

상행대동맥의 거대한 확장과 하행대동맥 박리를 동반한 Marfan 증후군 1례

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= Abstract =

A Case of Marfan Syndrome with Dissecting Aortic Aneurysm Involving Thoracic and Abdominal Aorta

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Marfan syndrome is an inheritable connective tissue disorder with protean clinical manifestations involving the ocular, skeletal and cardiovascular system. It is well established that the major cause of morbidity in Marfan syndrome is the dilation of the aortic root as well as the associated aortic regurgitation and dissection. Aortic dissection usually begins just above the coronary ostia in the ascending aorta and extends the entire length of aorta. We report a rare case of Marfan syndrome with aortic dissection which begins distal to the left subclavian artery in the descending aorta.

KEY WORDS : Marfan syndrome · Aortic aneurysm · Aortic dissection.

서 론

Marfan 증후군은 Marfan 1) Marfan 2) Marfan 3) Marfan 4) Marfan 5) Marfan 6-8) 9,10) Marfan 11) 1959 1

교신저자 : , 140 - 743 657
) (02) 709 - 9204,) (02) 749 - 2864

가 Marfan 1

증 례

: , , 20 .

: .

: 2

3 4 5

1

가

: 5

10

가 6

Marfan

가 :가 , ,

60

:

100/60mmHg, 75/ ,

36.8 , 160cm, 41Kg .

170cm ,

85cm 가 .

2

가

:

가 10.1g/dl

RPR

(rapid plasma reagin) TPHA(treponema pallidum

heamagglutination assay)가 , FTA -

ABS(fluoscent treponemal antibody - absorption)

. RA, CRP, ASO

X

(Fig.

1).

:

58mm valsalva

68mm

(Fig. 2).

4 1

:

70mm

(flap) (false lumen) (Fig. 3).



Fig. 1. The chest P-A view showed tortuous aorta and cardiomegaly with severe scoliosis.

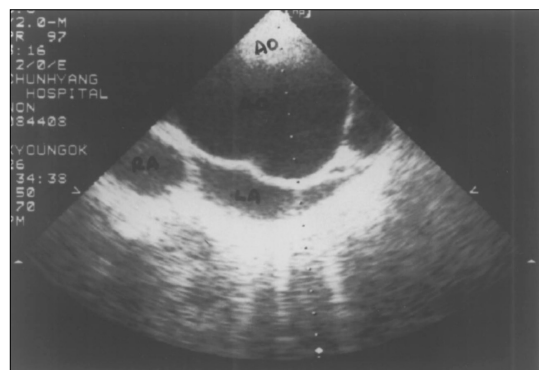


Fig. 2. Apical 4-chamber view showing marked dilatation of aortic root.

30/15(22)mmHg,
14mmHg, 138/31(67)mmHg
2.7L/
70mm

가

(true lumen)
(Fig. 4).

:

3

70mm

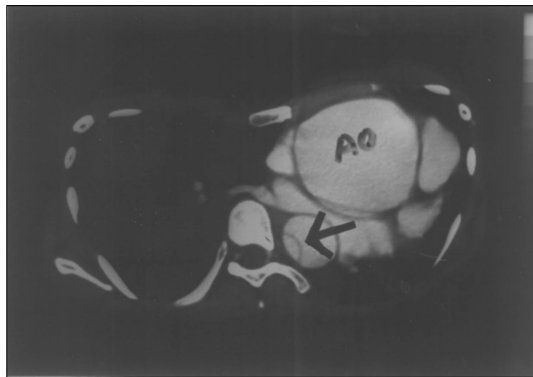


Fig. 3. The chest CT showed marked dilatation of ascending aorta and dissection of descending aorta with intimal flap (arrow).

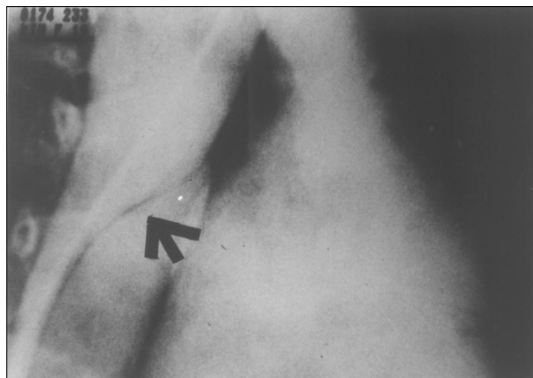


Fig. 4. Aortography showed marked dilatation of ascending aorta and intimal flap with true and false lumen (arrow).

고찰

Marfan

가

가

가

4).

Marfan

Marfan

2).

Marsalese ¹²⁾ 84

38%

, 65%

, 74%

13)

19

Marfan

17 (89%)

88%,

42%

가

4).

Valsalva

가

14).

Roman ²⁾ Valsalva

가

3).

70mm

50mm

60mm

15).

Marfan

가

| | | | |
|----------------------|-------------------|--------|-----------------|
| DeBaakey | graft | 4). | composite |
| 10% | 가 | 가 | , |
| 가 | 가 | 가 | , |
| 24 (67%), | 36 | 가 | 55mm |
| 5 (14%) | 1986 | 11) 1 | composite graft |
| 가 | 1987 | 13) 17 | 2) . Gott 15) |
| 가 | 1 | 가 | composite graft |
| | | | 1%, 10 |
| | | 76% | |
| | | 가 | |
| Pyeritz 16) | | | |
| 가 | 60mm | | 요 약 |
| . Gott 15) | Marfan | | |
| 가 | 14 | 13 | |
| 60mm | | | |
| 4). Roman 2) | 가 | Marfan | , |
| | | | |
| | | 가 | |
| | | | |
| 가 | 가 | | |
| 가 | 가 | | |
| 가 | 2). Marsalese 12) | 81 | |
| Marfan | 99 | 84% | |
| 가 | | 48%가 | |
| | | 35 | |
| , Murdoch 17) | Johns Hospkin | | |
| 257 | Marfan | 40 | 50 |
| | 80% | | |
| (negative inotropic) | | | |
| 3). | | | |
| 가 | 가 | | |
| | | | |

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