Case Report

Adult Endocardial Fibroelastosis in Japan

A Case Report and a Review

Einosuke Ueda and Tatsuo Kokubu

Incidence of endocardial fibroelastosis in infants is not so rare but is rare in adults. The etiology and pathogenesis of this cardiac lesion is unknown. There is difference of opinion referable to whether the lesion in the infants developed due to the same etiology as that in the adults. In 1956 Guraieb reviewed 24 cases of endocardial fibroelastosis in adults from America and England, and concluded that the pathologic lesions were identical with those of infants, and the process of this disease in adult was considered to be congenital.

It is true that there is some confusion about the term of endocardial fibroelastosis. Davies described in his review of obscure diseases affecting the mural endocardium “it is necessary to apply strict criteria, and the term ‘fibroelastosis’ in this review is restricted to those lesions in which there is the thickened fibroelastic layer of endocardial proliferation as seen in infants with or without hypertrophy of the layer of smooth muscle.” We summarized, according to him, the only cases which were reported as endocardial fibroelastosis or endomyocardial sclerosis with marked proliferation of elastic fiber in the endocardium.

In this paper are presented an adult case of fibroelastosis recently observed in our clinic and a review of the report of this disease in Japan.

A total of seventeen cases, including our case (Case No. 17), which were considered as endocardial fibroelastosis in adults (16 years of age and over) now has been reported in Japan. A brief summary of the clinical and pathologic findings of these cases is given in Table I.

Three cases were female and fourteen male, and this sex distribution was nearly same as that of the report of Guraieb. The age of our cases varied between 16 and 74 years. A case of 74 year-old man was the oldest case of this disease in the world so far as we know. The heart usually was enlarged; only three of these cases weighed less than 400 grams. A systolic murmur was the most common type as mentioned by Guraieb, however, in three cases there were no murmurs. The blood pressure was described in eleven of seventeen cases. Six of these eleven cases showed lower than 110mmHg in systolic blood pressure, and seemed to show a tendency of low blood pressure. An electrocardiogram was made in thirteen of seventeen cases. Four of these thirteen cases showed intraventricular conduction delay, two showed low voltage, none of them showed normal ECG. In 1955 Vlad, Rowe and Keith published a detailed description of the electrocardiographic findings in endocardial fibroelastosis. Since then these findings have been certified by many authors. The typical electrocardiograms showed left ventricular hypertrophy and negative T wave in V5,6. In our cases negative T wave was observed only in two cases. This phenomenon might be the difference
### Table I

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Reporter</th>
<th>Year</th>
<th>Age</th>
<th>Sex</th>
<th>Heart weight</th>
<th>Heart sound</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Yamamura, T.</td>
<td>1959</td>
<td>74</td>
<td>M</td>
<td>340</td>
<td>—</td>
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<td>2</td>
<td>Yamamura, T.</td>
<td>1959</td>
<td>53</td>
<td>M</td>
<td>530</td>
<td>—</td>
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<tr>
<td>3</td>
<td>Yamamura, T.</td>
<td>1959</td>
<td>24</td>
<td>M</td>
<td>412</td>
<td>—</td>
</tr>
<tr>
<td>4</td>
<td>Takashima, K.</td>
<td>1960</td>
<td>24</td>
<td>M</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>5</td>
<td>Kobayashi, T.</td>
<td>1961</td>
<td>64</td>
<td>M</td>
<td>500</td>
<td>Gallop rhythm</td>
</tr>
<tr>
<td>6</td>
<td>Okada, R.</td>
<td>1961</td>
<td>19</td>
<td>F</td>
<td>—</td>
<td>No murmur</td>
</tr>
<tr>
<td>7</td>
<td>Seki, I.</td>
<td>1962</td>
<td>20</td>
<td>M</td>
<td>470</td>
<td>MI murmur</td>
</tr>
<tr>
<td>8</td>
<td>Watanabe, R.</td>
<td>1963</td>
<td>37</td>
<td>M</td>
<td>650</td>
<td>Impure</td>
</tr>
<tr>
<td>9</td>
<td>Murakami, H.</td>
<td>1964</td>
<td>33</td>
<td>M</td>
<td>510</td>
<td>Systolic murmur in apex</td>
</tr>
<tr>
<td>10</td>
<td>Asakawa, H.</td>
<td>1964</td>
<td>39</td>
<td>F</td>
<td>300</td>
<td>No murmur</td>
</tr>
<tr>
<td>11</td>
<td>Yoshida, T.</td>
<td>1964</td>
<td>40</td>
<td>M</td>
<td>650</td>
<td>Gallop rhythm, relative TI.</td>
</tr>
<tr>
<td>12</td>
<td>Murao, S.</td>
<td>1966</td>
<td>43</td>
<td>M</td>
<td>390</td>
<td>AI murmur</td>
</tr>
<tr>
<td>13</td>
<td>Kawai, N.</td>
<td>1966</td>
<td>40</td>
<td>M</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>14</td>
<td>Murao, S.</td>
<td>1967</td>
<td>16</td>
<td>M</td>
<td>815</td>
<td>Early syst. murmur (2) in apex, IIIrd and IVth sound</td>
</tr>
<tr>
<td>15</td>
<td>Kashi, T.</td>
<td>1967</td>
<td>50</td>
<td>F</td>
<td>—</td>
<td>No murmur</td>
</tr>
<tr>
<td>16</td>
<td>Takashima, T.</td>
<td>1967</td>
<td>47</td>
<td>M</td>
<td>520</td>
<td>Systolic m. in apex, IIIrd sound.</td>
</tr>
<tr>
<td>17</td>
<td>Ueda, E.</td>
<td>1967</td>
<td>31</td>
<td>M</td>
<td>630</td>
<td>Systolic m. in all ostium.</td>
</tr>
</tbody>
</table>

