

두경부를 침범한 랑게르한스 세포 조직구증의 치료*

김 일 만

Treatment of Langerhans Cell Histiocytosis of Head and Neck

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Objective : The author presents a retrospective analysis of 11 patients with Langerhans cell histiocytosis (LCH) of head and neck to describe its optimal treatment strategies.

Methods : This study involved five male and six female patients with a median age of 11 years. Sites of involvement included calvarium (9 cases), temporal bone (1 case), and cervical node (1 case). Ten patients had a solitary symptomatic lesion in the time of presentation. The localized LCH of calvarium (eosinophilic granuloma) was treated by surgery alone (8 cases) or surgery plus radiotherapy (1 case). An infant presented with multifocal disease received systemic chemotherapy. The patient who had extensive temporomastoid lesion with aural mass underwent radical mastoidectomy and cranial irradiation due to incomplete resection.

Results : The time from initial symptom to diagnosis ranged from 10 days to four months (mean, 54 days). In cases with calvarial involvement, a painful lump with tenderness was the most common symptom at admission and a typical punched out lesion was seen on skull radiographs. No recurrence of symptoms or relapse of radiological evidence was observed during follow up, except for one infant who died of progressive multisystemic LCH.

Conclusion : It is suggested that complete surgical excision is the treatment of choice for unifocal cranial LCH, leaving radiotherapy for possible relapse. Systemic chemotherapy can also play an important role in the control of more extensive diseases in children.

KEY WORDS : Eosinophilic granuloma · Langerhans cell histiocytosis · Treatment strategy.

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(Langerhans cell histiocytosis)

23,31)

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가

uloma)

19),

15),

16),

2)

(eosinophilic gran-

26),

7),

12),

11)

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가

4,9,10,20,30,32)

대상 및 방법

1987

2001

2002

결 과

11, 1

가

5 : 6 11

(1 ~ 32) 10

6 9 4 53.6

3 63.7

가

“ (well) ” 5 “ (no recurrence) ”

가 2 9 4 가

3 2 1 10 가 1

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9

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8 x 4mm 40 x 40mm

1

11

Table 1. Summary of 11 cases with Langerhans cell histiocytosis of head and neck

Age(yrs), Sex	Location	Presentation(duration)	Treatment	Follow-up (mos)	Outcome
1mo,M	Multifocal(skull, chest, back, leg)	Multiple subcutaneous lesions(1mo) no organomegaly	Excision, chemotherapy	6	Remission, not progressive
24,F	Parietal	Scalp mass, posttraumatic headache(4mos)	Total resection, radiotherapy (900cGy)	20	Well
8,F	Orbitofrontal	Pain on eyebrow(2mos)	Total resection, autograft cranioplasty	26	Well
10,F	Occipital	Occipitalgia(1mo)	Total resection, cranioplasty	47	Well
8,M	Occipital	Lump with pain(2mos)	Total resection	89	No recurrence
11,M	Temporomastoid	Bloody otorrhea, otalgia postauricular swelling(2mos)	Biopsy, excision and radical mastoidectomy, radiotherapy (600cGy)	83	No recurrence
30,F	Lateral neck	Palpable cervical node(15days)	Excisional biopsy	110	No recurrence
27,M	Parietal	Lump with tenderness(10days)	Total resection	111	No recurrence
32,F	Temporal	Tender scalp mass(15days)	Total resection	110	No recurrence
20,M	Parietal	Progressive lump enlargement(3mos)	Total resection	88	No recurrence
11mos,F	Temporal	Temporalis swelling(2mos)	Excision, refused further treatment	2	Died of multisystemic disease(skull, lung, lymph node)

mo : month, mos : months

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 (Table 1).
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 (Fig. 1A,
 B).
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 T1 T2

