Acquired Pulmonary Artery Stenosis

Report of a Case Caused by Mediastinal Tumor

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SUMMARY

An acquired stenosis of pulmonary artery caused by benign cystic teratoma in a 22-year-old female is described. The pulmonary artery stenosis was manifested by the precordial systolic murmurs, distinct pulmonary artery-right ventricular pressure gradient, loud systolic murmurs in the pulmonary artery, and the characteristic picture of compression of the artery evidenced by pulmonary arteriogram. The removal of the tumor resulted in the acquired pulmonary regurgitation of significant degree. Short review of the literature was made and the complexity of the cardiac murmur was discussed.

Additional Indexing Words:
Mediastinal teratoma Bilateral stenoses of pulmonary artery Intracardiac phonocardiography Acquired pulmonary regurgitation

THOUGH increasing numbers of reports of acquired pulmonary artery stenosis have appeared, the extrinsic obstructive lesion caused by mediastinal tumor seems to be a rare phenomenon.11-12) This is a case report of such an instance, in which the hemodynamic, intracardiac phonocardiographic, angiocardiographic and surgical explorations are performed. Postoperative finding including an acquired pulmonary regurgitation is also demonstrated.

CASE REPORT

T. W., a 22-year-old female, was first admitted to the Second Department of Internal Medicine, Tokyo University Hospital on March 16, 1967 with recent complaints of oppression of anterior chest and slight cough. She has been diagnosed as having valvular disease since last year because of the loud heart murmur which was first noticed at the age of 12. However, she had no other cardiovascular symptoms, and the birth, growth, development and physical activity were normal. Her past
history was otherwise non-contributory, and the family history revealed nothing abnormal.

*Physical examination* revealed a relatively thin but healthy looking woman (height, 146.3 cm.; weight, 40 Kg.), slightly hoarse and coughing very intermittently. Pulse was regular and the rate was 104/min. Blood pressure was 120/74 mm.Hg. Her color was good and there was no cyanosis, no clubbing of the fingers or toes, no peripheral venous distension and no enlarged glands in the neck. Jugular venous pulsation appeared normal, the “a” wave being dominant. The apical impulse was poor in intensity and was not displaced. On the other hand, a distinct parasternal impulse was felt in the 2nd and 3rd left interspaces laterally, where a systolic ejection murmur of grade 3/6 intensity was heard. No definite ejection sound was heard. The first heart sound had no abnormality. The second heart sound was loud and split during inspiration and the pulmonic component (IIIP) showed no accentuation. The third heart sound was heard within the apex with a quasi-musical mid-systolic ejection murmur of grade 2/6. Another heart murmur, which was recognized after recording of the phonocardiogram, was present along the upper left sternal border, and this was also an ejection murmur with delayed accentuation. Decreased breath sounds and vocal fremitus and dullness with tympanitic resonance were present over the left lateral portion of the chest. There were no abnormal findings in the abdomen and others.

*Laboratory data* disclosed nothing abnormal including urinalysis, blood counting, biochemical as well as various serological studies.

The chest X-ray film (Fig. 1A) showed a homogeneous density of sharp delineation in the left upper lobe, which was initially considered to be marked dilatation of
main pulmonary artery. Tomography revealed that this shadow is continuing from the cardiac shadow and is located anteriorly in the mediastinum. The electrocardiogram (Fig. 2A) showed marked right axis deviation, leftward shift of the transitional zone, and small R wave with deep S in the left precordial leads, all of which are indicative of right ventricular hypertrophy. The phonocardiogram (Fig. 3) revealed the 2

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![Fig. 2. Electrocardiograms. A: preoperative, B: postoperative (5 months later).](image)

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![Fig. 3. Preoperative phonocardiograms and precordial low-frequency tracings (mechanocardiograms) simultaneously recorded. Note the difference of the configuration of the 2 systolic murmurs.](image)
kinds of basal ejection systolic murmur separately, one of which had definitely delayed accentuation. The IIP was much less than the IIA (aortic component). Neither abnormal ejection sound nor atrial sound was recorded. The precordial displacement cardiograms revealed a distinct heave, which was maximally recorded outside the Erb's point. The functional test using methoxamine did not elicit any other cardiac murmurs.

