Isolated Levocardia Associated with Absence of Inferior Vena Cava, Lobulated Spleen and Sick Sinus Syndrome

A Case Report

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SUMMARY

We evaluated a 68-year-old male patient with isolated levocardia without intracardiac anomaly. The patient’s condition was complicated by the absence of the inferior vena cava, a lobulated spleen and sick sinus syndrome. Isolated levocardia without intracardiac anomaly is very rare and only 25 cases of this disease have been reported, to our knowledge. In general, it is accepted that cardiac rhythm disorder is frequently observed in cases of isolated levocardia and/or absence of inferior vena cava. However, there are few cases of isolated levocardia without intracardiac anomaly complicated by the absence of the inferior vena cava, a lobulated spleen and apparent sick sinus syndrome. (Jpn Heart J 1998; 39: 235–241)

Key words: Isolated levocardia, Sick sinus syndrome, Pacemaker

Isolated levocardia is an extremely rare congenital heart disease. Its incidence is reported to be about 0.4–1.2% of all congenital heart disease.1,2) Most cases of isolated levocardia are frequently associated with intracardiac anomalies. Only 25 cases of isolated levocardia without intracardiac anomalies have been reported.3,4) The malformation responsible for isolated levocardia is frequently complicated by cardiac rhythm disorder. Out of 25 cases, only 2 cases associated with apparent sick sinus syndrome have been reported.1,5) In this report, we present a rare case of isolated levocardia without intracardiac anomaly, which was complicated by sick sinus syndrome, absence of the inferior vena cava and a lobulated spleen.

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CASE REPORT

A 68-year-old man was admitted to our hospital because of an abnormal electrocardiogram (ECG). The patient's family history includes an aunt with visceral inversion and dextrocardia. The patient had an appendectomy in 1968, with his appendix found in the left lower abdominal cavity. He was diagnosed as having diabetes mellitus in 1982. Two years prior to his current admission the patient was found to have an arrhythmia, but was not treated because of a lack of symptoms. In December 1991, during preoperative tests prior to cataract surgery, the patient was found to have an arrhythmia and was referred to our hospital for further examination.

The patient's general health was good with a blood pressure of 116/60 mm Hg and a pulse rate of 42 beats/min and regular. No abnormalities were detected on auscultation of the heart and lungs. The liver and spleen were not palpable, and edema of the extremities was not observed. Howell-Jolly bodies were not seen in peripheral blood smears. Except for elevation of the fasting blood sugar (198 mg/dl), biochemical examination revealed no abnormal findings. Chest X-ray photogram (Figure 1) showed a cardiothoracic ratio of 49%, and the cardiac shadow was located on the left side. An azygos knob (arrow) was observed on the right margin of the cardiac silhouette. A chest computed tomograph demonstrated normal tracheal anatomy. ECG showed sinus bradycardia of 42/min. In leads II, III, aVF and V1, T waves showed low voltage and were flat or inverted (Figure 2-a). On Holter ECG, paroxysmal supraventricular tachycardia followed by sinus arrest for 6.12 seconds was recognized (Figure 2-b). Echocardiographic findings showed normal-shaped cardiac ventricles and valves.

Figure 1. Chest X-ray. The cardiac shadow was located on the left side. An azygos knob (arrow) was observed on the right margin of the cardiac silhouette.
Figure 2. Electrocardiographic findings. a) Heart rate was 42/min. (sinus bradycardia). T waves showed low voltage and were flat or inverted in leads of II, III, aVF and V1-4. b) Sinus arrest for 6.12 seconds following paroxysmal supraventricular tachycardia was recognized on Holter ECG recording. c) Sinus node recovery time obtained from overdrive suppression test was apparently prolonged to 3.83 second after rapid pacing at 120/min.

To evaluate whether other cardiac disease was present or not, cardiac catheterization and angiography were performed. The catheter inserted via the right femoral vein reached the superior vena cava, right atrium and right ventricle through the dilated azygos vein. Pressure and oxygen saturation in each cardiac chamber were normal, and intracardiac shunt was absent. As shown in Figure 3, venography from the inferior vena cava revealed azygos continuation of the inferior vena cava. His bundle ECG was attempted to examine the function of the atroventricular (A-V) node. However, we could not direct the catheter tip to the His bundle because of the abnormal position of the inferior vena cava. Sinus node recovery time obtained from the overdrive suppression test was apparently prolonged to 3.83 seconds after rapid pacing at 120/min (Figure 2-c).

