CD3
 CD3 F1, TCR 1 T -
 7)8)
 가 NK - T - , T -
 NK - , 3
 (indeterminate large granular
 1)9)
 lymphocyte) Burkitt
 EBV B -
 PTL 10)
 AIL, EBV가
 11) AIL (transfor - ma -
 tion) TNKL EBV가 7)
 EBV 가

: Hemophagocytosis T/NK

²⁾¹⁶⁾

EBER
EBV PTL
가
가
cyclophosphamide가
CHOP(cyclophosphamide, adriamycin, vincristine, prednisolone), BACOP(bleomycin, adriamycin, cyclophosphamide, vincristine, prednisolone), C-MOPP(cyclophosphamide, vincristine, procarbazine, prednisolone)

³⁾
TNKL
(nasal)
(nasal type) TNKL
9) TNKL
2
TNKL (mi-
dline facial destructive disease)
가
가 가
가
Kim ¹²⁾
AIL TNKL
Ko ¹³⁾ CD56
EBV
Park ¹⁴⁾ EBV 21
(76%), T
(76%)
1998 Ko ¹⁵⁾ 1994 1996
1548 REAL classification
T- 7 48
129 (8.7%) , 74.1%,
가 13.8%, Waldeyer (Waldeyer 's tonsillar ring) 11.2%
(sarcoidosis),
Wegener
, aggressive NK - cell
leukemia, , blastic or monomorphic
NK - cell leukemia/lymphoma, enteropathy - associated T - cell lymphoma, T -

(monocyte - macrophage - histiocyte) 가
가
가
TNKL
²⁾³⁾
EBV가
^{4) Cruz¹⁷⁾}

중심 단어 : T - EB

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