Extreme Left Atrial Enlargement

Some Characteristic Features

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ENLARGEMENT of the left atrium is well known as one of the earliest manifestations of rheumatic mitral valvular dysfunction. On rare occasions this chamber may become enormous, expanding to the right and posteriorly to form a huge sac that encroaches upon adjacent structures and may eventually rest against the right chest wall. The clinical course of patients who develop such extreme left atrial enlargement is remarkably uniform, and the diagnosis can be suspected from the presence of a number of characteristic symptoms and signs.

Although Hewett in 1849 \(^1\) is generally credited with the original description of aneurysmal dilatation of the left atrium in rheumatic mitral disease, the chamber in the case which he reported was actually only the size of "a large orange." The first instance of truly extreme left atrial enlargement was that of Owen and Fenton in 1901.\(^2\) They described a 40-year-old woman with an 18-year history of progressive dyspnea. Because the classical signs of a right pleural effusion were present, thoracentesis was performed, and to their surprise the chest tap yielded gross blood. Clinically this patient was thought to have an aneurysm of the thoracic aorta, but subsequent postmortem examination revealed severe mitral insufficiency and a giant left atrium containing 900 ml. of blood. It was evident that the enlarged left atrium had produced the clinical signs interpreted as pleural fluid. Thus the first reported case dramatically illustrates one of the pitfalls in the management of patients with this condition. There have been numerous reports of extreme left atrial enlargement since this initial one 60 years ago, although the degree of atriomegaly in many of the subsequent cases has frequently been less marked.\(^3\)\(^-\)\(^18\)

It is the purpose of this report to present the clinical features of 10 patients with extreme left atrial enlargement seen at the Massachusetts General Hospital, six of whom were also studied post mortem, during the past 30 years. One of these cases (M.D.) has been separately reported.\(^19\) In order to establish uniformity within the group, we included only those patients whose left atrium either touched the right chest wall or extended to within approximately 1 cm. of it on the routine 6-foot postero-anterior roentgenogram or orthodiagram. The x-rays of the ten patients are shown in figures 1 to 3. Measurements of blood volume and left atrial capacity are reported when such information was available.

Observations

Some of the pertinent clinical features are summarized in table 1.

Age and Sex

There were six women and four men in the group. Only one patient, a woman aged 42, still survives. Of the nine who have died, the average age at the time of death was 40 and the oldest was 55.

Incidence

The incidence of this condition can be only approximated. Four of the 10 patients were treated in the past at the House of the Good Samaritan and are among the half-dozen or so who are known to have developed extreme left atrial enlargement in the 3,500 persons followed at that institution since 1921 for the treatment of rheumatic heart disease.

Previous Rheumatic Fever

All except one patient (P.W.) had had at least one known episode of acute rheumatic
fever with carditis, and six patients had had multiple attacks. All the patients, with the above exception, had suffered at least one severe, prolonged attack, and two patients (V.K. and J.L.) had had progressive symptoms of cardiac failure dating from their first and only known episodes of acute rheumatic fever. In the remaining seven patients, the interval between the first attack of rheumatic fever and the first clear-cut cardiac symptoms averaged 12 years.

**Symptoms**

Some degree of congestive heart failure was invariably present in all patients for several years prior to death. The one surviving patient is in chronic mild failure. Despite the enormous heart size and chronic failure, however, many patients did remarkably well for years. For example, the patient who is still alive does all of her housework and leads an active life. The oldest patient in the series (G.H.) was in chronic heart failure for over 25 years, was a semi-invalid only in her later years, and actually died at the age of 55 of bronchopneumonia complicating a subdural hematoma after a fall. Only one patient (V.K.) was essentially a complete invalid during the last 2 years of her life as the result of her heart disease.

Chest pain was present in nine patients and in five it was severe enough to constitute the

**Figure 1**

Hoarseness attributable to paralysis of the left vocal cord, presumably the result of pressure of a dilated pulmonary artery against the left recurrent laryngeal nerve (Ortner's syndrome), was present in only one patient. In no instance did the enlarged left atrium itself cause any nerve compression.

