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－ 증 례 보 고 －

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= Abstract =

Familial Hemifacial Spasm
- Case Report -

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The authors report two cases of familial hemifacial spasm according to the clinical and three dimensional short-range magnetic resonance angiography(3D- SRMRA) findings. In the family of the first case, there were five patient's with acquired hemifacial spasm in successive generations. Three male and two female patients in successive generations of the same family developed acquired hemifacial spasm. Four patients were on the left side and one was on the right side. In the family of the second case, there were three patient's with acquired hemifacial spasm in successive generations. Two brothers developed left- sided hemifacial spasm. 3D- SRMRA finding of the probands demonstrated that both anterior inferior cerebellar artery in first case and anterior inferior cerebellar artery & vertebral artery in second case offend the 7th cranial nerve respectively. The presence of familial clustering of these rare disorders suggest an underlying genetic predisposition. All family pedigrees suggest that a pattern of autosomal-dominant inheritance with partial penetrance.

KEY WORDS Familial hemifacial spasm· Genetic predisposition· Autosomal- dominant inheritance.

서　론

6)

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7　　　(root
exit zone, REZ)

증　례

증 례 1 :

36

* 　　1999　10　15

5

(platysma)

. 3D- SRMRA   (anterior
inferior cerebellar artery)

(Fig. 1).   3
(microrvascular decompression)

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6   .

,

3
5   (Fig. 2)
73   4
3D- SRMRA

(Fig. 3A).
45   4
. 3D- SRMRA
(posterior inferior cerebellar artery)
(Fig. 3B).
48   30   3

3D-
SRMRA   .   52
10

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Fig. 1. 3D-SRMRA(three dimensional short-range magnetic reson-ance angiography) imaging of the proband in case I, showing horizontal section through the region of pontomedullary junc-tion. This demonstrate that both AICA(anferior inferior cerebe-llar artery) offend the facial nerve.
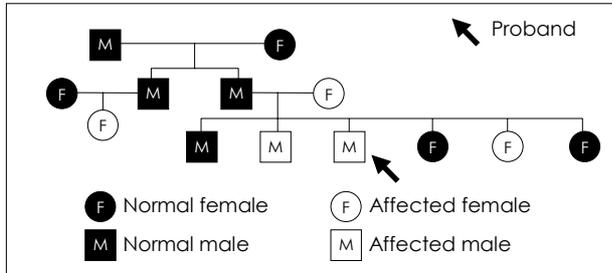
Fig. 2. The Pedigree of case I(proband arrow) showing five patients with acquired hemifacial spasm(white box) in succe-ssive generations. M male, F female.

증 례 2 :
56
.   10
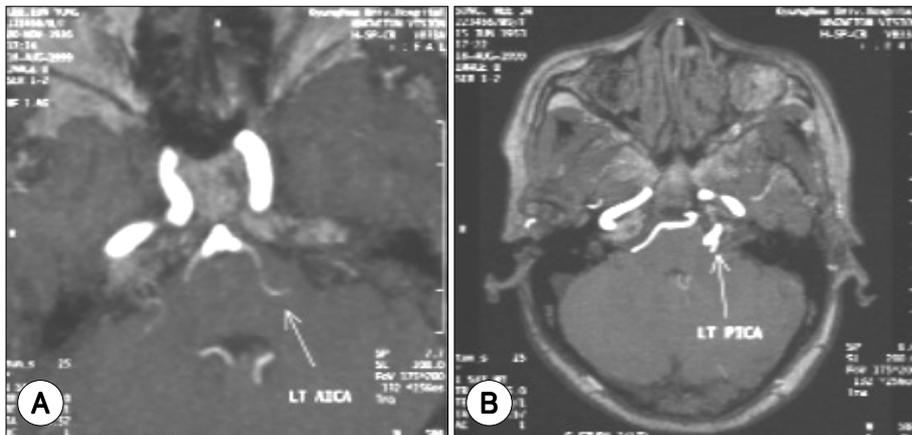
Fig. 3. 3D-SRMRA(three dimensi-onal short-range magnetic reson-ance angiography) imaging of another ones in case I, showing horizontal section through the re-gion of pontomedullary junction. A demonstrates that Lt. AICA(an-ferior inferior cerebellar artery) offends the facial nerve. B de-monstrates PICA(posterior inferior cerebellar artery) offends.
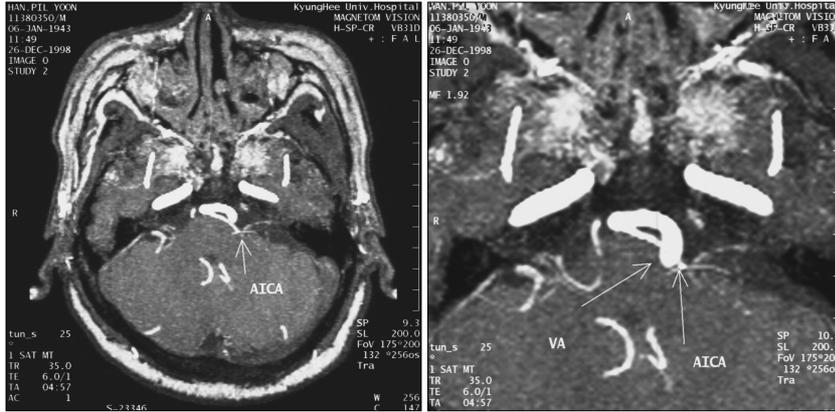
Fig. 4. 3D SRMRA(three dimensional short-range magnetic resonance angiography) imaging of the proband in case II, showing horizontal section through the region of pontomedullary junction. This demonstrate that AICA(anferior inferior cerebellar artery) and VA(vertebral artery) offend the facial nerve.
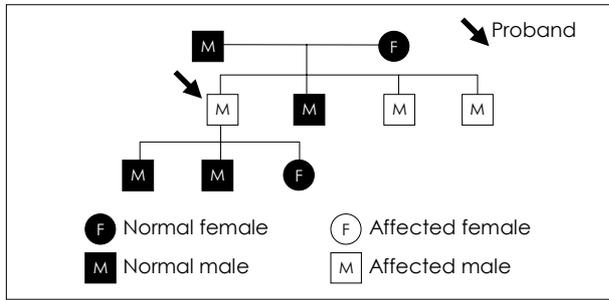
Fig. 5. The Pedigree of case II(proband　arrow) showing three patients with acquired hemifacial spasm(white box) in successive generations. M　male, F　female.

. 3D- SRMRA

(Fig. 4).

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3

2

(Fig. 5)　　　　　49　　6

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고　　찰

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(platysma)　　　　　　　.

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(tics),

(asymmetric blepharospams),

(facial myokimia),　　　　　(Meige's syndrome)

[6].

120

. 1947　Campell　Keedy[3]

.

.

(ephaptic transmi-

ssion)　　　　(axonic excitation)

[10]. Moller　[9]

(kindling

phenomenon)

(antidromic stimu-

lation)

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결 론

5

. Friedman [6]   3

5

2   20

Carter [4]

3   3

Carter [4]

...

. 1991   Coad [5]

2   3

(anterior inferior cerebellar

artery)   redundant loop

. Micheli [8]

Barbosa [2]

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[7].   ,   Bell   (Bell's palsy)

[1].   Fig. 2   Fig. 5

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1

2

. 3D-SR-

MRA

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