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Lipomatous Hypertrophy of the Interatrial Septum With Cutaneous Lipomatosis

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A mass was identified in the atrial septum by clinical imaging in a patient who had multiple large lipomas of the skin. Multislice computed tomography and magnetic resonance imaging indicated that the interatrial mass was a fatty infiltration. In order to discriminate the myocardial lesion from myxoma and other benign conditions and malignant tumors, percutaneous transvenous biopsy was performed. Microscopic examination demonstrated the cardiac mass to be lipomatous hypertrophy of the interatrial septum (LHIS), and the skin lesions, simple lipomas. The LHIS is itself a rare condition, and its development in patients having multiple ponderous lipomas, or lipomatosis, could not be found in a previous report. This may be the first published case of LHIS, which likely represents cardiac involvement of lipomatosis. (*Circ J* 2007; **71:** 986–989)

Key Words: Cardiac tumor; Cutaneous lipoma; Interatrial septum; Lipomatosis; Pathology

Lipomatous hypertrophy of the interatrial septum (LHIS) is a rare benign cardiac lesion characterized by the presence of non-encapsulated mass of the fatty tissue that infiltrates through the atrial septum. A definitive diagnosis is rarely attained during life. Lipomatosis, also a rare condition, is defined as a diffuse overgrowth of mature adipose tissue affecting a large portion of an extremity or portions of the trunk. We report a rare case of LHIS combined with lipomatosis, in which a diagnosis of LHIS was established during life by the use of cardiac imaging including multislice computed tomography (MSCT) and echocardiography, and percutaneous transvenous biopsy of the interatrial septum.

Case Report

A woman aged 27 years was admitted to Ishinomaki Municipal City Hospital for abdominal pain in June 2005. She had urgent surgery under a diagnosis of perforated duodenal ulcer. A cardiac tumor was incidentally found on computed tomography during the admission, and she was referred in July 2005. The family history showed that her mother and brother and sister were obese, but upon echocardiography, each revealed normal cardiac imaging. Physical examination of the present patient disclosed moderate obesity with a body weight of 98.8 kg, height of 166 cm, and a calculated body mass index of 35.9, but she proved not to have any metabolic disorders such as hyperlipidemia or diabetes mellitus. There were 2 soft tissue tumors, one in the left anterior chest wall, slightly elevated and measuring 24×14 cm, and the other in the left arm and presenting as a pendulous mass, approximately 25×19 cm in diameter (Fig 1). In both these sites, the emergence of the tumors had been preceded by faint pigmentation of the skin. The patient did not remember exactly her age when she noticed these lesions, but thinks it was when she was nearly 7 years old. Thereafter the pigmentations continued to become less remarkable, disappearing by the age of 13 when, however, the area began to bulge, slowly growing into masses during the 14 years that followed. In addition, multiple smaller elevations were found crowding around the original tumors. The heart and respiratory sounds were normal. Systemic blood pressure was 120/85 mmHg. A basal ECG showed sinus rhythm, with a heart rate of 65 beats/min; there were P-wave abnormalities consisting of amplitude <1.0 mm in lead Ⅲ, duration >100 ms, and a "notching" configuration (Fig 2). The chest X-ray disclosed mild cardiac enlargement (Fig 3). A large mass in the right atrium extending along the atrial septal wall was disclosed by both transthoracic and transesophageal echocardiography performed on the 2nd day of admission (Fig 4). More detailed information was provided by MSCT scanning, which visualized a tumor, 48 mm in maximal diameter, arising from the atrial septum (Fig 5). The transverse image at the level of the cardiac chambers demonstrated a typical waisted mass at the level of fossa ovalis, producing a dumbbell shape and a wedge-shaped lateral part where the density indicated that the mass consisted of adipose tissue. Another image showed the LHIS projecting "finger-like" fat along the coronary sinus. In a coronal image, the inferior vena cava was showed encased in the tumor and mildly compressed. The amount of subepicardial fat was greatly increased and appeared to be united with the interatrial mass. Neither metastasis nor continuous invasion of the tumor in the lung or mediastinum was confirmed. Both right and left cardiac catheterization revealed normal cardiac function, including a mean pressure of the superior vena cava 6 mmHg, of the inferior vena cava 7 mmHg, right atrium 6 mmHg, PCWP 10 mmHg and the pressure of right ventricle 23/7/8 mmHg, PA 21/15/16 mmHg, left ventricle 123/–9/13 mmHg and Ao 120/74/

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Fig 1. The 2 soft tissue tumors: 1 in the left anterior chest wall, slightly elevated and measuring 24×14 cm, and the other, in the left arm and presenting as a pendulous mass, approximately 25×19 cm in diameter.

