

## 측두골에 발생한 섬유성 이형성증 1례

가<sup>1</sup>, 채세용<sup>1</sup> · 노혜일<sup>1</sup> · 이종우<sup>1</sup> · 류재영<sup>1</sup> · 정은선<sup>2</sup>

### A Case of Fibrous Dysplasia of the Temporal Bone

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#### — ABSTRACT —

Fibrous dysplasia is a bone disorder of unknown etiology in which normal bone is replaced with fibrotic tissue and disorganized immature bone trabeculae. The disease has three variants ; monostotic, polyostotic, and Albright's syndrome. Monostotic fibrous dysplasia especially involving mainly squama in temporal bone is rare. The clinical course of them is unpredictable. Monostotic form arises usually in childhood and often come to an arrest at puberty. Polyostotic form without endocrine dysfunction tends to appear at a slightly earlier age than the monostotic form and may progress into middle age. Mostly their involvement is unilateral. Monostotic form involving temporal bone is associated with conductive hearing loss attributable to compression of the external auditory canal. Potential problems include cholesteatoma, recurrence and malignant transformation. We report a case of monostotic fibrous dysplasia of temporal bone involving mainly squama, which is successfully treated with canaloplasty and skin graft. We also observed the electron microscopic findings. (**J Clinical Otolaryngol 2000;11:109-114**)

**KEY WORDS** : Fibrous dysplasia · Temporal bone · Squama · Electron microscopy.

#### 서 론

1937 McCune Bruch<sup>1)</sup>가  
1938 Lichtenstein “fibrous dysplasia”  
McCune - Albright's syndrome 3  
가 . 70% 10%가

: 1999 10 9

: 2000 2 12

: , 442 - 060

가

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3)

4)

#### 증 례

31 가 6

10

가

10

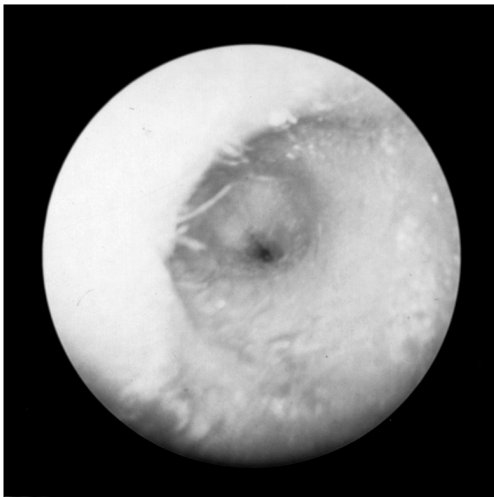


Fig. 1. The bony external auditory canal is narrowed circumferentially.

가  
가  
가  
1.5 mm (Fig. 1).  
20 dB, 8 dB  
B  
glass appearance)  
(Figs. 2 and 3).  
Tc-99m  
(Fig. 4).