At this point, the tentative or provisional diagnosis was mild to moderate pulmonary artery stenosis due to the compression of extracardiac mass. The absence of either pulmonic ejection sound or accentuation of IIP (and also the absence of regurgitant diastolic murmur) made it difficult to suspect the huge dilatation of pulmonary artery. On the similar bases, valvular pulmonic stenosis was also denied.

Fig. 4. Lung scan. A: preoperative, B: postoperative (20 days later).

Fig. 5. Pressure tracings in the pulmonary artery (PA) and the right ventricle (RV).
Fig. 6. Intracardiac phonocardiograms. RPA = right pulmonary artery, LPA = left PA, RV = right ventricle, 3L = 3rd left interspace on the precordium. L = low, M = medium and H = high frequency PCG.
She underwent the pulmonary function test which showed decreased vital capacity (1.62 L; 56% predicted normal). There was no evidence of an obstructive disturbance and the analysis of arterial blood revealed no abnormality. The lung scan, using $^{131}$I labeled macroaggregates of serum albumin ($^{131}$IMA), showed no visualization of the left lung, indicating virtually no pulmonary arterial flow to the left lung (Fig. 4A).

The right heart catheterization revealed mild elevation of right ventricular systolic pressure (RV=36/-2-0 mm.Hg) and distinct pressure gradient in the pulmonary artery (PA=24/7, mean 13 mm.Hg) (Fig. 5). However, the exact or localized site of the stenosis was not clarified with confidence, partly because of the bout of premature beat on the pullback tracing. Intracardiac phonocardiography revealed the intense systolic murmur within the right pulmonary artery and this was slightly attenuated in the main pulmonary artery and finally disappeared in the right ventricle (Fig. 6). The left pulmonary artery was thought to be severely stenosed, because the insertion of the catheter very easily provoked the artificial friction sounds.

The pulmonary arteriography demonstrated stenosis and compression of the main pulmonary artery from above and almost complete occlusion of the left pulmonary artery (Fig. 7). The thoracic and abdominal aortography demonstrated no abnormality and the mediastinal lesion was separate from the aorta.

She was transferred to the Department of Thoracic Surgery to remove the mediastinal tumor. *At thoracotomy,* a large tumor with tense capsule was found, which was firmly adherent to the pericardium, pleurae, and the other structures including pulmonary artery. The entire tumor was removed with difficulty and the pericardium over the pulmonary conus was resected (approximately $6 \times 7$ cm. in

![Fig. 7. Pulmonary arteriogram demonstrating the compression of pulmonary artery.](image)

*The operation was performed by Prof. Seiji Kimoto, director of the Department of Thoracic Surgery, University of Tokyo, on September 22, 1967.*
The thrill felt preoperatively on the pulmonary artery disappeared after operation. The tumor was benign cystic teratoma by histological examination (Fig. 8).

Postoperative course: The patient was doing well postoperatively and the complaints were abolished. The X-ray of the chest (Fig. 1B) revealed the complete removal of the tumor. The electrocardiogram (Fig. 2B) and lung scan (Fig. 4B) were normalized. However, auscultation of the precordium 1 week after thoracotomy disclosed a loud basal diastolic murmur which had extreme intensification during post-inspiratory phase. The murmur had medium frequency and moderate duration. The held expiration tended to abolish the murmur. On the other hand, the systolic murmur decreased in intensity (Fig. 9). Intracardiac phonocardiography

![Fig. 8. Incised teratoma.](image1)

![Fig. 9. Phonocardiograms taken postoperatively.](image2)
performed 4 months later demonstrated an only residual systolic murmur in the pulmonary artery and a loud diastolic regurgitant murmur in the outflow tract of the right ventricle (Fig. 10). An atriosystolic murmur of brief duration was also recorded on the precordium and this was most clearly recorded in the inflow tract of the right ventricle. No tricuspid regurgitant murmur was present in the tricuspid valve area. Cineangiocardiography also revealed the presence of the pulmonary regurgitation probably due to the dilatation of the pulmonary artery including the valve ring.