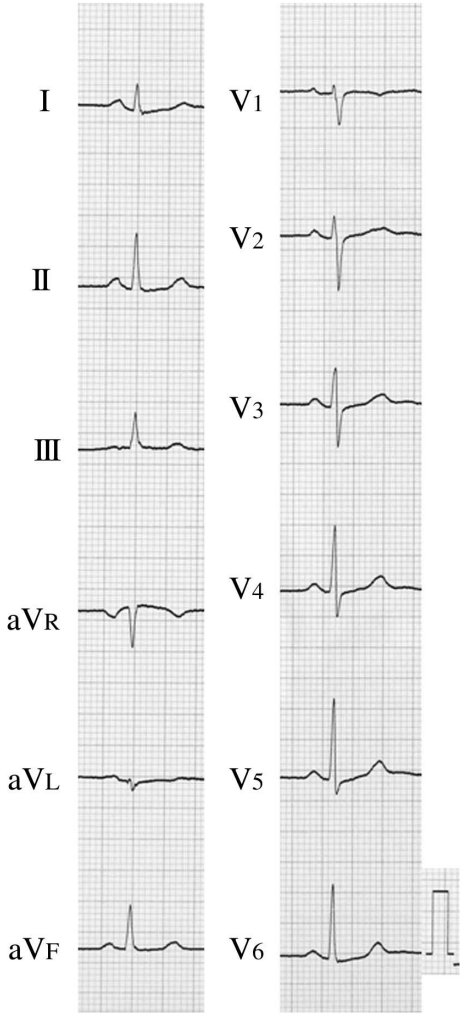

Fig 2. 12-leads ECG showed sinus rhythm (heart rate 65 beats/min) and P-wave abnormalities consisting of amplitude <1.0 mm in lead III, duration >100 ms, and "notching" configuration.

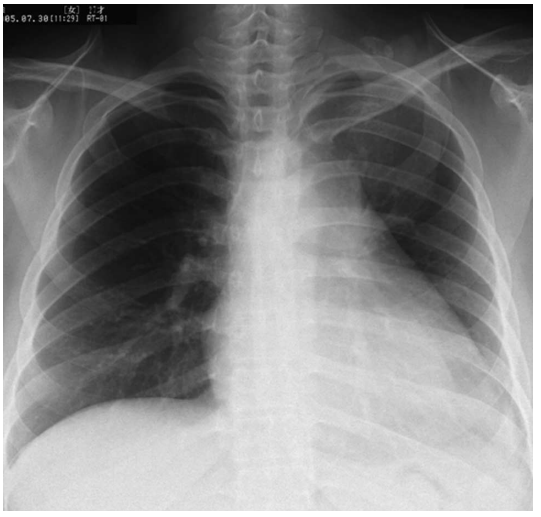

Fig 3. Chest X-ray shows mild cardiomegaly.

95mmHg. The CI was calculated at 3.0 $L \cdot min^{-1} \cdot m^{-2}$. Coronary angiography showed no signs of stenosing coronary disease. Percutaneous biopsy of the atrial mass was performed on the 5th day, using a Biopsy Forceps (T-REX®; Boston Scientific, Natick, USA), and 6 specimens were obtained.

Of these, 3 comprised, of a mass of mature adipose tissue, subendocardial collagenous zone, which was markedly thickened and in which small amounts of cardiac myocytes were intertwining with the fat cells. The other 3 specimens were taken from the myocardium and here too, mature adipose tissues were found creeping through the mesh of the myocytic network (Fig 6). On the whole, the myocytes were markedly thickened. In some places they were haphazardly oriented, giving an impression of fiber disarray (Fig 6), but this seemed attributable to the fatty infiltration, which transformed the otherwise bundle-forming myocytes. No signs of injuries were confirmed in the myocytes themselves, except for several contraction bands that were likely to have been produced when the specimens were excised. The adipose cells were seemingly normal, with little nuclear atypia. There were no lipoblasts or brown fat tissue. In every respect, the findings were thought to be consistent with what has been described in previous reports of LHIS.

The soft tissue tumor in the presternal chest wall was surgically excised. Microscopically it presented quite similar finding to the mass in the atrial septum; that is, increased subcutaneous adipose tissue entirely composed of mature adipose cells (Fig 7), a picture corresponding to