1999 3 24

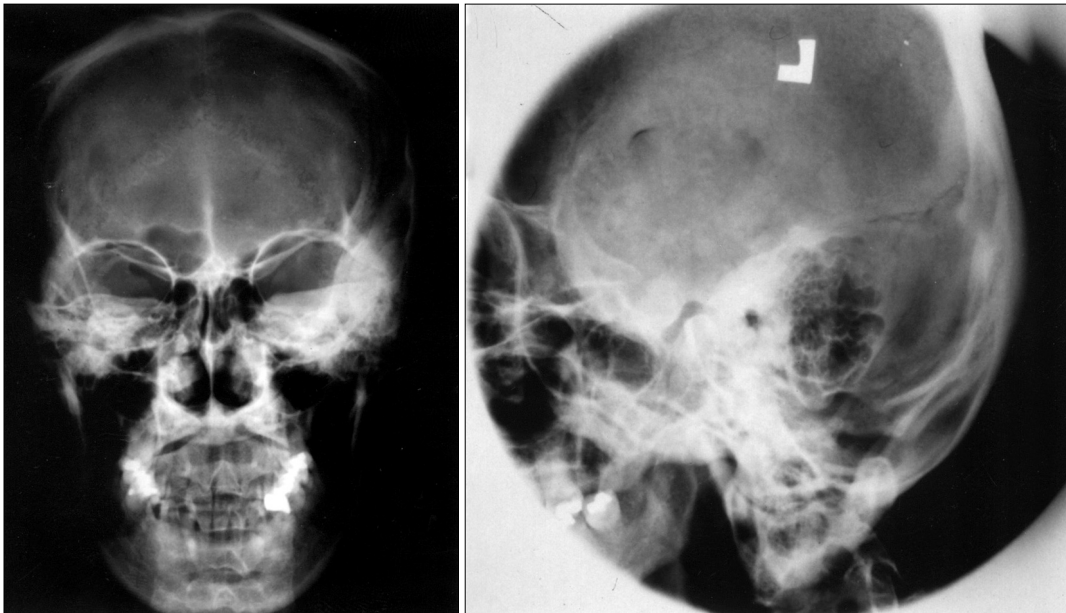
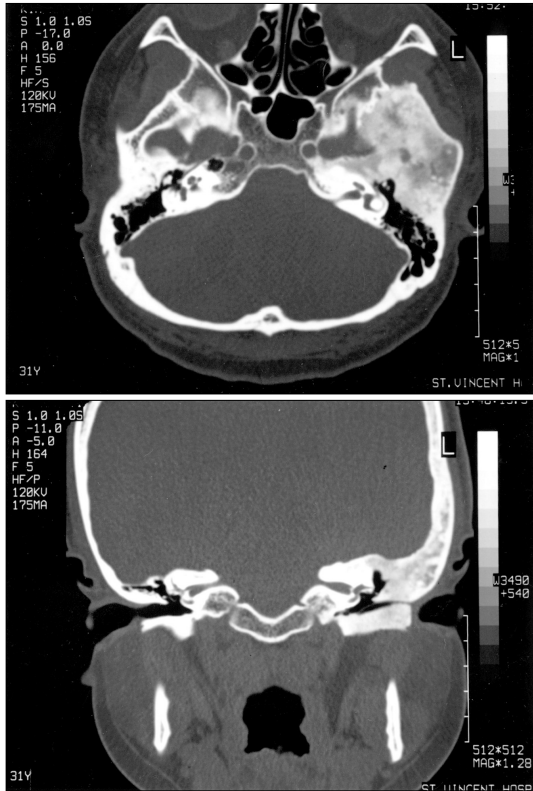


Fig. 2. Skull AP and law's view show marked overgrowth and sclerosis of bone in the squamous, petrous part of left temporal bone with ground glass appearance. Mastoid part is preserved.



**Fig. 3.** Axial CT scan at the level of internal acoustic meatus (top) shows expanding lesion of squamous and anterior region of petrous part of the left temporal bone produce sclerotic ground glass appearance. Note sparing internal acoustic meatus and the inner ear, facial canal and mastoid air cells. Coronal CT scan at the level of external acoustic meatus (bottom) shows expanding sclerotic lesion of squamous and tympanic part of temporal bone with thinning of cortex.

(cancellous bone)

10

(woven bone)  
(Fig. 5).

(nucleoli)

가

가

(rough endoplasmic reticulum)  
(Fig. 6).

11

(Fig. 7)

3  
가

고 찰

5)  
6) 7)  
14 ,  
11 , McCunn - Albright's 8  
McCunn - Albright's  
가 .

(fibula),

100%

가

18%

10)

(56%),  
(42%)<sup>8)</sup>

9) 1987 76 가  
가

(50%),

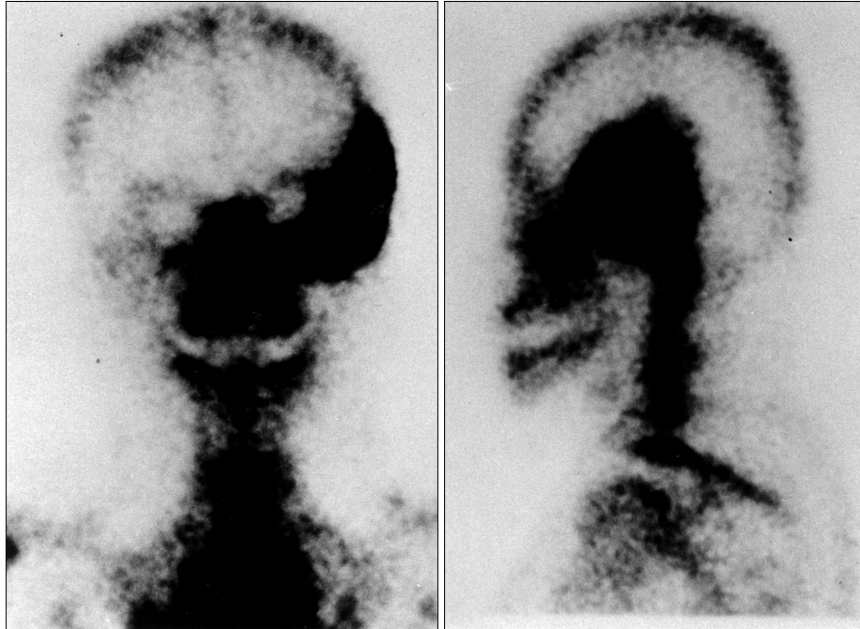
8)

