

말단 거대증을 동반한 가족성 심장 점액종

- 복합성 점액종 1예 -

강석민 · 정남식 · 변영섭 · 임세중 · 장병철* · 조상호**

한 경 훈

= Abstract =

Familial Cardiac Myxoma with Acromegaly (Complex Myxoma)

- A Case Report -

Seok Min Kang, M.D., Namsik Chung, M.D., Young Sup Byun, M.D.,
Sejoong Rim, M.D., Byung Chul Chang, M.D.,* Sang Ho Cho, M.D.**

Department of Cardiology Division Yonsei Cardiovascular Center, Cardiothoracic Surgery,*
Pathology,** College of Medicine, Yonsei University, Seoul, Korea

Kyung Hoon Han, M.D.

Department of Internal Medicine, Han Kook Hospital, Cheju, Korea

Background : Cardiac myxomas are rare benign tumors of the heart. Although cardiac myxomas are histologically benign, they may be lethal because of their strategic position. Most cases are sporadic, but rare familial occurrence has been described.

Patients and Results : The left atrial myxoma with cerebral embolism was diagnosed in the 21 year old female and the left atrial myxoma with acromegaly due to pituitary adenoma was subsequently diagnosed in her 19 year old male sibling. The myxoma in the male patient was successfully excised.

Conclusion : Echocardiography can be used effectively in the diagnosis of atrial myxoma, detection of its possible recurrence, and screening other members of the family.

KEY WORDS : Familial cardiac myxoma · Acromegaly · Echocardiography.

서 론

(cardiac myxoma)

가 ,

1845 King¹⁾
, 1951
가
1955

2), , :
 (sporadically), Krause oria)
 1971 가 가 162cm, 56kg , 100/60
 (familial cardiac myxoma) mmHg, 64 /
 3-8) 가 ,
 (cutaneous myxoma), (cutaneous)
 (mucocutaneous pigmentation), :
 (Cushing's syndrome) .
 (acromegaly) :
 (complex myxoma) 9-13) .
 가 가 (nonfamil- : 7700mm³(
 ial cardiac myxoma) 60.5%), 10.4gm/dl, 32.2%,
 가 291,000/mm³ Na 1
 (autosomal dominant) 40mEq/L, K 4.1mEq/L, Cl 105mEq/L, CO₂ 24 mM/L
 14-15) 1977 ,
 23) .

가 .
 (movable) (friable) 2.5 × 4.3cm
 (high echogenic) 가
 (Fig. 2).

증 례 2 :

: , 19 .

:

: 4 (3)

3 , ,

증 례 1 :

: , , 21 .

: 96 4 18

: , ,

가 : 가 8

, 20

. 1 1

:



Fig. 1. Photograph of the 21 year old female who has spotty brown pigmentation on forehead, below eyes, on cheek, nose and the lips.

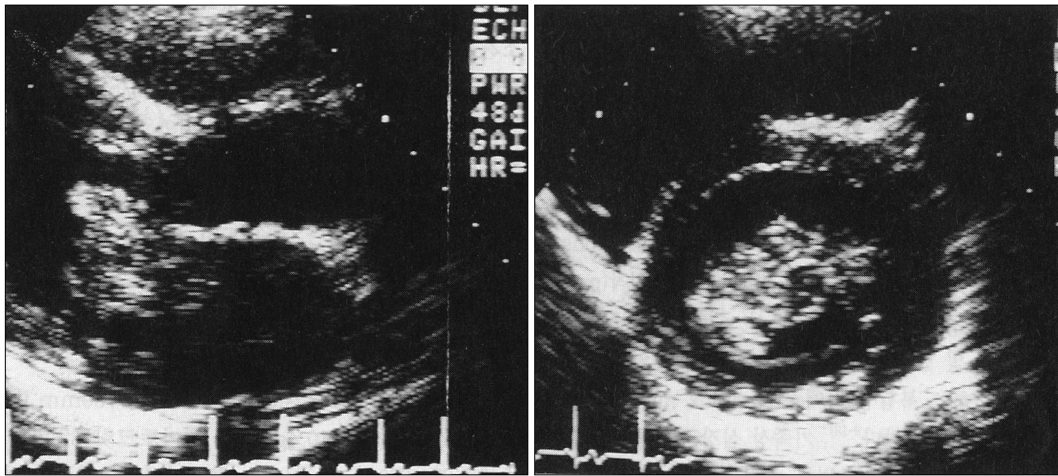


Fig. 2. Transthoracic 2-D echocardiogram(parasternal long and short axis view) showing a movable, echogenic mass in the left atrium which moves into the left ventricle during diastole.