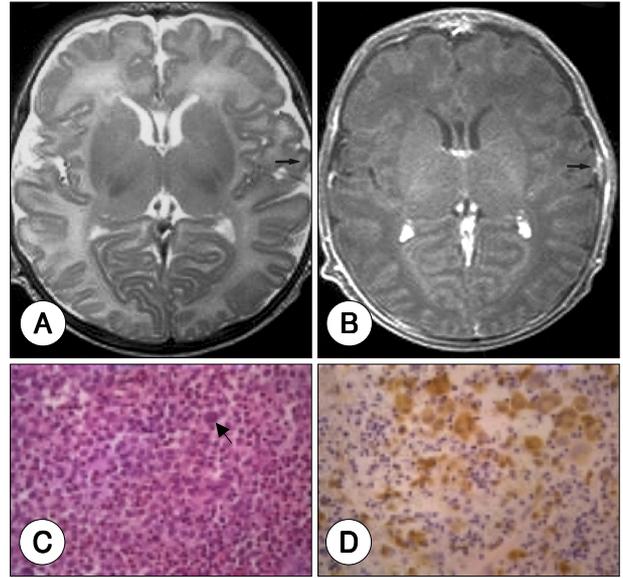


Fig. 1. Case 1. Axial brain magnetic resonance images(A and B) revealing a plaque-like soft tissue lesion(arrow) with epidural and subgaleal extension that has isointensity and uniform gadolinium enhancement in the left temporal area. Photomicrographs of histopathological examination(C and D). Hematoxylineosin stain(C) demonstrating multinucleated large pale-staining Langerhans cell histiocyte(arrow) with eccentric nuclei and nuclear grooves. There are foci of necrosis, fibrosis and scattered eosinophils and lymphocytes(original magnification x 200). Positive immunohistochemical staining for antibody to S-100 protein(D) is seen as dark brown areas in both the nucleus and cytoplasm of the Langerhans cells(original magnification x 400).

Birbeck
 (Fig. 1C, D)
 증 례 2 :
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 (monostotic)
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 T1 T2

(Fig. 2).

(Fig. 3).

600cGy

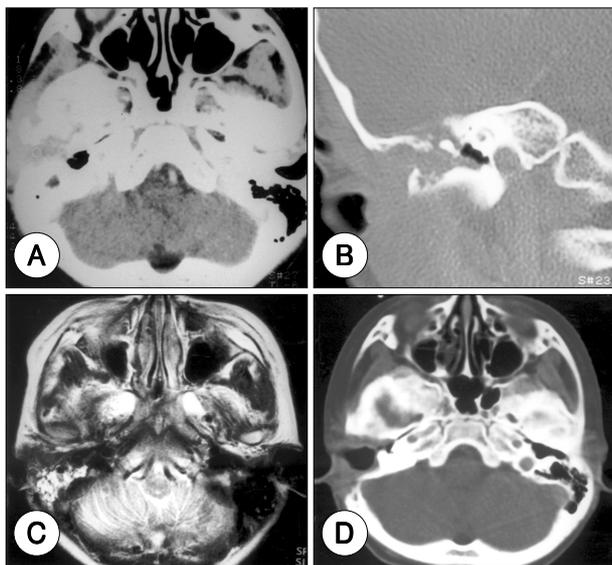


Fig. 2. Case 2. A and B : Axial enhanced and bone window coronal computed tomography(CT) scans showing osteolytic defect in right temporomastoid region with large, homogeneous, well enhancing soft tissue mass. Note the erosion of the ossicular chain and the floor of middle cranial fossa. C : Preoperative axial T2-weighted magnetic resonance image showing mainly hyperintense lesion filling the right mastoid antrum and air cells. D : Postoperative axial CT scan demonstrating no residual or recurrent mass.

증례 3 :

20

3

가가

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가

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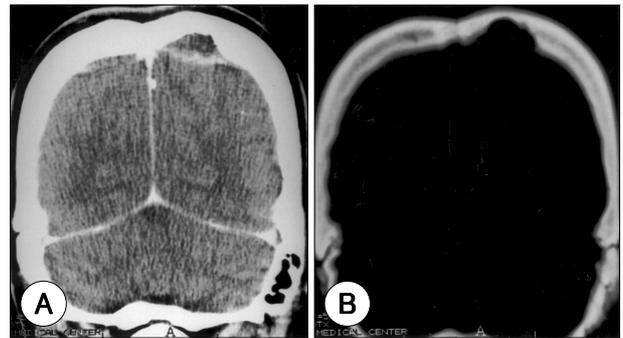


Fig. 3. Case 3. The eosinophilic granuloma in the left parietal bone is demonstrated. The lesion enhances moderately and a tail of dural enhancement is also seen(A). The inner table of calvarium is more destroyed than the outer, which has a bevelled appearance(B).

고찰

가

2

18,27,31)

가가

2,3)

가

Letterer - Siwe disease

13,17,18,25,32)

Birbeck

S - 100 CD1a

6,8,28)

2

Birbeck

가

xanthogranuloma),
xanthoastrocytoma),

(juvenile
(pleomorphic

가 21). 가 50~82% 가 21). 가 9). 가 2 (reossification) (remodeling) 13). 16% 가 5 29). 가 15,28) 가 22,27). 가 7,15,21,24). 가 14), Yasko 32) 가 5,14,21,22). 가 6 가 2 가 6), 3,6,13,29), 2 가 29). 2 가 2). 가 2), , 1000~2000cGy 가 가 가

가 가 가 2 (reossification) (remodeling) 13). 16% 5 29). 15,28) 가 2 가 2 (multifocal) 가 (multisystemic) 14), 16,24,27). 50~70% 1,15,17) 11 가 결론 : 2002 3 25 : 2002 7 31 : 700 - 712 194 : 053) 250 - 7335, : 053) 250 - 7356 E - mail : bach1158@dsmc.or.kr

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