50%

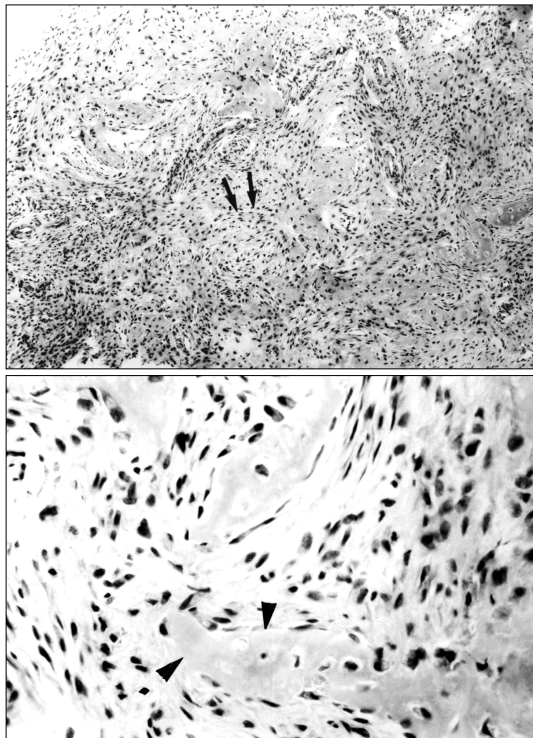
가<sup>8)</sup>

가가

11)

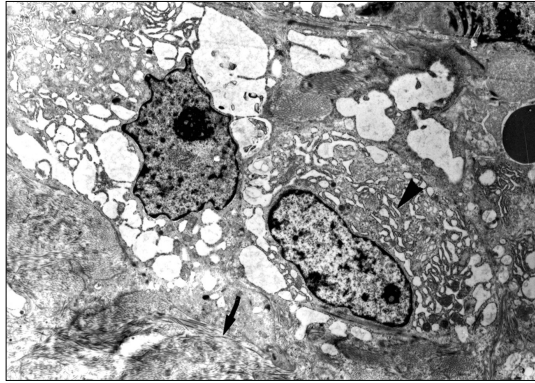


**Fig. 4.** Tc-99m HMDP imaging of skull AP and lateral view show intense hot uptake in the left temporal bone area.

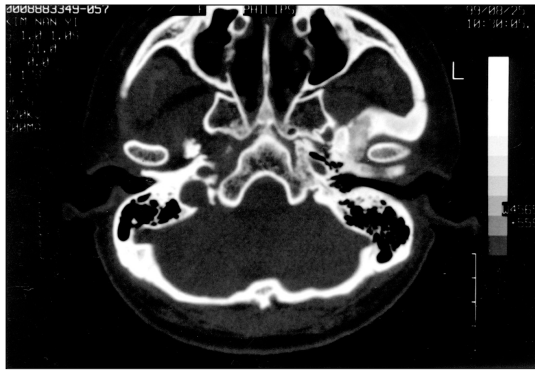


**Fig. 5.** Characteristic overgrowth of fibrous tissue (arrow) and haphazardly scattered trabeculae (arrow heads) of the woven bone (H-E stain, top ;  $\times 50$ , bottom ;  $\times 200$ ).

0.5%  
12)  
13)  
,  
17%  
가 10% 13)  
,  
(cystic) Paget  
가 13)14) Paget  
가 56%  
가 가  
23%  
21%  
4)  
67%가  
15) Tc-99m  
HMDP Ga-97  
16)  
T1



**Fig. 6.** Electron micrograph of the stromal cells of fibrous dysplasia reveals fibroblastic features of elongated nucleus, inconspicuous nucleolus. Abundant dilated rough endoplasmic reticulum (arrow head) and the surrounding collagen fibers (arrow) is shown (× 5000).



**Fig. 7.** Axial CT scan at the level of external acoustic meatus shows patent external auditory canal six months after canaloplasty.

, T2 가

16)

(woven bone)

가

10  
32

가 17) 14)

13)

가 가

가

, 6

14)

13)

가 가 16)

가 , 가  
 18) GNAS 가  
 가 , G- cAMP 가  
 가 interleukin - 6 가  
 가 19)  
 20)  
 가 가  
 가  
 중심 단어 :

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