A 60-year-old woman was found to have myelodysplastic syndrome (MDS) in 1999. She was transfused 2 units of packed red blood cells 4 times a month. From January 2004, she developed congestive heart failure, which presented dyspnea on exercise and pretibial edema. In May 2004, the patient was admitted under a diagnosis of congestive heart failure (total amount of packed red blood cells: 249 units).

The physical findings on admission: body temperature, 36.2°C; height, 159 cm; body weight, 45 kg; heart rate, 66/min and irregular; and blood pressure, 92/54 mmHg. Pansystolic murmurs (Levine 3) were audible at the apex. The respiratory sound was clear, and her liver was enlarged, with a 3cm span below the right costal margin on the midclavicular lines, but not palpable in the spleen.

Transthoracic echocardiography was performed using an Aplio (Toshiba Medical Systems Corp, Tokyo, Japan) with PST-30BT transducer. (Center frequency: 3.6 MHz) On two-dimensional and M-mode echocardiography (Figure 1), left ventricular wall motion was globally reduced, indicated as left atrial dimension of 38 mm, left ventricular diastolic dimension of 57 mm, left ventricular systolic dimension of 49 mm, functional shortening of 15 %, intraventricular septum thickness of 6 mm, left ventricular posterior wall thickness of 6 mm, and ejection fraction of 30 %. Also, small amount of pericardial effusion was observed. Color Doppler echocardiography showed moderate mitral regurgitation and severe tricuspid regurgitation.

We analyzed the left ventricular inflow velocity by pulsed Doppler echocardiography. E wave velocity, isovolumic relaxation time, and deceleration time were 90 cm/sec, 82 msec, and 185 msec, respectively. The pressure difference between the right atrium and ventricle was estimated as 38 mmHg using Bernoulli’s for-
Fig. 1  Two-dimensional and M-mode echocardiography of the present case. The reduced wall motion of left ventricle could be observed in M-mode image (Left Panel).

Fig. 2  Strain images of normal case. Left panel: Two-dimensional strain image of left ventricular short axis view. Right panel: Color M-mode strain profile on posterior wall. The transmural strain gradient with bright yellowish at endocardium could be observed in both of left ventricular short axis view and color M-mode (arrow).

Fig. 3  Strain image of the present case. Left panel: Gray Scale image of left ventricular short axis view. Center panel: Strain image of left ventricular short axis view. Right panel: Color M-mode strain profile on posterior wall. No strain was observed on epicardial side (arrow).
mula of tricuspid valve regurgitation observed with continuous wave Doppler echocardiography.

We analyzed tissue Doppler echocardiography using a TDIQ (analytical software) in this echo machine. Color M-mode strain imaging was obtained from the parasternal short-axis view in the end-systolic phase. Velocity data measured by tissue Doppler echocardiography was angle-corrected by designating the center of the left ventricular cavity. Time-integration of the velocity data gave the displacement of the target myocardium toward the left ventricular center. Spatial differentiation of the displacement gave strain, which was color-coded on end-systolic parasternal left ventricular short axis images. The larger strain value was expressed by bright yellowish color and smaller value by dark reddish color. Since normal myocardium has higher strain at endocardium, normal myocardium in left ventricular short axis view was expressed by the myocardial transmural gradient with bright yellowish at endocardium and dark reddish at epicardium. (Figure 2, left panel) The same gradient with bright yellowish at endocardium could be observed with color M-mode strain image. (Figure 2, right panel)

In the present case, the lack of color-coded strain was observed on epicardial side in left ventricle short axis view (Figure 3, center panel) , while the strain on endocardial side still remained. In color M-mode strain image, no strain was also observed on epicardial side (arrow) and the strain profile gradient frequently observed in normal myocardium was not recognized. (Figure 3, right panel)

T2-weighted MRI images confirmed myocardial deposition of iron. The deposition of iron over whole myocardium could be observed, however it was difficult to evaluate the transmural distribution of iron deposition on myocardium (arrow) (Figure 4).

Discussion

Hemochromatosis represents primary or secondary deposition of an excessive amount of iron in various organs such as the liver, spleen, bone marrow, lymph nodes, adrenal glands, and thyroid. It also deposits in the heart, thus, secondary cardiomyopathy causes congestive heart failure[1]. In the present patient, MDS required continuous blood transfusion; therefore, iron overload resulted in many organ deposition of iron.

The contraction of normal myocardium starts at end-diastole and is maximized at end-systole during cardiac cycle. In space, the contraction is minimum at epicardi-

um and increases as toward the endocardium[2,3]. When myocardial ischemia occurs, myocardial contractility on the endocardial side is initially affected, followed by that on the epicardial side. In addition, Sabbah et al.[4] implanted ultrasound crystal in an experiment, and reported that myocardial contractility on the endocardial side comprised 83 % of the left ventricular contractility (myocardial contractility on the epicardial side: 17 %).

Recent studies have evaluated regional myocardial function by tissue Doppler echocardiography. Myocardial strain imaging facilitates the evaluation of regional myocardial function without being influenced by cardiac rotation and translation[5].

Several studies have reported myocardial strain gradient imaging from the endocardial side toward the epicardial side with healthy volunteers[6-7], in patients with nonobstructive hypertrophic cardiomyopathy[8], cardiac lymphoma[9] and coronary artery disease[10-11].

We analyzed the radial myocardial strain values of the left ventricular short axis view.

Usually, the myocardial strain gradient is reduced on the endocardial side in the presence of myocardial ischemia.

However, in the present patient, the strain value on the epicardial side was reduced. This may have been because the lesion related to myocardial deposition of iron deteriorated the contraction on the epicardial side in the patient with cardiac hemochromatosis.

Olson et al.[12] reported that iron deposition was similar among the anterior, inferior, septum, and free

Fig. 4 T2-weighted MRI of the present case. The deposition of iron over whole myocardium could be observed, however it was difficult to evaluate the transmural distribution of iron deposition on myocardium (arrow)
Myocardial strain imaging of hemochromatosis

wall of the left ventricle in 14 autopsied cases. Buja et al.[13] indicated that iron deposition in the ventricular muscle was higher than in the atrial muscle, and that iron deposition was marked in 1/3 of the epicardial side.

In this study, myocardial strain imaging facilitated the analysis of left ventricular lesions involving the outer layer of the left ventricle to the inner layer in our patients with hemochromatosis. In the future, the association with the prognosis and treatment-related improvement should be serially examined. Myocardial strain imaging may facilitate the evaluation of other types of secondary cardiomyopathy in the early stage.

Hemochromatosis can be assessed by MRI images, however, it is not feasible to assess the transmural condition of myocardium by MRI images. Myocardial strain imaging has potential for assessing the transmural condition of iron deposition and the effect of MDS to the heart in an early stage.

References