A CASE OF ANOMALOUS PULMONARY VENOUS DRAINAGE FROM THE ENTIRE LEFT LUNG ASSOCIATED WITH COMPLETE HEART BLOCK

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A 22-year-old female patient presenting complete atrioventricular block and giant P waves in electrocardiogram had anomalous pulmonary venous drainage from the entire left lung. There was normal drainage from the right lung and no associated atrial septal defect or other intracardiac abnormalities. After a permanent pacemaker was implanted, she manifested signs and symptoms of heart failure. Although the anomalous pulmonary vein was anastomosed to the left atrium, intractable heart failure continued. She died six months later after surgical intervention. Postmortem examination revealed diffuse interstitial fibrosis throughout the myocardium.

ALTHOUGH many authors described the characteristic findings and classifications of the primary myocardial disease or idiopathic cardiomyopathy, the concept of the disease has been changing and yet not well established. For many cardiologists, the clinical diagnosis of the primary myocardial disease is one of exclusion and is suggested by the presence of cardiomegaly in relatively young subjects with left ventricular hypertrophy, conduction disturbances, or heart failure, without a preceding history of any cardiac diseases and without any clinical evidence of acquired valvular heart disease, congenital heart disease, or diseases of the coronary arteries.

On the other hand, there are eight criteria in making a diagnosis of cardiomyopathy at necropsy, four negative and four positive. The four positive features are: (1) cardiomegaly; (2) endocardial thickening; (3) mural thrombus; (4) scars or other lesions. The four negative features are: (1) no coronary artery disease or anomaly; (2) no valve disease or anomaly; (3) no hypertension; (4) no shunt inside or outside the heart.

Described here is a case showing abnormal electrocardiographic findings and developing severe heart failure which might be considered as myocardial origin and moreover being accompanied by partial anomalous pulmonary venous connection with the intact atrial septum. Since the heart failure continued after the operation, a clinical diagnosis of idiopathic cardiomyopathy seemed to be proper. But histopathological findings could not be consistent with the pathological criteria of cardiomyopathy. Then what is needed

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is the cooperative studies of the clinical cardiologists and pathologists for the elucidation of this obscure disease of the myocardium.

**CASE REPORT**

The patient is a 22-year-old female clerk. She had no previous history of rheumatic fever or syncopal attack, and her family history was negative for any heart disease. When she was 12 years old, she was told to have cardiomegaly on annual chest X-ray examination, and was diagnosed as slight mitral regurgitation by cardiac catheterization and angiocardiographic techniques. Ever since she has had no complaints, so she has been remained untreated until this time.

Two weeks prior to admission to our hospital, she noticed palpitation during her ordinary work, and visited a hospital. Physical examination revealed a heart rate of 30 per minute, rhythm irregular; consciousness was clear; no signs of heart failure. Electrocardiogram showed complete atrioventricular block for which isoproterenol and prednisone were given, but they were ineffective.

Two weeks later she was sent to our hospital for further examinations and the implantation of the permanent pacemaker. On admission to our hospital, her consciousness clear; no cyanosis; no edema; heart rate 30 per minute, irregular; and no heart murmur. Chest X-ray film revealed moderate cardiomegaly (cardiothoracic ratio 65%) and abnormal shadow was present in the left upper mediastinum (Fig. 1). Electrocardiogram showed giant biphasic P wave in lead V1 and complete atrioventricular block. The QRS complex was changed from left bundle branch block with right axis deviation to right bundle branch block with left axis deviation (Fig. 2) from day to day. Occasionally atrioventricular block was free because of medication of isoproterenol (Fig. 3).