**COMMENT**

The space-filling lesions in the mediastinum may result in the cardiovascular signs and symptoms which are usually manifested by the obstruction of superior caval vein. On the other hand, few cases were reported, in which the pulmonary artery was compressed and the loud heart murmur was developed. Such an acquired pulmonary stenosis may be caused by, in addition to mediastinal tumors, lung cancer, aortic aneurysm, pulmonary tuberculosis, rheumatic pericarditis, annular constrictive pericarditis, calcified lymphnodes, and so on. However, all of these possibilities are equally rare. Of these, mediastinal tumor has been a matter of interest, because of the considerable simulation to the congenital heart disease in respect to history, physical examination and routine laboratory examinations. A typical example was illustrated by Winter, and Shaver et al. collected 7
Table I. Survey of Cases with Acquired Pulmonary Artery Stenosis Produced by Mediastinal Tumor

<table>
<thead>
<tr>
<th>Authors</th>
<th>Age</th>
<th>Sex</th>
<th>Symptoms</th>
<th>Auscultation</th>
<th>Hemodynamics</th>
<th>Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Rusby</td>
<td>23</td>
<td>M</td>
<td>chest pain, palpitation, dyspnea, cough</td>
<td>SM in 2L</td>
<td>RV syst. 40–50</td>
<td>Hodgkin's disease</td>
</tr>
<tr>
<td>2. Maier</td>
<td>23</td>
<td>F</td>
<td>occasional dyspnea, fatigue on moderate exertion</td>
<td>harsh SM over entire precordium</td>
<td>RV syst. 45, PA 18/12</td>
<td>Hodgkin's disease</td>
</tr>
<tr>
<td>3. Fry et al.</td>
<td>23</td>
<td>M</td>
<td>precordial pain radiating to left arm and shoulder, increased fatigability</td>
<td>harsh, low-pitched, pulmonic SM, palpable thrill</td>
<td>RV syst. 40–50, PA 18/12</td>
<td>Hodgkin's disease</td>
</tr>
<tr>
<td>4. Wood</td>
<td>23</td>
<td>M</td>
<td>chest pain, cough</td>
<td>typical pulmonic SM with thrill</td>
<td>RV syst. 45, PA 18/12</td>
<td>Hodgkin's disease</td>
</tr>
<tr>
<td>5. Waldhausen et al</td>
<td>23</td>
<td>M</td>
<td>chest pain, cough</td>
<td>$^{3/4}$ SM in 2L and 3L, widely transmitted</td>
<td>RV syst. 45, PA 18/12</td>
<td>Hodgkin's disease</td>
</tr>
<tr>
<td>6. Winter</td>
<td>21</td>
<td>M</td>
<td>chest pain, dyspnea</td>
<td>$^{3/4}$ harsh ejection SM, transmitted into neck and back</td>
<td>RV syst. 45, PA 18/12</td>
<td>Hodgkin's disease</td>
</tr>
<tr>
<td>7. Babcock et al.</td>
<td>15</td>
<td>F</td>
<td>asymptomatic</td>
<td>$^{3/4}$ SM in 2L and 3L, widely transmitted</td>
<td>RV syst. 45, PA 18/12</td>
<td>Hodgkin's disease</td>
</tr>
<tr>
<td>8. Shaver et al.</td>
<td>27</td>
<td>F</td>
<td>chest pain, dyspnea</td>
<td>$^{3/4}$ to $^{1/2}$ ejection SM in 2L-3L</td>
<td>RV syst. 45, PA 18/12</td>
<td>Hodgkin's disease</td>
</tr>
<tr>
<td>9. Gough et al.</td>
<td>15</td>
<td>M</td>
<td>asymptomatic</td>
<td>loud pulmonic ejection SM and thrill, single II S</td>
<td>RV syst. 45, PA 18/12</td>
<td>Hodgkin's disease</td>
</tr>
<tr>
<td>10. ibid.</td>
<td>22</td>
<td>F</td>
<td>exertional dyspnea, chest pain, dull aching in the upper chest</td>
<td>moderate pulmonic ejection SM</td>
<td>RV syst. 45, PA 18/12</td>
<td>Hodgkin's disease</td>
</tr>
<tr>
<td>12. Ueba et al.</td>
<td>20</td>
<td>F</td>
<td>exertional dyspnea</td>
<td>basal SM</td>
<td>RV syst. 45, PA 18/12</td>
<td>Hodgkin's disease</td>
</tr>
<tr>
<td>13. Ogawa et al.</td>
<td>16</td>
<td>F</td>
<td>general malaise, exertional dyspnea</td>
<td>$^{3/4}$ to $^{1/3}$ SM along LSB</td>
<td>RV syst. 45, PA 18/12</td>
<td>Hodgkin's disease</td>
</tr>
<tr>
<td>14. Present Authors</td>
<td>22</td>
<td>F</td>
<td>chest oppression, slight cough</td>
<td>$^{3/4}$ ejection SM in 2L-3L laterally, delayed ejection SM in 2L</td>
<td>RV syst. 45, PA 18/12</td>
<td>Hodgkin's disease</td>
</tr>
</tbody>
</table>