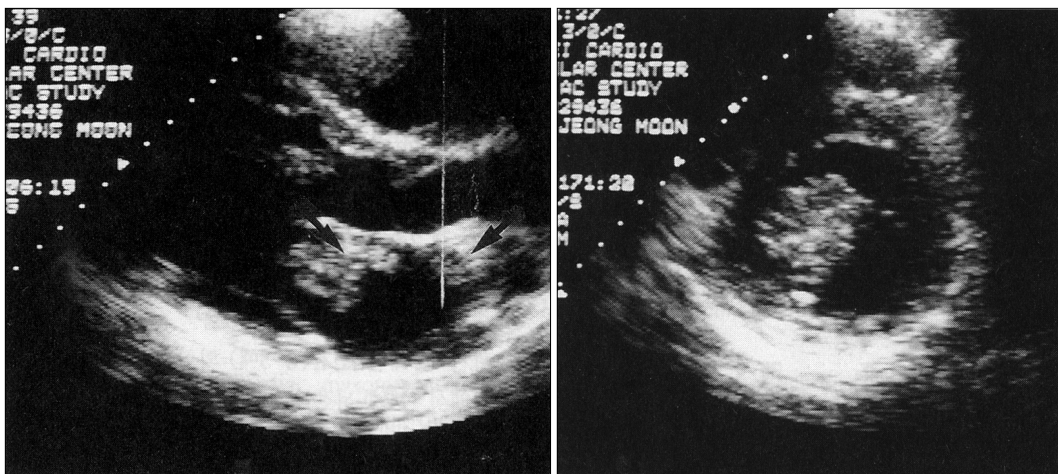


Fig. 3. Transthoracic 2-D echocardiogram(parasternal long and short axis view) showing the echogenic masses in the left atrium which moves into the left ventricle during diastole.

가 : 1
: 가

:
: 5100mm³(
180cm, 95kg . 54.5%), 13.3gm/dl, 39.8%,
140/80mmHg, 70 / 280,000/mm³ . Na
" tumor plop " 142 mEq/L, K 3.7mEq/L, Cl 108mEq/L, CO₂ 22

mM/L , 38mm :
가 , C - reactive protein (Fig. 5, 6).
: 6
somatomedin -
C 600ng/ml (: 56 177ng/ml) , 가 1cm
growth hormone 25.81ng/ml(: 0 5ng/ml) stalk 0.8×0.8cm
broad stalk 3×
: 1×1cm, 2×1×1cm
4.7×3.4cm 가 ,
12
가 (Fig. 3, 4).

