

측두골에 발생한 랑게르한스 세포 조직구증 2예

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Two Cases of Langerhans Cell Histiocytosis of the Temporal Bone

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-ABSTRACT -

The most well-known childhood histiocytosis, previously known as histiocytosis X, is Langerhans cell histiocytosis and includes the clinical entities of eosinophilic granuloma, Hand-Schüller-Christian disease and Letterer-Siwe disease. The hallmark of this disease is the presence of a clonal proliferation of cells of the monocyte lineage containing the characteristic electron microscopic findings of a Langerhans cell. The skeleton is involved in 80% of patients and bone lesions may be single or numerous and are seen most commonly in the skull, especially in the temporal bone. Prognosis is relatively favorable. We have experienced two cases of Langerhans cell histiocytosis with temporal bone involvement which were treated surgically in two children. (J Clinical Otolaryngol 2005;16:132-135)

KEY WORDS : Histiocytosis, Langerhans-cell · Temporal bone · Surgery.

서론

1941 Farber¹⁾
Hand - Schuller - Christian disease, Letterer - Siwe disease, Eosinophilic granuloma 3가

1953 Lichtenstein
X(histiocytosis X)
(Langerhans cell

histiocytosis)
(monocyte - macrophage system)
가

가
3) 5

11
2

: 2005 4 20
: 2005 5 30
: , 602 - 715

3가 1

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증례

증례 1 :
5 가 1

2 × 2 cm 가 (600 rad/6), 2 가 6 (squamous portion) 증례 2 : 11 가 (Fig. 1).

가 10 dB, 35 dB , 10 dB 가 CD1a S-100 (ma- (Fig. 2). stoid segment) (bony erosion) T2 , T1

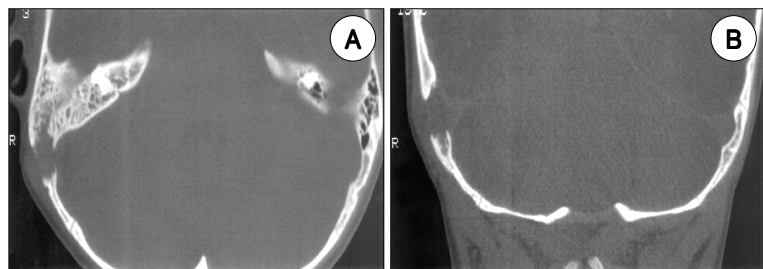


Fig. 1. Preoperative temporal bone CT in case 1. It demonstrates osteolytic lesion on squamous portion. A : Axial view, B : Coronal view.

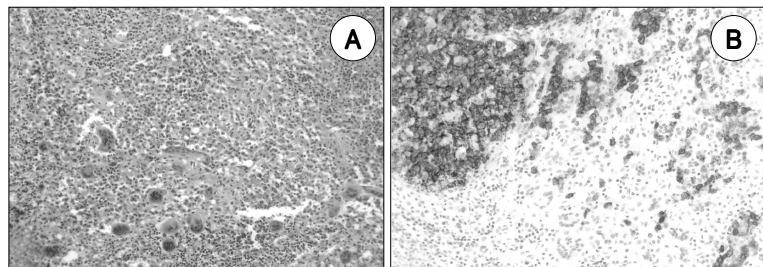


Fig. 2. Pathologic findings of Case 1. A : The osteolytic bone of case 1 shows infiltrates of histiocytes, eosinophils and multinucleated giant cells (H & E, ×200). B : The immunohistochemical stain shows positive reaction of Langerhans cells for CD1a (Immunohistochemical stain, ×200).

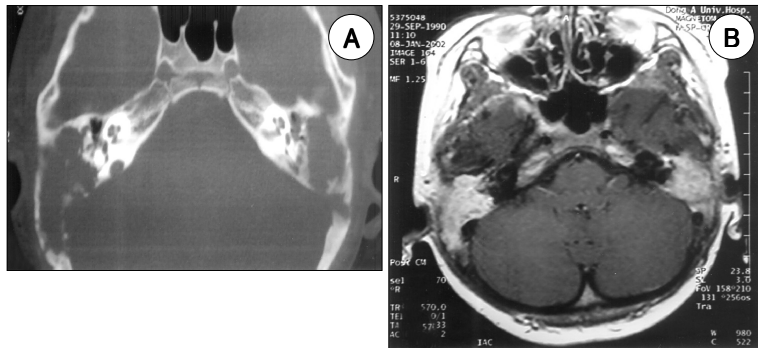


Fig. 3. Preoperative images in case 2. A : Axial temporal bone CT shows massive osteolytic lesion on mastoid area, squamous portion and tegmen. B : Axial T1-weighted MR image demonstrates a high signal mass lesion in both mastoid portion and middle ear cavity, infiltrating into sigmoid sinus and dura.

S
(Fig. 3).

5

80%

가

3)

10%

2)

가 가

6)

7

30%

25%

가

8 (etoposide)

2 6

가

(tegmen tympani)

6)7)

3)6)7)

Birbeck

granule

8)9)

CD1a,

S - 100 , CD4, CD11b, CD11c, EBM11, UCHM1,

5) T8+ KB61 HLA - DR 6)

가 가

3)

CD1a S - 100

15

50%가 5

2 가 3)6)

가

“punched out”

5

9)

13)14) 가 2

2 6

7)

3)10)11)

13)14) 1

2

S

가

11)

90%

13)

가

가 (2),

가

:

2

3)11) 16%

1

2

중심 단어 :

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