Fluorography of the gastrointestinal tract showed inversion of the stomach to the right side and an abnormal location of the intestinal tract. Computed tomography of the abdomen (Figure 4) showed that the hepatic part of the
Figure 3. Venogram from the inferior vena cava. The azygos continuation of the inferior vena cava was revealed. RA = right atrium; SVC = superior vena cava.

Figure 4. Abdominal computed tomograph. Dilated azygos vein (upper panel, ↑) and absence of inferior vena cava at hepatic part (lower panel, ▲) were revealed. The inversion of the stomach (St) and the lobulated spleen (S) to the right side were recognized. L = liver.

inferior vena cava was absent and that the azygos vein was dilated. The spleen was located on the right side and appeared to be lobulated. Morphological abnormality of the liver was also observed. The pancreas could not be clearly identified by means of either computed tomography or echography.

Thus, on the basis of these findings, isolated levocardia complicated by the absence of the inferior vena cava, a lobulated spleen and sick sinus syndrome was
diagnosed. Permanent pacemaker implantation was recommended to the patient. A dual chamber pacemaker was implanted and the patient was discharged in good condition.

**DISCUSSION**

In general, cardiac axis rotation in the cardiac germ and intestinal rotation are considered to occur around the 30th day of embryonic development. Isolated levocardia is thought to occur when there is a discrepancy among the directions of cardiac and intestinal rotation. Most cases of isolated levocardia are associated with intracardiac anomaly, such as transposition of the cardiac cavity, septal defect, transposition of the great vessels, pulmonary arterial constriction, abnormal pulmonary venous drainage and so on.

Anomalous connection of the great cardiac veins is also frequently noted in isolated levocardia. Absence of the inferior vena cava has been reported in about 30.7–58.0% of the cases. Abnormal anastomosis of the inferior vena cava is thought to be the basis for the occurrence of isolated levocardia. As a result, a defect in the hepatic segment and drainage from the superior vena cava through the right supracardinal vein (azygos vein) or left supracardinal vein (hemiazygos vein) to the right atrium is induced. The great cardiac vein is generated as a result of anastomosis of the hepatic, mesenteric, and postrenal segments of the vitelline vein and the cardinal vein. In this process, the symmetry is lost and right sided dominance becomes definite. This developmental period almost coincides with that of intestinal and cardiac axis rotation.

Therefore, isolated levocardia and absence of the inferior vena cava could be explained by some developmental anomaly occurring in almost the same stage of the embryonic period.

Isolated levocardia and absence of the inferior vena cava are known to frequently accompany various rhythm disorders. However, a few cases of isolated levocardia, complicated by an absence of the inferior vena cava and apparent sick sinus syndrome have been reported. Ogihara et al. suggest that because isolated levocardia and/or absence of the inferior vena cava are frequently associated with other intracardiac anomalies, the cause of rhythm disorders may be due to right atrial volume and pressure overload which induces tissue degeneration of the atrial muscle. In our patient, however, no other intracardiac anomalies were observed, and it seems difficult to attribute the rhythm disorder to volume and pressure overload of the right atrium. According to Patten, specially differentiated pacemaker cells are observed symmetrically at the sinus ostium of the bilateral common cardinal veins, and the sinoatrial nodes and A-V nodes are generated from the right and left sides, respectively. In case of
persistent left superior vena cava in which the left side of the great cardiac venous system was dominant. James and his colleagues\(^\text{16}\) demonstrated that the sinus nodes generated from the right pacemaker cells were hypoplastic. In our case, considering these reported facts, it is more probable and reasonable to conclude that the pacemaker cells did not differentiate normally during the process of development, producing hypoplastic sinus nodes, resulting in sick sinus syndrome.

Although a marked period of sinus arrest lasting over 6 seconds was noted in our case, there were no symptoms. Therefore his disease had not been discovered until quite late in spite of the presence of sinus node dysfunction.

The evaluation of the A-V nodal function could not be conducted in our case because of the abnormal location of the inferior vena cava. However, the presence of a dysfunctional A-V node can be anticipated in aged patients, and His bundle ECG, examination should be conducted whenever possible. Thus, the development of a new catheter electrode to examine such anomaly cases is needed.

**REFERENCES**