**Signs**

Extreme cardiomegaly was uniformly present.

All patients had the characteristic systolic murmur of mitral regurgitation, and in all but one (A.S.) this was grade III (on a scale of 0 to VI) or louder, with an accompanying thrill in six instances. Transmission of the murmur to the right lung base as well as to the left was specifically recorded twice. Similarly, all patients had an apical diastolic rumble, with an accompanying thrill in six. Four patients were thought to have a Graham Steell murmur of functional pulmonary insufficiency, and seven were considered to have tricuspid insufficiency.

Accentuation of the pulmonary second sound was present in all patients, and in six
Table 1

Salient Clinical Features in Ten Patients with Extreme Left Atrial Enlargement *

<table>
<thead>
<tr>
<th>Patient, age, sex</th>
<th>Status</th>
<th>No. attacks acute rheumatic fever</th>
<th>No. years known atrial fibrillation</th>
<th>Right chest pain</th>
<th>Dysphagia</th>
<th>Electrocardiogram</th>
<th>Cardiac diagnosis</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>I.C. 42, F</td>
<td>Alive</td>
<td>1</td>
<td>19</td>
<td>+</td>
<td>0</td>
<td>AF, Axis +90°</td>
<td>MR, ms, ai, tr</td>
<td>Frequent hemoptysis</td>
</tr>
<tr>
<td>M.D. 28, M</td>
<td>Dead</td>
<td>3</td>
<td>11</td>
<td>++</td>
<td>0</td>
<td>AF, Axis +100°†</td>
<td>MR, ms, tr, as</td>
<td>Died suddenly</td>
</tr>
<tr>
<td>A.S. 36, M</td>
<td>Dead</td>
<td>6</td>
<td>16</td>
<td>++</td>
<td>++</td>
<td>AF, Axis +90°</td>
<td>MR, ms</td>
<td>Died after insertion of Starr valve prosthesis</td>
</tr>
<tr>
<td>E.D. 37, M</td>
<td>Dead</td>
<td>3</td>
<td>4</td>
<td>++</td>
<td>++</td>
<td>AF, Axis +100°†</td>
<td>MR, ms, tr</td>
<td>Frequent hemoptysis</td>
</tr>
<tr>
<td>V.K. 37, F</td>
<td>Dead</td>
<td>1</td>
<td>5</td>
<td>+++</td>
<td>0</td>
<td>A. flutter</td>
<td>MR, ms, tr</td>
<td>Left renal embolus, hospitalized for last 4 mos. of life; died suddenly</td>
</tr>
<tr>
<td>J.S. 39, F</td>
<td>Dead</td>
<td>1</td>
<td>11</td>
<td>+++</td>
<td>+</td>
<td>AF, Axis +100°</td>
<td>MR, ms</td>
<td>Died suddenly</td>
</tr>
<tr>
<td>J.L. 41, F</td>
<td>Dead</td>
<td>2</td>
<td>24</td>
<td>+</td>
<td>0</td>
<td>Nodal rhythm Axis +120°</td>
<td>MR, MS, TR</td>
<td>Splenic and left renal emboli; died after emergency appendectomy</td>
</tr>
<tr>
<td>P.W. 41, M</td>
<td>Dead</td>
<td>0</td>
<td>9</td>
<td>?</td>
<td>0</td>
<td>AF, intraventricular block †</td>
<td>MR, ms, TR</td>
<td>Marked ascites, died suddenly</td>
</tr>
<tr>
<td>M.G. 49, F</td>
<td>Dead</td>
<td>2</td>
<td>9</td>
<td>+</td>
<td>0</td>
<td>AF, Axis +110°</td>
<td>MR, ms, as, ai</td>
<td>Chronic congestive failure for over 25 years; paralysis of left vocal cord. Died of subdural hematoma and bronchopneumonia</td>
</tr>
<tr>
<td>G.H. 55, F</td>
<td>Dead</td>
<td>7</td>
<td>25</td>
<td>+</td>
<td>++</td>
<td>Nodal rhythm LVH, RVH</td>
<td>MR, ms, tr</td>
<td></td>
</tr>
</tbody>
</table>

