Ampulla (‘Takotsubo’) Cardiomyopathy of Both Ventricles


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An 84-year-old woman was admitted to hospital with chest pain at rest. An electrocardiogram showed ST-segment elevation in leads II, III, aVF and V2-6, and the 2-dimension echocardiogram showed apical ballooning akinesis and basal hyperkinesis of both ventricles. ⁹⁹ᵐTc-tetrofosmin myocardial single photon emission computed tomography (SPECT) showed severely reduced uptake in the apex. Coronary angiography did not show any organic stenosis, and epicardial coronary spasm was not provoked by the ergonovine loading test. Left ventriculography showed apical ballooning akinesis and basal hyperkinesis, which were also apparent on right ventriculography. The coronary flow velocity pattern showed rapid diastolic acceleration and deceleration times, and the coronary flow reserve measured with a Doppler guide wire was severely decreased. ⁹⁹ᵐTc-tetrofosmin myocardial SPECT showed improvement in the findings after 14 days, and the coronary flow velocity pattern and coronary flow reserve improved after 30 days. Left and right ventriculography both revealed mild improvement in the wall motion. These findings suggested that a microcirculation disturbance caused ampulla (‘Takotsubo’) cardiomyopathy. (Circ J 2004; 68: 1076–1080)

Key Words: Ampulla (Takotsubo) cardiomyopathy; Doppler guide wire; Microcirculation disturbance; ⁹⁹ᵐTc-tetrofosmin myocardial SPECT; Ventricles

A heart syndrome with acute onset and characterized by chest symptoms, elevated ST segment on electrocardiogram (ECG), transient balloon-like asyn-ergy in the apical regions and hyperkinesis in the basal regions on left ventriculography, minimal myocardial enzymatic release, and no significant luminal narrowing of the coronary artery, was originally named by Sato et al as ‘Takotsubo’ cardiomyopathy⁴ after its distinctive appearance on left ventriculography of this syndrome. In Japan, the takotsubo, which is a unique fishing pot (‘subo’) with a round bottom and narrow neck, is used for trapping octopus (‘tako’). Since then, many reports of the disease have been published, but the details of its mechanism are still unclear. We describe a rare case of ampulla cardiomyopathy of both ventricles, in which the microcirculation disturbance was evaluated using ⁹⁹ᵐTc-tetrofosmin myocardial single photon emission computed tomography (SPECT) and Doppler guide wire.

Case Report

An 84-year-old woman was admitted to hospital with chest pain at rest. She had a history of cerebral infarction, Parkinson’s disease and right femoral neck fracture, but no coronary risk factors. Physical examination revealed blood pressure of 92/56 mmHg, and S3 sound and moist rales were audible. An ECG showed ST-segment elevation in leads II, III, aVF and V2–6 (Fig 1) and chest X-ray showed mild cardiomegaly with a cardiothoracic ratio of 55%. The leukocyte count was elevated at 15,700/µl. The serum concentrations of AST (84IU/L), lactate dehydrogenase (453IU/L), creatine kinase (CK) (633IU/L, max 862IU/L), CK-MB (69IU/L, max 82IU/L), C-reactive protein (13.2mg/dl) and brain natriuretic peptide (1,010 pg/ml) were also elevated. Urinalysis revealed Escherichia coli. The concentrations of catecholamines and antibodies of various viruses were normal. Two-dimensional echocardiogram showed apical ballooning akines is and basal hyperkinesis of the left and right ventricles, ⁹⁹ᵐTc-tetrofos min myocardial SPECT showed severely reduced uptake in the apex (Fig 2) and coronary angiography did not show any organic stenosis other than in a diagonal branch (Fig 3). Left ventriculography showed apical ballooning akinesis and basal hyperkinesis; the ejection fraction (LVEF) was 33% (Fig 4). The pressure gradient between the outflow tract and the apex of the left ventricle was not detected. Right ventriculography also showed apical ballooning akinesis and basal hyperkinesis (Fig 4). The coronary flow velocity pattern and the coronary flow reserve (CFR: ratio of the peak hyperemic to resting coronary flow velocity) were measured with a Doppler guide wire in the middle portion of the left anterior descending artery after administration of intracoronary adenosine triphosphate disodium (Fig 5). The coronary flow velocity pattern showed systolic reverse flow, and rapid diastolic acceleration and there
Fig 1. ECG in the acute phase shows ST-segment elevation in leads II, III, aVF and V2–6, which persisted in leads II, III and aVF after 30 days.

Fig 2. 99mTc-tetrofosmin myocardial SPECT in the acute phase shows severely reduced uptake in the apex, which only mildly improved after 14 days.

