Case Reports

A Case of Anomalous Inferior Vena Cava with Azygos Continuation Associated with Situs Inversus Totalis and Acquired Mitral Valvular Disease

Hirokuni Beppu, M.D., Satoru Matsushita, M.D., Shin-ichi Kimata, M.D., and Kazuhiko Murata, M.D.

ANOMALOUS inferior vena cava with azygos or hemiazygos continuation has been considered to be a very rare anomaly in the past. However, since the development of cardiac catheterization and angiocardiography, this congenital abnormality has been reported with increasing frequency. This anomaly is usually associated with other cardiovascular defect, and there are few reported cases without other malformation of the heart. The following is a description of a case of anomalous inferior vena cava with hemiazygos continuation associated with situs inversus totalis, acquired mitral valvular disease, orthostatic proteinuria and no other demonstrable congenital cardiac malformation.

CASE REPORT

A 18-year-old girl was admitted to the hospital for the evaluation of cardiac status. The patient has been told to have a dextrocardia since childhood. An enlargement of the heart was first noticed at an annual check-up when she was 15 years old. She had been well until February 1963, when cough, dyspnea and orthopnea developed. She visited a hospital where a diagnosis of mitral valvular disease was established. The patient was referred to our cardiac unit on April 12, 1963. At that time, chest X-ray films demonstrated dextrocardia, an enlargement of the left atrium and a slight pulmonary vascular congestion. A grade 3 apical systolic murmur and a grade 1 to 2 apical diastolic murmur were audible. The urine gave a + to ++ test for protein. The patient was given digitoxin, and she had been without symptoms thereafter except for intermittent bouts of dyspnea. She was admitted to our department for a check-up of the cardiac condition and albuminuria on September 21, 1965.

Physical examination revealed a well-developed, well nourished girl without apparent discomfort. There were no signs of cyanosis, clubbing or dyspnea. The
temperature was 36.7°C, the pulse 67 and the respirations 18. The blood pressure was 114/56 mmHg. There was no chest deformity. The cardiac dullness was not enlarged. The apex beat was felt in the fourth interspace at the midclavicular line. A systolic thrill was felt near the apex. The second sound was split and accentuated at the second right interspace. An opening snap was audible. There was a grade 3 harsh systolic murmur best heard at the right sternal border in the second interspace. A grade 3 to 4 systolic murmur and a grade 3 diastolic rumble with presystolic accentuation were present at the apex. A phonocardiogram confirmed the above auscultatory findings (Fig. 1). There were no venous hum, jugular venous pulsation or goiter. The lungs were clear and the liver was not palpable. There was no venous dilatation or peripheral edema.

Chest X-ray film showed dextrocardia with slight enlargement of the transverse diameter of the heart and protrusion of the left atrium. There was an unusual prominence of the left upper part of the cardiac silhouette which was proved to represent the dilated, anterior-coursing azygos vein by angiocardiography (Fig. 2). The electrocardiogram was consistent with dextrocardia. The PR interval was slightly prolonged to 0.24 sec. (Fig. 3). The urine gave a ++ test for protein on the first day of admission, but negative on and after the second hospital day. The sediment contained one white cell per 3 to 10 highpower fields but no red cells. The hematocrit was 31% and the red cell count was $382 \times 10^4$. The white cell count was 6,100, with 77.5% neutrophils and 22.5% lymphocytes. Other laboratory examinations were normal. The kidney function tests were within normal limits. A radiological examination of the gastrointestinal tract was not remarkable except for situs inversus totalis.

Cardiac catheterization was first attempted via the right saphenous vein. The catheter was advanced from azygos vein through the superior vena cava into the right atrium. (Fig. 4) It was possible to advance the catheter tip in the liver, but
Fig. 3. Electrocardiogram.

Fig. 4. A catheter was advanced from azygos vein through the superior vena cava into the right atrium. Right atrium, right ventricle and pulmonary artery were opacified by contrast medium injected from the catheter.

