

## 제 1 형 다발성신경섬유종과 동반된 접형골 형성부전 1 예

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## A Case of Sphenoid Dysplasia Associated with Neurofibromatosis Type 1

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## -ABSTRACT-

Sphenoid dysplasia is rare and has reported to occur in 13.5% of patients with neurofibromatosis type 1 (NF-1). It is characterized by a defect in all or part of the greater wing, an elevation of the lesser wing of sphenoid, the distortion of the sella and the enlargement of the ipsilateral orbit. A 33-year-old man was admitted to our hospital with right zygomatic, maxillar fracture after trauma to right periorbital area. While treating the patient, pulsatile exophthalmos was found, and through computer tomographic scan, the absense of the wing of the sphenoid bone was found, along with the enlargement of the orbital cavity. He had skin problems since he was very young and a long history of decreased visual acuity in his right eye, but went untreated for them. After treating for the right zygomatic, maxillar fracture, and further discussion with the neurosurgery, he underwent sphenoid wing reconstruction with a number of surgicels and gelfoams. Postoperatively, his pulsating exophthalmos was not seen. We report successful treatment of sphenoid dysplasia associated with neurofibromatosis type 1. (J Clinical Otolaryngol 2004;15:303-306)

KEY WORDS : Sphenoid bone · Neurofibromatosis 1 · Exophthalmos.

## 서 론

1 (NF - 1)  
(pathognomic)<sup>1)2)</sup>

<sup>3)</sup>

NF - 1

가

## 증 례

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: , 600 - 730

33

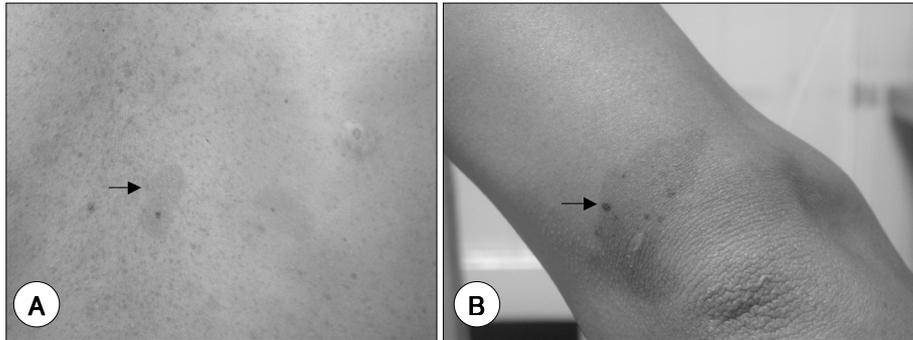
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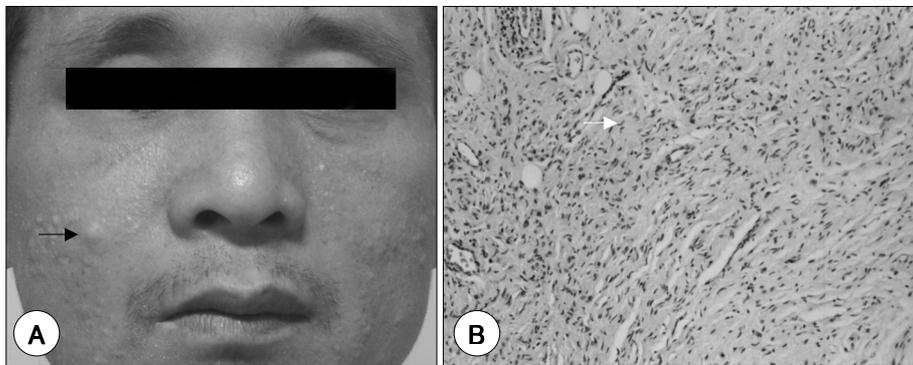
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**Fig. 1.** Photography of cafe-au-lait-spot. A : on back, B : on elbow joint.



**Fig. 2.** A : Neurofibromas on right cheek of patient (arrow). They were biopsied for histologic examination, B : Section shows interacting bundles of elongated cells having wavy dark-staining nuclei (white arrow) and collagen-rich stroma (x 100, H-E stain).

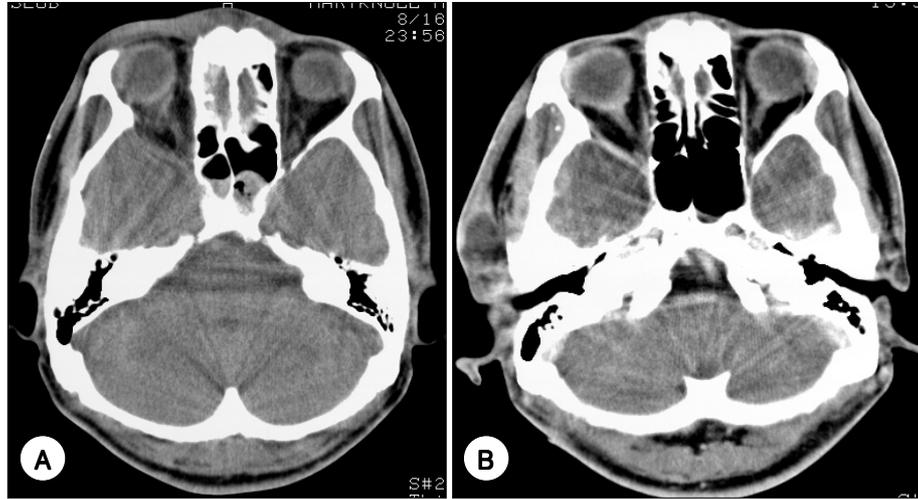
NF - 1                    cafe - au - lait spot  
 (Fig. 1),                    가  
                                   (Fig. 2A).  
 0.3,                    0.8                    , Hertel  
 13 mm,                    13 mm                    가  
 ,  
 가  
 (Fig. 2B).  
 ,  
                                   (Figs. 3 and 4).  
 7

20  
 (frontotemporal approach)



**Fig. 3.** Coronal CT scan showing the defect in lesser and greater wings of right sphenoid bone (arrow).

Fig. 4. Axial CT scan of right sphenoid dysplasia. After 10 days later from operation, right orbit was divided from right temporal lobe with surgicels and gelfoams. A : Preoperative view, B : Postoperative view.



surgicel gelfoam (Fig. 4).

8가 , 가 1  
 Recklinghausen . Recklinghausen  
 cafe - au - lait spot,  
 Lisch  
 . Cafe - au - lait spot

3  
0.5

고 찰

가

5 mm ,  
 15 mm cafe - au - lait spot 6  
 (freckling),

<sup>3)5)</sup> 1929 Dandy<sup>4)</sup>

Lisch , 1 가

2가

NF - 1 1~7%

<sup>5)</sup> NF - 1

<sup>7)</sup> NF - 1

<sup>4)</sup>

7~10

가

6가

6

NF - 1

<sup>6)</sup>

<sup>5)</sup>

NF - 1 <sup>5)6)</sup>

<sup>8)</sup>

NF - 1  
 methyl methacrylate  
 lastic reconstruction)<sup>10)</sup>  
 (autogeneous reconstruction)  
 .<sup>11)</sup> Papay <sup>12)</sup>  
 -  
 6  
 gelfoam  
 surgicel  
 중심 단어 : 1

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