Fig 3. Coronary angiography does not show any organic stenosis other than in a diagonal branch.
was rapid deceleration. The average peak velocity was 22 cm/s, and it did not improve after administration of adenosine triphosphate disodium (CFR=1.0). The findings were the same in the left circumflex and right coronary artery. 123I-3-methyl-iodo-phenyl pentadecanoic acid (BMIPP) myocardial SPECT showed severely reduced uptake in the apex (Fig 6) and 123I-metaiodobenzylguanidine (MIBG) myocardial SPECT also showed severely reduced uptake in the apex; the uptake ratio of heart to mediastinum was 1.72 and the washout rate was 57.9 % (Fig 6). 

Severely reduced 99mTc-tetrofosmin myocardial uptake, the coronary flow velocity pattern and degradation of the CFR suggested that the ampulla cardiomyopathy may have been caused by a microcirculation disturbance. Therefore, we instituted treatment with an antiplatelet agent (ticlopidine 200 mg/day and aspirin 162 mg/day), KATP channel opener (nicorandil 15 mg/day), angiotensin-converting enzyme inhibitor (imidapril 5 mg/day), diuretic (azosemide 60 mg/day and spironolactone 25 mg/day) and nitric acid (isosorbide dinitrate 40 mg/day), after which her symptoms...
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The 99mTc-tetrofosmin myocardial SPECT findings showed mild improvement after 14 days (Fig 2), but after 30 days, the ST-segment elevation in leads II, III and aVF was still appearing on the ECG (Fig 1). The coronary flow velocity pattern improved in 3 vessels and in particular, the CFR improved to 1.5 in the left anterior descending artery (Fig 5). Abnormalities in the bilateral ventricular wall motion remained, but the LVEF improved from 33% to 48% (Fig 7). Epicardial coronary spasm was not provoked by the ergonovine loading test. An endomyocardial biopsy was taken from the septum of right ventricle, but showed only fibrosis without the hallmark findings of myocarditis, such as infiltration of lymphocytes, macrophages, plasma cells, eosinophils, giant cells, and polymorphonuclear cells.

**Discussion**

Ampulla (‘Takotsubo’) cardiomyopathy is a syndrome with acute onset of ST-segment elevation and subsequent T wave inversion, mimicking acute myocardial infarction, but with a normal coronary angiogram. Left ventriculography reveals apical ballooning akinesis and basal hyperkinesis (and occasionally normokinesis), which gives the heart a balloon shape. There are only a few reports of ampulla cardiomyopathy of the right ventricle. Ampulla cardiomyopathy is thought to be caused by epicardial multivessel coronary spasm, microcirculation disturbance, or catecholamine overload; and acute myocarditis has also been suggested as a cause of the ventricular wall motion. However, the mechanism remains unclear. In the present case, organic coronary stenosis, except in a diagonal branch, epicardial multivessel coronary spasm and catecholamine overload were not observed and it is possible that the patient’s medications for Parkinson’s disease (levodopa 750 mg/day and bromocriptine mesilate 7.5 mg/day) provoked β-receptors and caused the cardiomyopathy. The antibody titers of various viruses were also normal and the histology results did not suggest myocarditis. Psychological and physical stress could be implicated because she was...
bed-ridden with a urinary tract infection, and 123I-MIBG myocardial SPECT showed reduced uptake. 8

99mTc-tetrofosmin myocardial SPECT is used for myocardial perfusion imaging and can be performed in an emergency. Detailed coronary perfusion examination can be carried out using a Doppler guide wire and both these examinations are used in the investigation of various heart diseases. It has been reported that the coronary flow velocity pattern is normal and the CFR is normal or decreased in diseases. It has been reported that the coronary flow velocity pattern is normal and the CFR is normal or decreased in diseases. In the present patient, the findings from the ECG, 99mTc-tetrofosmin myocardial SPECT, coronary angiogram and Doppler guide wire suggest that a microcirculation disturbance, similar to that seen with reperfusion injury, caused the ampulla cardiomyopathy.

The left ventricular wall motion in a patient with ampulla cardiomyopathy should improve either in a week or after several months. 10 We think that it will take a relatively long time to normalize the bilateral ventricular wall motion in the present patient because the 99mTc-tetrofosmin myocardial SPECT, the CFR and the bilateral ventriculography findings had only improved mildly after 30 days.

The detailed mechanism of the characteristic ventricular shape of this syndrome is yet to be elucidated. The distribution of sympathetic nerves and the density of receptors in the left ventricular myocardium of animals is not uniform, being high in the basal segment and low in the apex for the receptors. 17 This nonuniformity of the sympathetic nervous system in the left ventricle could create a wall motion similar to ‘Takotsubo’ when the sympathetic nerves become tense. 18 The findings from 99mTc-tetrofosmin and 123I-MIBG myocardial SPECT suggest that the nonuniformity of the sympathetic nervous system also contributes to disturbance of the microcirculation, such as a microvascular spasm. The findings from left and right ventriculography suggest that there is nonuniformity not only in the left ventricle, but also in the right ventricle. Ampulla cardiomyopathy can therefore occur in both ventricles. We need to investigate further cases because the extent of the wall motion abnormality may be useful for determination of prognosis.

References