Table I. Commonly Associated Anomaly of Anomalous Inferior Vena Cava with Azygos or Hemiazigos Continuation

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Persistence of left superior vena cava</td>
<td>24</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>23</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>19</td>
</tr>
<tr>
<td>Transposition of great vessels</td>
<td>19</td>
</tr>
<tr>
<td>Pulmonary stenosis or atresia</td>
<td>17</td>
</tr>
<tr>
<td>Dextrocardia</td>
<td>13</td>
</tr>
<tr>
<td>Anomalous pulmonary venous return</td>
<td>13</td>
</tr>
<tr>
<td>Single atrium</td>
<td>10</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>9</td>
</tr>
<tr>
<td>Cor biloculare</td>
<td>8</td>
</tr>
<tr>
<td>Persistent atrioventricularis communis</td>
<td>5</td>
</tr>
<tr>
<td>Cases reported as an isolated anomaly</td>
<td>7</td>
</tr>
</tbody>
</table>

(Total: 75 cases)
the right ventricle could not be entered. The catheterization study via the right arm vein revealed a right ventricular pressure of 40/0 mm.Hg. The catheter could not be advanced into the pulmonary artery. No step up of oxygen content was demonstrated up to the ventricular level. Oxygen saturation of the femoral artery blood was 96.2% and cardiac index was 2.53 L./min./M.²

Angiocardiography revealed a great vessel, which ascended along the right side of the spine, crossed it at the level of the diaphragm, ascended behind the heart and then emptied into the superior vena cava (Fig. 5). A lateral angiogram showed a typical “candy cane” appearance of the anomalous vein (Fig. 6). During the course of her admission she was asymptomatic and was discharged on October 16, 1965.

COMMENT

The anomalies of the systemic venous system has not been well recognized until recent development of cardiac catheterization and angiocardiography. The most frequent anomaly of the systemic venous system is persistent left superior vena cava which is reported to be observed in 3 to 10% of the catheterized patients.1),3) The reported incidence of anomalous inferior vena cava with azygos or hemiazygos continuation, on the other hand, is between 0.2 to 1.0% of the cases with congenital heart disease.²,4) There are 75 reported cases of this anomaly,¹,3,4¹⁻⁹ including 13 cases in Japan⁵⁻⁹ up to the present. The incidence of this anomaly in general population is not known. Adachi¹⁰ found no cases in 1,055 autopsies. The most probable explanation for the development of this anomaly is the failure of union of the hepatic and subcardinal segment of the inferior vena cava.¹¹ Sixty-eight of 75 reported cases were associated with other congenital abnormality of the cardiovascular
system. As shown in Table I, the commonly associated anomalies are persistent left superior vena cava, atrial septal defect, ventricular septal defect, transposition of great vessels, pulmonary stenosis or atresia, dextrocardia, and anomalous pulmonary venous return. Abnormal position of the heart is not infrequently observed. The associated cardiac anomaly was not described in 7 of 75 cases.

Although mirror image dextrocardia with situs inversus was present in the present case, no other congenital anomaly of the heart was demonstrated. The mitral valvular disease was considered to be of acquired origin. However, a possibility of ventricular septal defect or pulmonary stenosis could not be entirely excluded because of the presence of a harsh systolic murmur at the base, although the systolic pressure of the right ventricle was not remarkably elevated. An increased pulmonary vascularity is another finding against pulmonary stenosis. Unfortunately, a catheter could not be advanced into the pulmonary artery.

As far as the orthostatic proteinuria is concerned, it may be postulated that an abnormal course of renal vein is a contributing factor, but renal venography was not attempted.

**Summary**

A case of anomalous inferior vena cava with azygos continuation associated with mirror image dextrocardia, situs inversus and acquired mitral valvular disease is reported. The anomaly of the inferior vena cava was incidentally discovered during cardiac catheterization. No other congenital anomaly of the cardiovascular system was demonstrated.

**Acknowledgment**

The authors are grateful to Associate Prof. Akira Tasaka at the Radiology Department for angiocardiography. Thanks are also due to Prof. Kiku Nakao and Associate Prof. Masao Ikeda for reviewing the manuscript.

**References**