Table 1 was the results of cardiac catheterization. The elevated left ventricular end-diastolic pressure and the increase in the pressures of the right side heart were noted. Pulmonary arteriography (Fig. 4) was performed and it revealed the
A Case of Anomalous Pulmonary Venous with Complete Heart Block

Left

Right

Fig. 2. Electrocardiograms before pervenous pacing shows complete atrioventricular block with giant biphasic P wave with negative component in lead V1. The QRS complex changes from the left bundle branch block pattern with right axis deviation (left) to the right bundle branch block pattern with left axis deviation (right).

anomalous pulmonary vein from the entire left lung draining into the left innominate vein and this was identified as the abnormal shadow in the chest X-ray plain film. The left ventriculography demonstrated moderate mitral regurgitation (Fig. 5). The ejection fraction calculated from

TABLE I  PREOPERATIVE CARDIAC CATHETERIZATION DATA

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressure (mmHg)</th>
<th>EDP = end-diastolic pressure; m = mean.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left ventricle</td>
<td>130–124/46–40</td>
<td></td>
</tr>
<tr>
<td>Aorta</td>
<td>126–122/80–74</td>
<td></td>
</tr>
<tr>
<td>Pulmonary capillary (wedge)</td>
<td>20–16 (m)</td>
<td></td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>44–38/18–14</td>
<td>28–26 (m)</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>44–40/10–8</td>
<td></td>
</tr>
<tr>
<td>Right atrium</td>
<td>15 (m)</td>
<td></td>
</tr>
</tbody>
</table>

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the biplane left ventriculogram was 34.7%. All the four cardiac chambers were enlarged, but interatrial communication was not found both by oxygen saturation step-up method and by angiogram.

No contributory informations were obtained as to the etiology of the complete heart block and the cardiomegaly. Other laboratory examinations offered the following results: Hemoglobin 15.2g. per 100 ml.; W.B.C. 7300 per c.mm.; platelets 285,000 per c.mm.; urine normal. Biochemistry showed serum total protein 6.2 g., globulin 2.3 g., blood urea nitrogen 17 mg., and total cholesterol 135 mg. per 100 ml. The electrolytes were normal. The LE cell test, Raynaud's phenomenon, antinuclear antibody, and antibody to DNA were all negative. Complement fixation tests for certain viruses and toxoplasma were also negative.

A diagnosis of cardiomyopathy was made.

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Fig. 4. Late film following pulmonary artery injection. The left pulmonary vein converge to form a vertical vein (black arrow) which connects to the left innominate vein, eventually entering the superior vena cava.

Fig. 5. Left ventricular injection in ventricular diastole. Left ventricle is dilated and the wall of the left ventricle is thick. Regurgitation of contrast material into left atrium is also seen.
Fig. 6. Non-specific diffuse myocardial fibrosis (left). (HE × 63) Mural fibrosis of small arteries and remained muscle fibers are seen among the fibrous replacement of muscle (right). (HE × 170)

Two weeks after her admission to our hospital, a permanent pacemaker was implanted. After one month, she noticed leg edema, and symptoms of heart failure gradually developed, so the operation of the correction of the partial anomalous pulmonary venous connection; i.e., the anomalous pulmonary vein from the entire left lung was anastomosed to the left atrium. At the same time myocardial biopsy was done, but the specimen was not contributory to etiological diagnosis.

After the successful operation, however, dyspnea became slowly progressive, and she had been in congestive heart failure, which did not respond to the ordinary treatment using digitalis glycosides and diuretics. About six months after the operation, she suddenly developed shock and died.

At autopsy the heart, weighing 560 g., grossly showed hypertrophy and dilatation, strikingly in the right ventricle and both sides of the auricle. Histological examination revealed diffuse interstitial fibrosis throughout the entire myocardium, in which isolated muscle fibers remained. Myocardial fibrosis was prominent around the small coronary arteries which showed mural thickening, but no inflammatory cells were found both in the arterial wall and in the fibrous tissue (Fig. 6). There was no atherosclerotic change of the coronary arteries up to the extent of the anterior or posterior descending coronary artery. Some of the remained muscle fibers were rather hypertrophic, but no remarkable vacuolization of muscle fibers nor irregular muscle arrangements were found in the myocardium.

Semi-serial sections revealed the marked fibrosis in the left bundle branch (Fig. 7), moreover, between the atrioventricular bundle and the bundle branches the continuation of the conduction fibers were blocked with the fibrous tissue.