* Figures apply to status presently or at time of death.  
† Only standard leads I-III available.  
A, confirmed by autopsy; MR, mitral regurgitation; MS, mitral stenosis; AS, aortic stenosis; AI, aortic insufficiency; TR, tricuspid insufficiency (upper case denotes a major, lower case, a minor lesion); O, none; +, slight; ++, moderate; ++++, severe; AF, atrial fibrillation; LVH, left ventricular hypertrophy; RVH, right ventricular hypertrophy.
the mitral first sound was also increased. An opening snap of the mitral valve was recorded in only one patient (V.K.).

Vigorous precordial pulsations were usually present. Two patients (E.D. and J.S.) were noted to have systolic pulsations of the right posterolateral chest wall, and one (G.H.) had a systolic expansion noted in the second and third right interspaces anteriorly.

Dullness to percussion at the right posterior lung base was noted in the nine patients in whom a description of the chest findings was available. This was associated with diminished to absent tactile fremitus in this area. These findings were indistinguishable from pleural effusion, and actually two patients (G.H. and J.L.) erroneously had thoracentesis which produced gross blood from puncture of the left atrium, although no other mishap resulted.

In general, these patients were of small stature, undernourished, and prone to cardiac cachexia.

Clinical Diagnosis

All patients in this series were thought clinically to have mitral insufficiency as the major valvular lesion, with some degree of accompanying mitral stenosis present in all and tricuspid insufficiency in seven. Slight aortic stenosis was thought to be coexistent in two cases, and mild aortic insufficiency in two. The clinical diagnosis was uniformly confirmed in six patients at postmortem examination.

Electrocardiograms

Complete 12-lead electrocardiograms were available in seven patients, and the standard limb leads in the remaining three. The pertinent features are recorded in table 1.

There was a striking similarity among the tracings, all but one showing the pattern of right axis deviation and, in the seven cases in which all 12 leads were available, left ventricular hypertrophy as well. Clear-cut combined ventricular hypertrophy was present in two instances and was suggested by the combination of left ventricular hypertrophy and right axis deviation in the remainder.

Atrial fibrillation had been present in all cases for many years. Two women (J.L. and G.H.), however, eventually developed an established nodal rhythm, which was not the result of digitalis toxicity. Both had previously manifested a multiplicity of arrhythmias, including atrial flutter, nodal tachycardia, complete atrioventricular block, bigeminy, and paroxysmal atrial tachycardia with block. These arrhythmias, in contrast to the nodal rhythm ultimately established, were often produced by digitalis, to which both were unusually sensitive.

The rare manifestation of changing from atrial flutter to paroxysmal atrial tachycardia with block when digitalis administration was excessive was repeatedly documented in one case (V.K.).

Roentgenograms

By definition the patients in this series had extreme enlargement of the left atrium, which extended either to the right chest wall or to within approximately one centimeter of it. The cardiothoracic ratio was over 88 per cent in each case. Fluoroscopically, moderate to marked biventricular enlargement was uniformly noted. The pulmonary artery segment was usually quite prominent, and the pulmonary vessels were variably congested. Calcification of the mitral valve was observed twice.

The esophagus was displaced posteriorly in all patients, to the left in six, and to the right in two. Passage of barium was frequently delayed at the point of dorsal angulation of the esophagus at the superior level of the cardiac silhouette. Marked elevation of the left main-stem bronchus was present in all cases, producing flattening of the carinal angle, which usually approached 180 degrees. Atelectasis of the right lower lobe was apparent on eight roentgenograms.

Blood Volume

Blood volume was determined in five patients with the Evans-blue dye method. It was markedly increased in all five instances (table 2), averaging 180 per cent above the predicted values.

Cardiac Catheterization

Right heart catheterization was performed
in only two patients (I.C. and A.S.). The resting pulmonary wedge pressures were moderately elevated, measuring 24 and 22 mm. Hg, respectively, as was the pulmonary artery pressure (51/25 and 55/20 mm. Hg, respectively). The pulmonary arteriolar resistances were only slightly increased.