SM = systolic murmur, RV = right ventricle, PA = pulmonary artery (M : main, R : right, L : left), Gr. = RV-PA pressure gradient, CO = cardiac output. Pressures are expressed by mm.Hg.
similar cases from the literature. Review of the 14 reported cases (Table I) revealed that teratoma (7 cases) and thymoma (4 cases) are the frequent causes, and most of the patients belonged to young age and the sex distribution was predominant in female. These are simple but important differential diagnostic clue to the other causes of pulmonary artery compression, such as lung cancer or aortic aneurysm.

The compression of pulmonary artery by mediastinal tumor is characterized by systolic murmur masquerading as organic heart disease. This was first described by Rusby, but the correct interpretation was attempted by Fry et al. The phonocardiogram was presented in several reports, however, it may be noted that the precordial murmur may not be simple, but may be composed of 2 or more different murmurs as shown in the present case. The intracardiac phonocardiogram revealed different type of ejection systolic murmurs at the various sites of pulmonary artery, and this is not the case of simple congenital heart disease. Though the different murmurs were discovered in the present case, the sites of maximal audibility of these 2 murmurs were in the quite vicinity each other. Careful auscultation is, therefore, recommended in doubtful cases.

Pulmonary regurgitation following the removal of the tumor was not anticipated and it has unexplained cause. The most probable explanation is the damage of the pulmonary valve ring secondary to the surgical procedure. The absence of the pericardium over the outflow tract of right ventricle and also the already thinned pulmonary arterial wall are both contributory factors, but the exact cause is still unknown.

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REFERENCES

Notice of the Journal of Electrocardiology

The first issue of the Journal of Electrocardiology, a quarterly interdisciplinary journal devoted to the study of electricity produced by the heart or applied to it, was published in April, 1968. It contains 15 papers including Editorials, Experimental and Clinical Studies, an Instrumentation Section edited by L. A. Geddes, Ph.D. and L. E. Baker, Ph.D. of Baylor University and R. Ware, Ph.D. of the Southwest Research Institute, an ECG-Pathological Section edited by R. C. Scott, M.D. of the Cincinnati General Hospital and a Review Section edited by Dr. P. Kezdi. The Editors are Dr. Z. Z. Zao of the Cox Coronary Heart Institute, Kettering, Ohio, and Dr. E. Lepeschkin of the University of Vermont, Burlington, with Dr. P. Kezdi of the Cox Institute and Dr. L. Werkö of Stockholm as Co-Editors. The Editorial Board includes 49 prominent scientists and clinicians from the U.S.A. and 35 from abroad. The second issue, which will contain papers and also abstracts of electrocardiological papers hitherto scattered among medical, surgical, physiological, pharmacological, biophysical and engineering journals selected by over 40 Abstract Editors, is in preparation. Information concerning manuscripts and subscriptions can be obtained from the Editorial Office, Box 2225, Kettering, Ohio 45429.