between in children and adults.

Many efforts to establish the etiology of the disease has been performed in every report. Takatsu tested antibodies in the serum against several viruses, such as polio virus, Coxsackie virus, encephilitis japonica virus, mumps virus and herpes virus, but none of the cases tested revealed positive reaction. Only in the case No. 10, C-reactive protein was one plus, the antistreptolysin O titer of the serum was 330 Todd unit, but no inflammatory lesions was found in the heart autopsied.

Histological finding revealed small difference in each other. In case No. 5 and No. 14 the pathological changes of the myocardium were prominent and change of the endocardium seemed to be secondary. None of all these cases showed the findings strongly suggesting inflammatory origin.

The most interesting case was Case No. 13. This 16 year old boy was diagnosed by means of endocardial biopsy. Although the histological findings showed rather strong fibroelasticosis of the endocardium, the patient, a sports player, had no subjective complaints.

### Case report

A 31 year-old man visited our clinic on April 21, 1967, with chief complaints of strong dyspnea and foot edema. The patient had been well until three years before when he felt easy fatigability and palpitation on exertion. A syncope attacked him two times last year but precise condition could not be obtained. The subjective complaints has not been so strong that he had been able to go to the usual work until severe dyspnea attack fell him down on this April. He was admitted in some hospital, the doctor of which sent the patient to our clinic for exact diagnosis. The patient died sud-

A CASE OF ENDOCARDIAL FIBROELASTOSIS

Table I

<table>
<thead>
<tr>
<th>EGG</th>
<th>Remarks</th>
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|     | B. P. 170/? 144/90 110/78  
Thrombosis of the cerebral artery.  
Marked coronary sclerosis.  
Diagnosed as myocardial infarction 6 years before.  
Coronary sclerosis  
Diagnosed as valvular disease when 15 years old. |
|     | B. P. 134/98. Lesions of the endocardium were considered secondary to myocardial change. |

| Ventricular extrasystole, T flat in V₅₋₆, ST dep. in V₆ | Laboratory test normal.  
B. P. 104/80 98/82, RA-, CRP-, ASLO-,  
Referred as endomyocardial sclerosis.  
B. P. 100/60, CRP+, ASLO 330 Todd unit |
| LAD, ILBBB | B. P. 127/76. Cell infiltration in the myocardium.  
B. P. 102/68, Lues, Small coronary arterial sclerosis. |
| Auricular fibrillation | B. P. 128/70, Diagnosis was made by using endocardial biopsy.  
B. P. 120/60, Partial fibroelastosis  
B. P. 90/60, Total protein 4.2 gm/dl |
| LBBB | B. P. 90/70, GPT 104, GOT 81, I. I. 19  
B. P. 100/80, CRP-, ASLO-, RA-,  
Total protein 4.9 gm/dl |
| ILBBB |  
qs in V₂₋₄, ST dep. in II, aV₇, V₃₋₅,  
T inserted |
| LAD | B. P. 90/70, GPT 104, GOT 81, I. I. 19  
B. P. 100/80, CRP-, ASLO-, RA-,  
Total protein 4.9 gm/dl |
| Coronary T in V₂₋₄ |  
QS in V₁, V₂  
Intraventricular conduction delay  
LVH, QS in V₅₋₆  
Low voltage, T, flat, auricular fibrillation  
Ischemic T in V₄₋₅ |

denly before hospitalisation.

There was neither history of jaundice nor of rheumatic fever.

Physical examination revealed a well developed, moderately nourished, and pale colored man with slight edema on eye lid and marked edema on the lower extremities. There was no clubbing of the fingers. Pulse was 56 per minute with occasional extrasystole, blood pressure 100/80mmHg, and respirations 32 per minute. There was no enlargement of the lymph nodes. The heart was markedly enlarged to the both sides. Soft systolic murmurs were audible in all ostium, second pulmonic sound was accentuated with marked splitting. The abdomen was distended without any obvious ascites.

The liver was palpable 5cm below the right costal margin with tenderness.

The laboratory test revealed a slightly positive urinary protein. Urinary sugar was negative. The sediment of the urine was normal. The hemoglobin was 12.1 gr/dl and the white cell count was 6200 per cubic mm. The blood sedimentation rate was 11 mm. per one hour and 28 per two hours. The serum sodium was 137 mEq/L, the potassium 4.3 mEq/L, the chloride 110 mEq/L and BUN 35 mg/dl.