There were no changes in the sinus node and the atrioventricular node. The moderate fibroelastosis of the endocardium was found in both auricles. All the four cardiac valves were intact.

There were no significant changes in other organs such as the liver, kidney, lung and endo-
crine organs.

**Discussion**

Disorders which affect the myocardium have been classified in several ways. Some authors employ the term "primary myocardial disease" when the myocardium is the principal site of the involvement. Such terms as "idiopathic cardiac hypertrophy", "idiopathic cardiomyopathy" and "myocardosis" are almost synonymous with primary myocardial disease. The cardiomyopathies are defined as disorders of the heart muscle of unknown or obscure causes.

Primary myocardial disease is among the many conditions which lead to various manifestations of cardiac failure. The symptomatology is such that this condition now takes a specific place in the differential diagnosis in heart diseases.

The electrocardiographic changes encountered are not diagnostic, but abnormal P waves are commonly seen in patients with primary myocardial disease and suggest biventricular enlargement with intraventricular conduction disturbances. The atrioventricular block of varying degree has been also reported to occur. Since this patient had both atrial enlargement and secondary fibroelastosis in their endocardium, the giant biphasic P wave with negative component in lead V₁ might be suggestive of the diagnosis.

It seemed more proper from a clinical standpoint that this patient was a case of primary myocardial disease because of the presence of the abnormal electrocardiographic findings including conduction disturbances and of the severe heart failure which might be myocardial origin. Marked elevation of the left ventricular end-diastolic pressure supports the latter evidence. Histopathological findings showed the non-specific diffuse myocardial fibrosis. Mural thickening of the small arteries was prominent in and around the fibrotic foci in the myocardium and no inflammatory change was noted around both arteries and their walls.

It is considered that the vascular change might be secondary one resulting from the myocardial fibrosis, though it could not be excluded, of course, that it may be due to an old inflammation in the myocardium.

James described the hereditary medial necrosis of small coronary arteries in which the primary lesion is focal degeneration of smooth muscle cells in the tunica media and the secondary lesions are often intimal proliferation over the collapsed tunica media. This medial necrosis of small coronary arteries (0.1 to 1.0 mm in diameter) has been observed in a number of heritable diseases including familial cardiomyopathy. He suggests that hereditary medial necrosis may be an etiologic factor in the development of obscure cardiomyopathies, especially if there are associated arrhythmias and conduction disturbances, bouts of syncope or sudden death. Moreover, when this hereditary medial necrosis involves the small coronary arteries with associated cardiac defects requiring surgery, this may be a further contributing factor to both surgical and postoperative morbidity and mortality.

In our case, a persistent elevation of the right atrial pressure also may contribute in acceleration of the fibrosis throughout the myocardium. The prominent fibrosis around the small arteries with thickening of the wall has not yet described in the cases of primary myocardial disease. In general, cases with congenital anomalies of the heart belong to the secondary myocardial disease according to the pathological criteria of the cardiomyopathy.

The patient has also intact atrial septum. Partial anomalous pulmonary venous drainage is often associated with atrial septal defect. A review of the literature reveals only 41 cases of anomalous pulmonary venous drainage with intact atrial septum. The anomalous pulmonary venous drainage is hemodynamically very similar to a secundum atrial septal defect with a left to right shunt, whether heart failure occurs or not in the patients with anomalous pulmonary venous connection depends upon the pulmonary blood flow and the change in the pulmonary vasculature. Because of developing heart failure, anomalous pulmonary venous connection was corrected, but the patient remained in the states of heart failure after the operation. It seemed, therefore, more difficult to consider that this congenital heart anomaly was the cause of the heart failure in our case.

Finally, of additional importance is that partial anomalous pulmonary venous connections encompassing all of the pulmonary veins on the left side are relatively rare. According to Mason et al., this condition has not been recognized previously in conscious patient.

Since this patient has the congenital heart anomaly, a partial anomalous pulmonary venous connection, the heart of this patient does not belong to the primary myocardial disease based on the pathological criteria but it is a case of...
idiopathic diffuse myocardial fibrosis with prominent fibrous change around the arteries.

REFERENCES