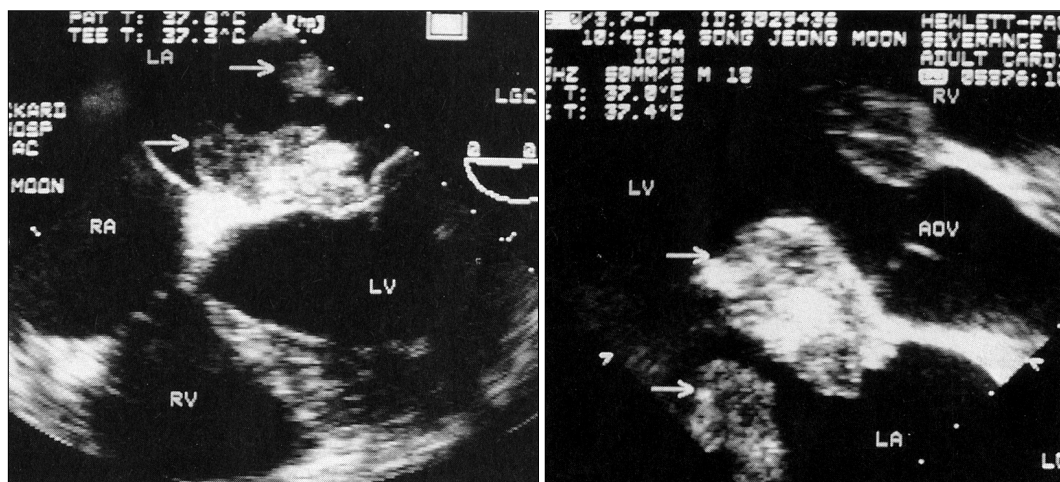


Fig. 4. Transesophageal echocardiogram showing multiple movable echogenic masses in the left atrium which were attached to the interatrial septum and the posterolateral side of the left atrium.

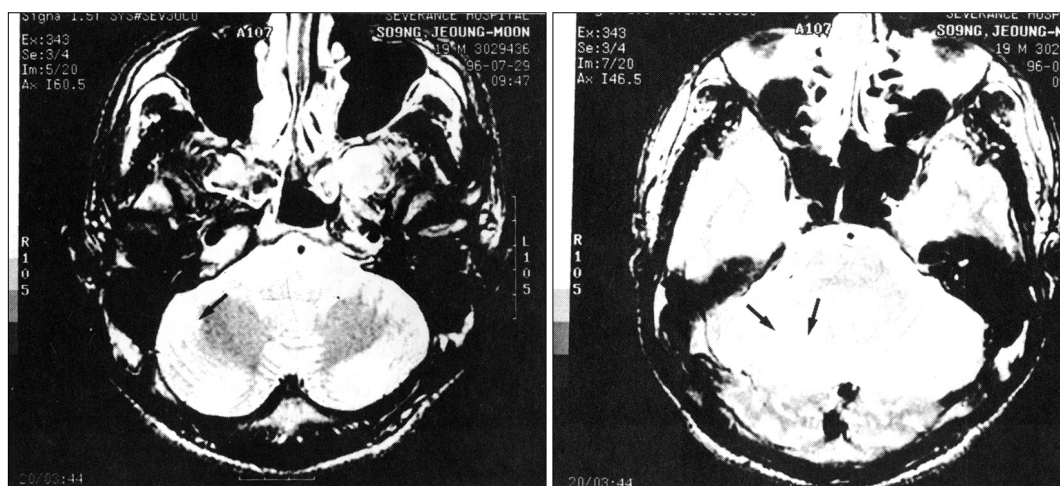


Fig. 5. Brain MRI(T₁-weighted image) showing multiple lacunar infarctions in the cerebellar hemisphere.

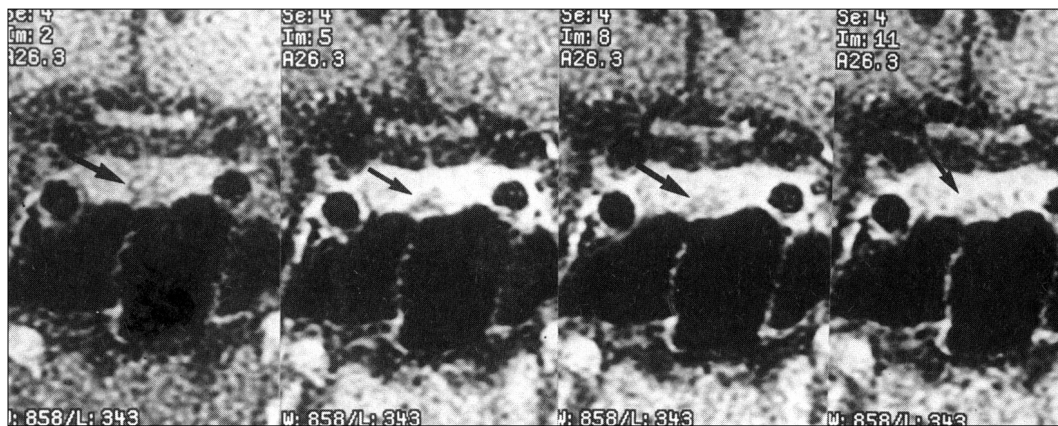


Fig. 6. On dynamic image, small sized delayed enhancing lesions are seen at the midline and the right lateral portion of the pituitary gland suggesting small adenoma (T₁-weighted Brain MRI).