The serum total protein was 4.9 gr/dl and the electrophoretic pattern of the serum showed as follows: albumin 58.9 per cent, α₁-globulin 3.3 per cent, α₂-globulin 7.5 per cent, β-globulin 12.6 per cent, γ-globulin 17.7 per cent. The bromsulphalein retention was 34 per cent after 45 minutes, SGOT 28 u., alkaline phosphatase 14 King-Armstrong units. The serological test were as follows: C-reactive protein was negative, ASLO 40 Todd unit, RA-test negative, Waaler-Rose reaction 14 dils.

The electrocardiogram showed low voltage in all limb leads and QS patterns in I, aVF, V₅, V₆ with occasional ventricular premature beat (Fig. 1). Chest X-ray film demonstrated a markedly enlarged, globular heart (cardiothoracic ratio: 78 per cent) with signs of slight lung congestion (Fig. 2).

Clinical course: The patient was administered the digitalis preparation with little effect. He was found to be dead in the morning on July 1, 1967.

Autopsy findings: The heart was markedly

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Fig. 1. Electrocardiogram taken two weeks before death.

Fig. 2. X-ray film of the chest, showing the enlarged globular heart.

Fig. 3. View of the left ventricle, showing marked endocardial thickening with pearly gray color.

Fig. 4. A microscopic view of the endocardium of the left ventricle with fibroelastic thickening. Weigert stain X 20

enlarged and weighed 630 gm. The heart was consisted markedly dilated right atrium, markedly dilated right ventricle and markedly hypertrophied left ventricle. (Fig. 3) Mural thrombi were found in the both atrium. Neither coronary sclerosis nor coronary occlusion were found. The inner surface of the left ventricle was smooth and pearly gray color, with leathery hardness.

Microscopically, the increase of the thickness of the endocardium was due to proliferation of both collagen and elastic fibers (Fig. 4) Some times the fibroelastic tissue within the endocardium extended for a short distance into the myocardium. In many of the thicker areas of endocardium there was either a layer of cardiac muscle, completely surrounded by the fibroelastic tissue. In other part of the heart muscle some of them showed fatty degeneration and picnolysis.

The aorta appeared to be normal either macroscopically or microscopically. The lung was edematous. The right weighed 670gm. and the left 560 gm. A few thrombi already organized were found in the both lung. The liver weighed 1270 gm. The liver cells showed picnolysis, and significant diffuse fibrosis were found in the both lobes suggesting the long lasting congestion of the liver.

The right kidney weighed 110 gm., left 130 gm. The several hollows suggesting old infarctions were found on the surface of both kidneys.

Pathological diagnosis was as follows;
1) diffuse endocardial fibroelastosis of the left ventricle with mural thrombi in the both atrium. 2) old infarction in kidneys. 3) diffuse fibrosis of the liver.

**DISCUSSION**

Recently much attentions are paid upon the "Primary Myocardial Disease" (PMD) which might involve endocardial fibroelastosis. TAKATSU reported twenty cases diagnosed as PMD clinically. Endocardial biopsy was performed in fifteen of these twenty cases and two of these cases, both 16 year old boys, were diagnosed as endocardial fibroelastosis. This data suggest about tenth of clinically diagnosed PMD may be endocardial fibroelastosis.

SELLERS, KEITH and MANNING pointed out the characteristic clinical and electrocardiographic patterns, even though we could not differentiate fibroelastosis from other PMD. Our case revealed normal laboratory test except low value of total serum protein, 4.9 gm/dl. The electrophoretic pattern was normal. The same phenomenon was observed in case No. 15, the total serum protein was 4.2 gm/dl. WUHRMANN named the disease of myocardial degeneration as "Myokardose" which might originate from low serum albumin levels. OKADA found the correlation between the thickness of subendocardial layer of the endocardium and the serum albumin level, that is, the lower the serum albumin the thicker the endocardium. These results suggest that fibroelastosis could be produced by not only congenital but also acquired condition.

FISHER reviewed fifteen cases of endocardial fibroelastosis in children or infants. Microscopic findings of his cases seemed completely same as our cases.

Five of fifteen of his cases combined extra-cardiac anomalies, but non in our cases. Four of his cases showed valvular involvement, aortic valve alone in two cases, mitral valve alone in one case and combined involvement in one case. But not were described as valvular involvement in our cases. Many authors found no histological difference between adult and infants or children. We also could not find the difference microscopically between adults and infants. We could assume that the only difference between them was quantitative.

**SUMMARY**

The cases of endocardial fibroelastosis in adults reported in Japan are briefly summarized. Seventeen cases were found.

Main clinical signs were congestive heart failure without any remarkable murmur, ischemic signs of left ventricle on ECG.

A case of endocardial fibroelastosis, 31 year old man, was presented. No particular signs was found clinically except for the low value of serum protein, 4.9 mg/dl.

We assumed that the disease might be a
congenital, not acquired.

Acknowledgement

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REFERENCES


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