Left heart catheterization was also performed in patient A.S. by the retrograde aortic technic. Selective left ventricular angiography, which was attempted in order to assess the degree of mitral regurgitation, could not be accomplished, as the catheter was swept into the left atrium during systole. In our experience this phenomenon has been observed rarely and only in the presence of severe mitral insufficiency.

Cardiac Surgery

Mitral valvulotomy was performed in one patient (J.L.). The calcified leaflets formed a rigid orifice through which there was a high degree of regurgitation and moderate mitral stenosis. Although the valve was split so as to admit two and a half fingers, the regurgitation persisted.

A Starr valve prosthesis was inserted into a second patient (A.S.). At surgery the left atrium was estimated to contain about 2 liters of blood. Although the prosthesis was easily implanted, the patient developed refractory congestive failure and died 1 week after operation.

Postmortem Examination

Pertinent findings observed in the six patients who underwent postmortem examination are summarized in tables 1 and 3. Marked cardiomegaly was present in all cases. The four recorded left atrial volumes ranged from 1,200 to 2,500 ml. Mitral regurgitation was considered the predominant lesion in each case, although slight to moderate mitral stenosis was also present in all cases. Calcification of the mitral valve was present in all cases, and calcification of the left atrial endocardium near the mitral valve caused by a regurgitant jet was extensive in two patients (E.D. and J.L.). The wall of the left atrium was grossly normal in thickness in five cases but was quite thin, measuring 1 mm. in one (M.D.). No intra-atrial thrombi were present.

Marked compression of adjacent thoracic structures by the enlarged left atrium was evident. Atelectasis, caused primarily by direct

Table 2

<table>
<thead>
<tr>
<th>Patient</th>
<th>Predicted blood volume,</th>
<th>Measured blood volume,</th>
<th>Percent predicted</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>liters</td>
<td>liters</td>
<td></td>
</tr>
<tr>
<td>I.C.</td>
<td>3.8</td>
<td>6.5</td>
<td>171</td>
</tr>
<tr>
<td>G.H.</td>
<td>3.3</td>
<td>4.8</td>
<td>145</td>
</tr>
<tr>
<td>J.L.</td>
<td>2.9</td>
<td>7.4</td>
<td>255</td>
</tr>
<tr>
<td>A.S.</td>
<td>5.9</td>
<td>9.1</td>
<td>154</td>
</tr>
<tr>
<td>J.S.</td>
<td>3.6</td>
<td>6.6</td>
<td>183</td>
</tr>
</tbody>
</table>

* Taken as 9 per cent of body weight in kilograms.

Table 3

<table>
<thead>
<tr>
<th>Patient</th>
<th>Heart weight, Gm.</th>
<th>LA volume, ml.</th>
<th>RA volume, ml.</th>
<th>RV/LV thickness, mm.</th>
</tr>
</thead>
<tbody>
<tr>
<td>E.D.</td>
<td>1350</td>
<td>2500</td>
<td>250</td>
<td>4/13</td>
</tr>
<tr>
<td>M.D.</td>
<td>850</td>
<td>1760</td>
<td>660</td>
<td>9/20</td>
</tr>
<tr>
<td>M.C.</td>
<td>600</td>
<td>1500</td>
<td>—</td>
<td>4/20</td>
</tr>
<tr>
<td>V.K.</td>
<td>620</td>
<td>1200</td>
<td>250</td>
<td>5/20</td>
</tr>
<tr>
<td>J.L.</td>
<td>650</td>
<td>1500 (estimated)</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>G.H.</td>
<td>500</td>
<td>1500</td>
<td>—</td>
<td>—</td>
</tr>
</tbody>
</table>
compression of the adjacent lung, was noted in all instances, and, in addition, varying degrees of indentation and displacement of the trachea, main-stem bronchi, and descending aorta were present. The esophagus was severely displaced dorsally in all cases.

Microscopic examination of the lungs, liver, spleen, and kidneys revealed chronic passive congestion. Small renal infarcts were noted in three patients, and a splenic infarct in one.