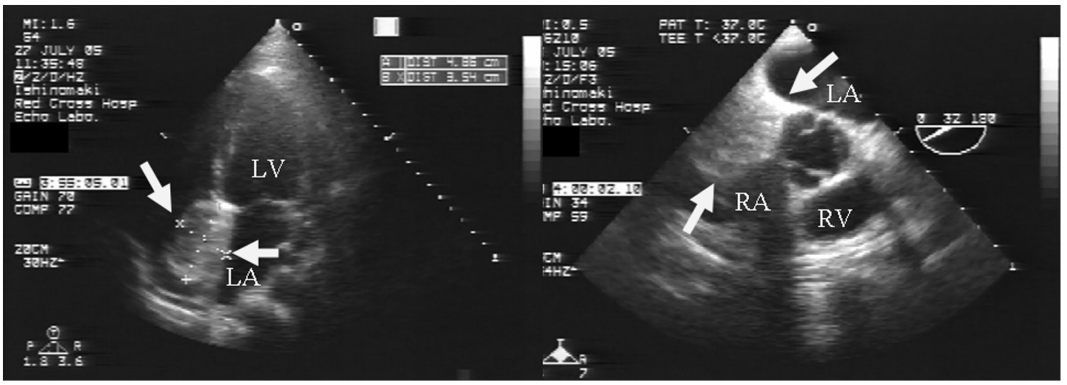

Fig 4. (A) Apical 4-chamber 2-dimensional echocardiogram demonstrates a large mass (48×35 mm) in the interatrial septum (arrows). (B) Transesophageal echocardiogram showing the interatrial solid mass adhered to the noncoronary cusp. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

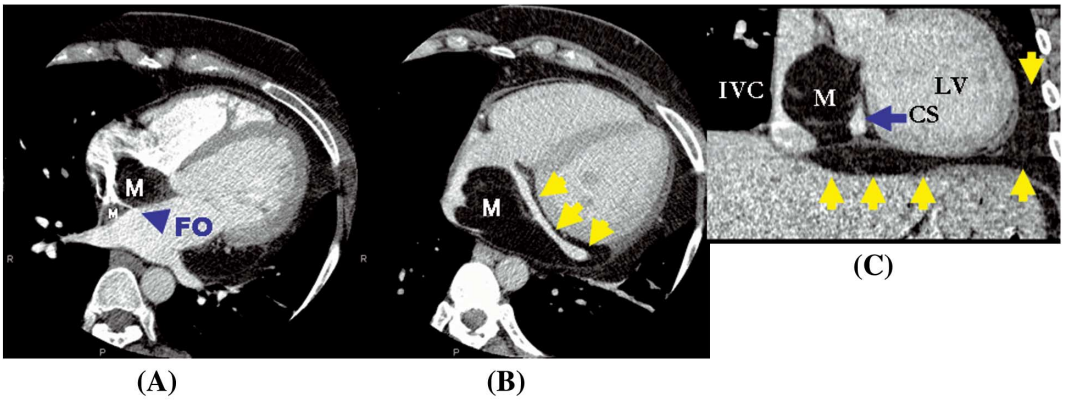

Fig 5. (A) Multislice computed tomography image show typical waist in the fatty mass at the level of the fossa ovalis, producing a dumbbell shape and wedge-shaped lateral part. (B) Lipomatous hypertrophy of the interatrial septum with a "finger-like" projection of fat along the coronary sinus (arrow). (C) Coronal image shows encasement and mild compression of the inferior vena cava. The coronary sinus is surrounded by the fat. Also note the increased pericardial fat (yellow arrowheads). M, fatty mass; FO, fossa ovalis; IVC, inferior vena cava; CS, coronary sinus; LV, left ventricle.

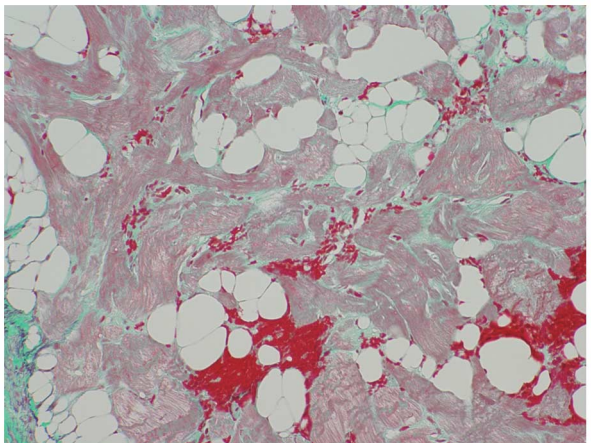

Fig 6. Microscopic findings of cardiac mass. Subendocardial collagenous zone is markedly thickened. Mature adipose tissue found creeping through the mesh of the myocytic network. The myocytes, variable in thickness and haphazardly oriented, give an impression of fiber disarray, but this seema attributable to fatty infiltration, which transformed the otherwise bundle-forming myocytes. The adipose cells themselves are seemingly normal with little nuclear atypia. There are no lipoblasts. In every respect, the findings are consistent with lipomatous hypertrophy of the interatrial septum.