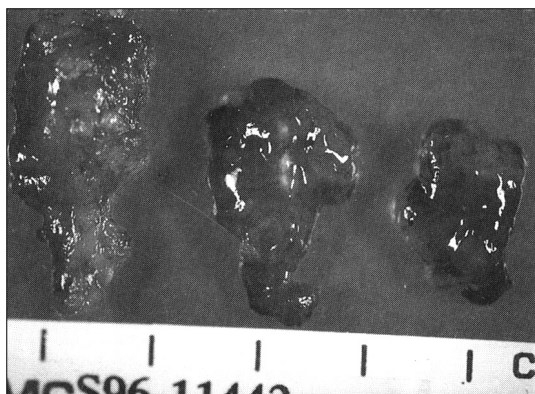


Fig. 7. Excised three myxomas appearing grossly soft, polypoid and gelatinous.

: 3 3 × 1.4 ×
1.0cm, 2.4 × 1.4 × 0.8cm, 1.5 × 1.1 × 0.7cm

가
(Fig. 7).

(Fig. 8),
(Fig. 9).

고 안

(foramen ovale)
(endocardium)

30 50% 가
. 1951

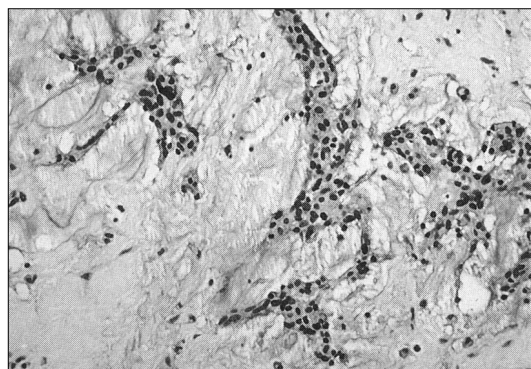


Fig. 8. Histologic section of a myxoma with characteristic abundant loose myxoid matrix in which polygonal cells are embedded and occasional blood vessels (hematoxylin and eosin, × 100).

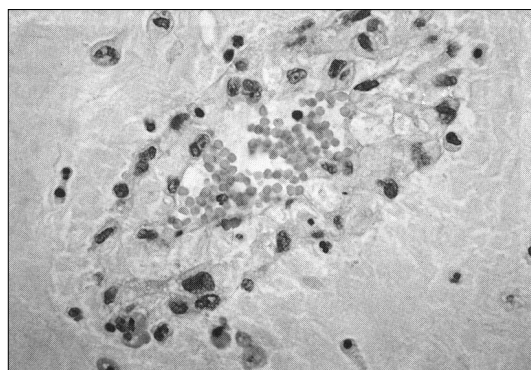


Fig. 9. Myxoma cells are characteristically lined around blood vessels (hematoxylin and eosin, × 100).

가 ,

가 5 6cm Kr -
가 75%, ause 1971 1992
15 20% 16), 3 4% 197가 13) 가
17,18) (27)
가
30 40% (embolism) (autosomal dominant)
19,20) 14,15) 가
(systemic embolism) (nonfamilial cardiac myxoma)
() , 14)
20) 가 ,
, , ,
, ,
(normal sinus rhythm) 24).
가 22)
, , , Raynaud's , ESR
가, CRP
interleukin - 6 , 가
가 가
21) (inadeq -
(sporadically) uate resection),
가 가 (intracardiac implantation),
4-7) 가 (familial cardiac myxoma) , 가 가
(com - 4 , 가
plex myxoma) , 가
(Cushing's syndrome), (acro - 12%, 22% , 가
megaly) (cutan - 1 3%
eous myxoma), (cutaneous lentiginosis) 10).
(testicular tumor) 가
10,14) 가 , ,
가 가 ,
가 가
가 가
4 6

요 약

가

plex myxoma)