Histologic examination of the wall of the left atrium was made in three cases. The left atrial myocardium was almost completely replaced by loose fibrous tissue in each instance, and the few remaining muscle fibers were hypertrophic (fig. 4). It is evident that the myocardium of the right atrial wall is hypertrophic, whereas that of the left is virtually absent and has been largely replaced by fibrous tissue.

Discussion

The most striking feature of these 10 patients was their ability to survive for years with this extraordinary degree of cardiac enlargement, accounted for primarily by the massively dilated left atrium. Normally, the adult left atrium contains approximately 140 ml. of blood. The most capacious left atrium ever described held 3 liters and was reported in 1904 by Minkowski in a patient with mitral stenosis and insufficiency. The left atrium of our patient E.D., with a capacity of 2.5 liters, is the second largest we have found in the literature, being equivalent in volume to that of the case reported by Müller in 1905.

The reason for the rightward enlargement of the left atrium becomes apparent when the anatomy of that chamber is viewed. Lying normally just under the tracheal bifurcation, the left atrium is the most superior and the most posterior of the cardiac chambers. Anteriorly it is partially bounded by the great vessels arising from the base of the heart. To the left, anteriorly and inferiorly, is the left

**Figure 4**

Microscopic sections of the walls of the right atrium (A) and left atrium (B) of patient J.L. P., pericardium; M., myocardium; E., endocardium. The pericardium of the left atrial wall is thickened, and the myocardium has been largely replaced with fibrous tissue.
extreme left atrial enlargement

The etiology of the right-sided chest pain prominently mentioned in the literature and present in nine of our patients is not entirely clear, although there are probably multiple contributing factors. It seems most logical to attribute it to the pressure exerted by the enlarged atrium against the parietal pleura and right chest wall. Relief of the discomfort by changing position, such as leaning forward, tends to support this thesis. Discomfort may be minimal to absent, however, even with extreme degrees of atrial enlargement. Liver engorgement may contribute to the pain, although it was not possible to correlate the presence or the severity of chest pain with hepatomegaly and, conversely, most patients with hepatic congestion do not exhibit this type of pain. Relief of the chest pain with diuresis, as shown in two of our patients, might be better related to diminished hepatic congestion than to a decrease in atrial size.

Aggravation of the pain with exertion in four patients suggests an anginal component. Daley and Franks describe a patient whose pain was located in the right shoulder and arm, was precipitated by exertion and breathing 10 per cent oxygen, and was relieved by nitroglycerin and rest. They and others have suggested that this may be "atrial angina," although it is possible that it may represent angina produced by ventricular ischemia and referred to the point of contact of the left atrium with the thoracic wall.

The esophagus, which is in direct apposition to the left atrium posteriorly, is markedly displaced dorsally. The reason for dysphagia in four of our patients is hence understandable. Indeed, in view of the extreme angulation of the esophagus demonstrated fluoroscopically and at postmortem examination, it is remarkable that this symptom was not more frequent or severe. Occasionally dysphagia has been marked, as in the case reported by Nichols and Ostrum of a woman who was subjected to a feeding gastrostomy because of dysphagia and the suspicion of an esophageal neoplasm but at autopsy showed merely esophageal compression from a massively enlarged left atrium.

Displacement of the adjacent bronchi with widening of the carinal angle is the rule, and in rare instances actual bronchial obstruction with atelectasis has been reported. More frequently atelectasis results from direct compression of the lungs by the enlarged heart. Respiratory function may be further impaired by the decrease in vital capacity produced by the extreme cardiomegaly. In most instances the enlarged left atrium produces on physical examination signs that are almost indistinguishable from those of a right pleural effusion. The mistaken impression of a right pleural effusion has frequently led to atrial puncture at thoracentesis, as occurred in Owen's and Fenton's original case and in two of our patients. Similarly, the proximity of the left atrium to the chest wall may produce detectable systolic pulsations of the right thorax.