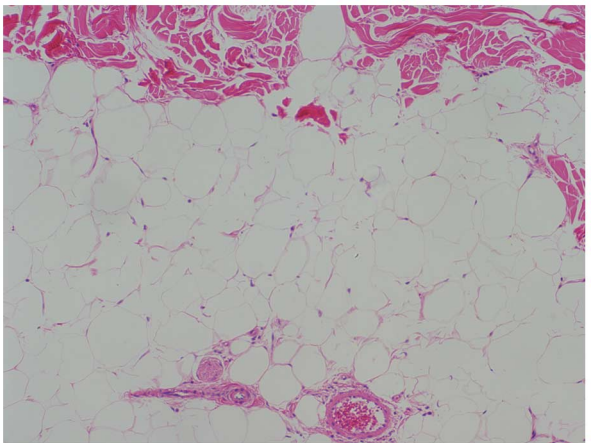

Fig 7. Microscopic findings of the soft tissue tumor of the chest wall. Similar findings as in the atrial septum; that is, it consists of increased subcutaneous adipose tissue entirely composed of mature adipose cells, a picture corresponding to well-differentiated lipoma.

well-differentiated lipoma.

Surgical treatment for the cardiac mass was not considered to be an indication because the mass was not creating any symptoms and there were no signs of circulatory obstruction. The patient, after being given a sufficient explanation about the benign nature of the cardiac and skin lesions, was discharged in September 2005 for subsequent follow-up. She remains asymptomatic 17 months after discharge, and the intra-atrial mass and skin lesions remain as they had been before.

Discussion

In pathological terms, LHIS is described as a nonencapsulated accumulation of mature adipose cells or of brown fat cells, with enlarged cardiac myocytes[1] The disease is considered when a deposit of fat in the atrial septum is confirmed at the level of fossa ovalis and exceeds 2 cm in transverse dimension. According to Burke et al[2] more than one-third of patients diagnosed as having this disease are obese, and there is an association between the patient's body weight and the thickness of the atrial septum. After its first postmortem description by Prior in 1964[3] about 200 cases have been reported, but there is speculation that in reality, the incidence is much higher. Reyes et al[4] found the condition in approximately 1% of autopsy cases, whereas analyses of transthoracic echocardiography by Pochis et al[5] revealed LHIS in as many as 8% of nonselected patients. In a study by Christoph et al using MSCT scanning, an incidence of 2.2% was given[6] There have been descriptions of the electrocardiographic abnormalities associated with LHIS: the characteristic "dome and dip" configuration of P waves in leads II, III and aV_F[7] The incidence of atrial arrhythmias in patients with LHIS who underwent autopsy postmortem is said to range from 40 to 70%[8] The abnormalities underlying arrhythmias were atrial fibrillation, atrial premature complexes, junctional rhythms or ectopic atrial rhythms.

Echocardiography, MSCT scanning, and magnetic resonance imaging have increasingly been used in an effort to obtain more detailed pathological information[9,10] However, the results obtained by these measures have scarcely added new information to what is already known; that is, there is an increased amount of epicardial and/or mediastinal fat. Thus, so long as diagnosis is based on conventional imaging alone, the differentiation of LHIS from other cardiac neoplasms or other proliferative disease might be difficult. Definitive diagnosis requires tissue sampling to exclude myxoma, thrombus, amyloidosis, and benign and malignant tumors. Thus, in the present case, percutaneous transvenous biopsy of the right ventricular endomyocardium[1,11] currently a commonplace procedure, was used to get samples of atrial tissue. Microscopic examination of the specimens provided results sufficiently convincing of a typical case of LHIS.

Lipomatous hypertrophy can cause compression of the superior or inferior vena cava. Only in rare cases is an interatrial mass large enough to produce altered hemodynamics and congestive heart failure and severe rhythmic disorders, in which case surgical correction should be considered, depending on the growth and size of tumor and its relation to the great vessels[1,12,13]

Besides LHIS, diffuse lipomatosis is a rare condition defined as a diffuse overgrowth of mature fat affecting a large portion of an extremity or portions of the trunk[1,14] Two disease types are recognized: in 1, disease usually begins in infant under 2 years of age, but in some cases it becomes apparent in adult. It typically involves both the subcutaneous tissue and the underlying muscles, and may be associated with bone involvement and osseous hypertrophy. The other type is multiple symmetric lipomatosis[1,15] a disease that almost always affects middle-aged men. There are large, symmetrical fatty masses primarily in the neck and shoulder areas, although they can emerge in the cheeks, breasts, upper arms, axillae, trunk and mediastinum. The deposits of bland fatty tissue involve both the subcutaneum and deep soft tissues, and may extend along the intermuscular space of the neck and upper chest, resulting at times in partial obstruction of the esophagus or trachea.

We speculate that in the present case, LHIS developed as a cardiac involvement of multiple lipomatosis, possibly of the latter of the above 2 types.

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