(com -

References

- 1) King TW : *On simple vascular growths in the left auricle of the heart. Lancet* 2 : 428-435, 1845
- 2) Crafoord C : *Discussion on late results of mitral commissurotomy. In International Symposium on Cardiovascular Surgery. Edited by CR Lam. Philadelphia, WB Saunders Company pp202, 1955*
- 3) Krause S, Adler LN, Reddy PS, Magovern GJ : *Intracardiac myxoma in siblings. Chest* 60 : 404-406, 1971
- 4) Liebler GA, Magovern GJ, Park SB, Cushing WJ, Begg FR, Joyner CR : *Familial myxomas in four siblings. J Thorac Cardiovasc Surg* 71 : 605, 1976
- 5) Powers JC, Falkoff M, Heinle RA, et al : *Familial cardiac myxoma : Emphasis on unusual clinical manifestations. J Thorac Cardiovasc Surg* 77 : 782-788, 1979
- 6) Kleid JJ, Klugman J, Haas J, Battock D : *Familial atrial myxoma. Am J Cardiol* 32 : 361-364, 1973
- 7) Farah MG : *Familial atrial myxoma. Ann Intern Med* 83 : 358-360, 1975
- 8) Siltanen P, Tuuteri L, Norio R, Tala P, Ahrenberg P, Halonen PI : *Atrial myxoma in a family. Am J Cardiol* 38 : 252-256, 1976
- 9) Carney JA, Hruska LS, Beauchamp GD, Gordon H : *Dominant inheritance of the complex of myxomas, spotty pigmentation and endocrine overactivity. Mayo Clin Proc* 61 : 165-172, 1986
- 10) McCarthy PM, Piehler JM, Schaff HV, et al : *The significance of multiple, recurrent and 'complex' cardiac myxomas. J Thorac Cardiovasc Surg* 91 : 389-397, 1986
- 11) Vatterott PJ, Seward JB, Vidaillet HJ, Su WPD, Oftedahl GL : *Syndromic cardiac myxoma : More than just a sporadic event. Am Heart J* 114 : 886-889, 1987
- 12) Bennett WS, Skelton TN, Lehan PH : *The complex of myxomas, pigmentation and endocrine overactivity. Am J Cardiol* 399-400, 1990
- 13) VanGelder HM, O'Brien DJ, Staples ED, Alexander JA : *Familial cardiac myxoma. Ann Thorac Surg* 53 : 419-424, 1992
- 14) Carney JA : *Differences between nonfamilial and familial cardiac myxoma. Am J Surg Pathol* 9 : 53-55, 1985
- 15) Farah MG : *Familial cardiac myxoma : A study of relatives of patients with myxoma. Chest* 105 : 65-68, 1994
- 16) McAllister HA Jr, Fenoglio JJ Jr : *Tumors of the cardiovascular system. Atlas of tumor pathology. 2nd series. Fascicle 15. Washington DC Armed Forces Institute of Pathology* 1-38, 1978
- 17) Meller J, Teichholz LE, Pichard AD, et al : *Left ventricular myxoma ; Echocardiographic diagnosis and review of the literature. Am J Med* 63 : 816-824, 1977
- 18) Hall RJ, Cooley DA, McAllister HA Jr, Frazier OH : *Neoplastic heart disease. 7th ed. New York. Mc-Graw-Hill, 1382-1415, 1990*
- 19) St John Sutton MG, Mercier LA, Giuliani ER, Lie JT : *Atrial myxomas ; A review of clinical experience in 40 patients. Mayo Clin Proc* 55 : 371-376, 1980
- 20) Goodwin JF : *The spectrum of cardiac tumors. Am J Cardiol* 21 : 307, 1968
- 21) Saji T, Yanagawa E, Matsuura H, et al : *Increased serum interleukin-6 in cardiac myxoma. Am Heart J* 122 : 579-589, 1991
- 22) Burch GE, Shewey LL, Harb JM : *Coxsackie B4 viruses and atrial myxoma. Am Heart J* 88 : 634-639, 1974
- 23) 박영배 · 이명목 · 김성윤 · 권인순 · 서정돈 · 이영우 · 이성호 · 안광필 · 김종환 · 한만청 · 김용일 · 함원근 : 좌심방 점액종의 1예 보고. 순환기 7 : 47-52, 1977
- 24) Wold LE, Lie JT : *Cardiac myxomas ; A clinicopathologic profile. Am J Pathol* 101 : 219-240, 1980