Despite the apposition of the enlarged atrium to the spine and rib cage, actual bony erosion is rare and has been recorded in only two patients. Ashworth and Jones reported a woman with marked pain in the right chest and axilla whose enlarged left atrium had eroded the fifth through ninth thoracic vertebrae. Similarly, one of Daley's and Franks' patients had erosion of the spine. None of our patients demonstrated this.

In all likelihood multiple factors contribute to the pathogenesis of this unusual entity. The features that seemed common to our patients were (1) a severe initial attack of rheumatic fever often with frequent recurrences (one case excepted), (2) the onset of atrial fibrillation at a relatively early age and continuing...
for many years, and (3) mitral regurgitation as the predominant valvular lesion, with a lesser degree of coincident mitral stenosis in most.

Rarely, mitral stenosis may produce left atrial enlargement to the degree found in this study. Kent and others reported 14 patients with "giant left atria," and mitral stenosis was the predominant lesion in five of these. It is not apparent from the report, and it seems unlikely, however, that the atria were enlarged to the extreme size of those in this study. Kent found no correlation between left atrial mean pressure and chamber size.

Despite the enormous size of these hearts, many of which seem to occupy fully 50 to 75 per cent of the thoracic cavity, the patients often do remarkably well. This may be due in part to the infrequent occurrence of significant pulmonary hypertension, an observation which others have cited and one that was confirmed in the only two of our patients who had cardiac catheterization.

Another interesting facet of the medical peculiarities of these individuals was the striking increase in blood volume in the five patients in whom it was measured. It seems logical to assume that this increase was due in large part to the reservoir of blood in the heart.

Systemic embolism is rare. There were only two minor embolic episodes in our series. A likely explanation is the predominance of mitral insufficiency, which produces enough turbulence in the huge atrial reservoir to prevent thrombus formation.

Occasionally, the right atrium may simultaneously achieve a large size, though the enlargement is never as great as that of the left atrium. This condition of extreme bilateral atriosepy has been reviewed recently by Rogers and Wittels. Clinically, except for the uniform presence of tricuspid insufficiency, these patients differ very little from those with extreme enlargement of only the left atrium.

Increase in left atrial size may be present in association with many other cardiac diseases, such as hypertension, aortic valvular disease, congestive heart failure from any cause, complete heart block, and a variety of congenital defects. Atrial enlargement in these conditions, however, is usually not of striking proportions.

Although extreme enlargement of the left atrium appears to be a rare occurrence in the course of rheumatic heart disease, it is likely that this unusual condition will be seen more frequently in the future as improvements in medical and surgical therapy prolong the lives of seriously ill cardiac patients.

Summary and Conclusions

Extreme left atrial enlargement to the right chest wall is a rare manifestation of chronic rheumatic mitral disease. The clinical and roentgenologic features of 10 cases observed over the past 30 years are presented, with the pathologic findings in six. These patients often survive in fair health for many years in spite of the enormous heart size. Rheumatic mitral insufficiency was the predominant lesion in all instances, with slight to moderate mitral stenosis also present.

Some of the characteristic manifestations of this condition include right-sided chest pain, dysphagia, signs simulating a right pleural effusion, and a marked increase in blood volume. Systemic embolism and significant pulmonary hypertension are infrequent. Possible etiologic mechanisms are discussed.

References


The Medical Profession: Human Understanding and Peace

Because of the nature of our profession, born to care for the health of men, of all men, protecting them against misfortune, we cannot remain coldly indifferent, like mere spectators, to the visible menace of our age.

Each day the world sickens more with distrust and fear, bringing us at times to the verge of despair. It falls to us to play our part to uproot these evils, to serve the cause of human understanding, to fight to ensure peace; that peace which is the slogan of no man because it is the banner of all men whatever their country, their colour, or their religion may be; that peace sought by him who follows Buddha, by him who believes in the God of Mahomet, and by him who prays to Christ; that peace to which all men of goodwill on the earth aspire.—Dr. Ignacio Chávez. Speech delivered at the Inaugural Ceremony of the IV World Congress of Cardiology. Universidad Nacional Autónoma De México, México, D.F., 1962, p. 10.