A CASE REPORT OF ISOLATED LEVOCARDIA WITHOUT INTRACARDIAC ANOMALIES ASSOCIATED WITH SICK SINUS SYNDROME

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A 42-year-old female with cardiomegaly showed bradycardia without syncope. Clinical data showed that she had an isolated levocardia with interruption of the inferior vena cava. Isolated levocardia was defined as a normally placed heart associated with situs ambiguus of other viscera. She did not have intracardiac anomalies.

Isolated levocardia without intracardiac anomalies, as in this case, has only been reported in 13 other cases. Isolated levocardia is often accompanied by severe complex intracardiac anomalies and, therefore, most of the patients have a short life span.

Situs ambiguus, especially left isomerism, is frequently associated with deteriorated sinus node function, and an interruption of the inferior vena cava may also be an indication of this phenomenon. Therefore, the patient's sinus node function was examined using an electrophysiological study and a 24-hour ambulatory electrocardiogram. Sick sinus syndrome was finally confirmed. (Jpn Circ J 1993; 57: 245–250)

Isolated levocardia is defined as a normally placed heart associated with either total or partial situs inversus or ambiguus of other viscera! Patients suffering from this condition tend to have a short life span because of severe complex intracardiac anomalies. In addition, situs ambiguus, especially left isomerism, is often associated with a disturbance of the cardiac conduction system or automaticity.

Key words:
Isolated levocardia
Sick sinus syndrome
Intracardiac anomaly
Interruption of inferior vena cava continuing to the azygos vein
Permanent pacemaker

This report describes an adult case of isolated levocardia without intracardiac anomalies, and in whom sick sinus syndrome was confirmed.

CASE REPORT

A 42-year-old female was admitted to Asahikawa Medical College Hospital in February 1991 for evaluation of cardiomegaly.

The patient had received a cystectomy for empty sella syndrome in October 1978, and a diagnosis of situs inversus was made by her family physician in 1988. At the time of the operation, the doctor had suspected that
the patient had sick sinus syndrome because the electrocardiogram had shown junctional escaped beats.

The medical history of her family was non-contributory as far as could be ascertained.

On physical examination, the patient had bradycardia (40 beats/min) and normal blood pressure (140/80 mmHg). A grade 2/6 systolic ejection murmur was heard in the second left intercostal space. The stomach was found by percussion at the right costal margin and the liver was palpable 5 cm below the xiphoid process.

An examination of the peripheral blood smear showed neither Howell-Jolly bodies nor Heinz bodies.

An electrocardiogram taken on admission revealed a slow atrial rhythm, at a rate of 42 beats/min, with left axis deviation of P waves (frontal plane P wave vector was $-45$ degrees) and sinus arrest with junctional escaped beats (Fig. 1).

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On the P-A chest X-ray film (Fig. 2), the apex of the heart was in the left of the thorax with a 60% cardio-thoracic ratio and the azygos arch was dilated. A gastric air bubble was also located on the right and the bilateral hyparterial bronchi were observed. A transthoracic two-dimensional echocardiogram showed normal-shaped cardiac ventricles and valves. However, a transesophageal echocardiogram suggested that the auricle of the right-sided atrium had an index finger configuration (morphologically “left atrial” shape) (Fig. 3). Neither a shunt nor regurgitation across the valves were observed by Doppler echocardiography. A bronchogram and chest tomogram disclosed that the right-sided lung was bilobed and the right-sided bronchus was longer, narrower and ran more horizontally than a normal right bronchus. These findings suggested that it was a hyparterial configuration which was morphologically similar to a left bronchus (Fig. 4). An abdominal computed tomogram demonstrated that three spleens were on the right side of the abdomen and the liver was located centrally and overlaid bilaterally (Fig. 5 A, B). A spleen scintigram also revealed multiple spleens on the right of the abdomen (Fig. 5 C). These findings indicated that the patient had left isomerism (bilateral leftsideness) with levocardia.

Cardiac catheterization showed normal pressure and blood oxygen saturation. The contrast angiogram showed that the inferior vena cava was interrupted in the hepatic portion and continued to the azygos vein (Fig. 6).

An overdrive suppression test showed that the sinus node recovery time was prolonged (8.03 sec). Twenty four-hour ambulatory electrocardiograms were recorded repeatedly. A sinus arrest with a duration of 8 seconds was observed (Fig. 7). Consequently a permanent pacemaker was implanted to prevent syncope.

DISCUSSION

If visceral heterotaxia exists, the position of the heart is generally dextroverted. In the present study, various data indicated that this patient had left isomerism associated with polysplenia and the apex of the heart was on the left. Therefore, we defined this case as isolated levocardia.

A case of this type is very rare, with an incidence of 0.88% in 3500 autopsy cases of congenitally malformed hearts. Campbell and Deuchar have reported that the incidence is about 1 in 22,000 of the general population. Isolated levocardia occurs due to a discordance between the heart and the abdominal viscera during embryonic development. Since this discordance of the heart usually leads to severe complex intracardiac anomalies; the prognosis of this disease is poor. As a result, there have been very few reports of isolated levocardia without intracardiac anomalies. Only 13 such cases have been reported to date.

Dickinson et al. have described the cardiac conduction system in situs ambiguus.
They reported that left isomerism tends to have a hypoplastic sinus node located abnormally. As a result, left isomerism frequently has a rhythm disturbance because the right-sided atrium is replaced by an anatomical “left atrium” instead of an anatomical “right atrium” that embryologically includes the sinus node. Momma et al. have also concluded that multiplicity and progressive slowing of the atrial rhythm are characteristic in patients with left isomerism.

 Interruption of the inferior vena cava continuing to the azygos vein is often related to left isomerism. However, cases of this type which did not involve left isomerism have been reported by Osaki et al. and Bharati and Lev. Cases of this type have also been accompanied by deteriorated conduction systems of automaticity.

Imai et al. have reported that isolated levocardia was frequently complicated by various rhythm disturbances, such as coronary sinus rhythm, atrio-ventricular nodal rhythm, left atrial rhythm and wandering pacemaker, and that cases with sick sinus syndrome had not been previously described in medical literature. They reported the first case of isolated levocardia with sick sinus syndrome in Japan. Although we initially believed that our patient had simple congenital anomalies and did not show any subjective symptoms, the search to evaluate the patient’s sinus node function was undertaken exclusively to identify whether or not rhythm disturbances existed. In this manner, sick sinus syndrome was finally confirmed. Some of the previously reported cases involving rhythm disturbances complicated by isolated levocardia may have been sick sinus syndrome.

The present case indicates that isolated levocardia can be identified through chest X-ray by analyzing the abnormality of the bronchial trees and the relationship between the position of the gastric bubble and the apex of the heart. Patients with isolated levocardia should have their sinus node function evaluated.

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**Fig. 6.** Contrast angiogram of the inferior vena cava. The lateral view (A) and anterior view (B) show an interruption of the inferior vena cava continuing to the azygos vein.

△: inferior vena cava

▲: azygos vein

○: superior vena cava

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**Fig. 7.** Twenty four-hour ambulatory electrocardiogram show a sinus arrest with a duration of 8 seconds.

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