A Case of Takotsubo Cardiomyopathy Complicated by Ventricular Septal Perforation

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The patient was a 73-year-old female who developed chest pain and dyspnea 16 days after her husband passed away. ST segment elevation was detected on V2–5 by electrocardiography and emergency coronary arteriography was done for suspected acute myocardial infarction. No coronary arterial stenosis was present and ventricular septal perforation and takotsubo cardiomyopathy were diagnosed by left ventriculography. The perforation was closed and the patient was discharged 23 days after surgery. This patient had a very rare case of takotsubo cardiomyopathy, which was complicated by ventricular septal perforation and was saved by surgical treatment. (Circ J 2008; 72: 1540–1543)

Key Words: Ampulla cardiomyopathy; Takotsubo cardiomyopathy; Transient left ventricular apical ballooning; Ventricular septal perforation

The first case of takotsubo cardiomyopathy was reported by Sato et al in 1990¹. The incidence of this disease is high in postmenopausal women with psychosomatic stress. The onset pattern is very similar to that of acute myocardial infarction, such as sudden chest pain and dyspnea, electrocardiographic changes, and abnormal left ventricular (LV) wall movement, but LV systolic failure occurs without significant coronary arterial stenosis.^{1–7} Ventricular septal perforation is known as a severe complication following acute myocardial infarction, but it rarely complicates takotsubo cardiomyopathy. Thus, we report this rare case, in which the patient was saved by surgical treatment.

Case Report

The patient was a 73-year-old female, with a height of 150 cm; body weight, 58.3 kg; blood pressure, 110/68 mmHg; heart rate, 70 beats/min (regular); and respiratory rate, 14 beats/min. The patient was under treatment for hypertension and diabetes. She was vomiting and had an asthmatic attack after her husband passed away. The symptoms were improved by inpatient treatment at the Internal Medicine Department, but chest pain and dyspnea suddenly occurred after 16 days, and ST segment elevation was noted on V2–5 of electrocardiography (ECG, Fig 1). The elevation suggested acute myocardial infarction and emergency coronary arteriography was done. No coronary lesion was present (Fig 2a), but ventricular septal perforation, the absence of apical contraction, and excess base contraction were noted, showing takotsubo-type wall movement, on left ventricu-

lography. Furthermore, the right ventricle and pulmonary artery were imaged via left-to-right ventricular shunt (Fig 2b). Pressure data (mmHg) were: pulmonary artery, 42/28; pulmonary capillary wedge pressure, mean 15; right ventricle, 44/9; and left ventricle, 95/12. The myocardial enzymatic release concentrations were not elevated (Table 1). On chest X-ray examination, the cardiothoracic ratio was increased (62%) and pulmonary congestion was present (Fig 3). Levine 4/6 pansystolic murmur with the strongest point at the apex was heard, and the respiratory sound was accompanied by moist rale on auscultation. A small apical defect was detected by ²⁰¹Tl myocardial scintigraphy and marked reduction of anteroapical accumulation was detected by ¹²³I-MIBG myocardial scintigraphy (Fig 4). Echocardiography detected a 25×12-mm ventricu-

Fig 1. Electrocardiography. The left electrocardiogram (ECG) was recorded during chest pain. ST segment was elevated on V2–5. The right ECG was recorded 14 days after the onset. Negative T waves were present on V1–6.

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Fig 2. (a) Coronary angiography (LAO view); no significant stenosis was present. (b) Left ventriculography before surgery; Takotsubo-type heart wall movement was observed. A left-to-right ventricular shunt was imaged.

Table 1 Changes in Laboratory Test Values

	On admission	At time of chest pain onset	After cardiac catheterization	One day after the onset of chest pain
CK (IU/L)	58	73	65	58
AST (IU/L)	27	42	31	18
ALT (IU/L)	29	29	25	17
LDH (IU/L)	246	305	270	242
CRP (mg/dl)	2.46	0.58	0.54	1.12
WBC (/ml)	7,090	10,400	9,370	7,400

CK, creatine kinase; AST, aspartate aminotransferase; ALT, alanine aminotransferase; LDH, lactate dehydrogenase; CRP, C-reactive protein; WBC, white blood cells.

lar septum defect near the apex, showing a left-to-right shunt. LV ejection fraction (LVEF) was 49.0%; pulmonary to systemic flow ratio (Qp/Qs), 2.96; Qp, 15.7l; and Qs, 5.3l (Fig 5). Takotsubo cardiomyopathy was diagnosed based on the above findings.

Surgery was done 22 days after the onset of ventricular septal perforation. Extracorporeal circulation was established by blood withdrawal via the superior vena cava and inferior vena cava and supply from the ascending aorta, and antegrade cardioplegic solution was infused. Under cardiac arrest, the apex was incised. A perforation measuring 25×12 mm was present in the ventricular septum near the apex. The ventricular septum of the perforated region was not fragile, but relatively firm. A Teflon patch was attached to the perforated region and closed with a section of equine pericardium. As for the LV incision, felt was applied to the inner and outer sides of the incision to close it by sandwiching.

On pathological examination, the ventricular septum showed relatively new myocardial infarction, in which only nuclei were lost, and an old infarct lesion accompanied by hemosiderin deposition was present in the root of the papil-

Fig 3. Chest X-ray photograph. The cardiothoracic ratio was increased (62%) and pulmonary congestion was noted.

Fig 4. Myocardial scintigraphy. The upper and lower columns present ^{201}Tl and ^{123}I -MIBG myocardial scintigraphy, respectively.

Fig 5. Echocardiography. A 25×12-mm defect was present in the ventricular septum near the apex, showing a left-to-right shunt.

lary muscle (Fig 6). No congenital LV diverticulum was present.

The postoperative course was smooth, and T waves on V₁₋₄ ECG were normalized on day 20 after surgery. LVEF was 65% and the LV wall movement was normal on echocardiography on day 21 after surgery. Left ventriculography was performed on day 22 after surgery, and the LV wall movement was normal (Fig 7). The patient was discharged on day 23 after surgery.

Discussion

In 1990, Sato et al reported a disease with a reversible course in which the onset pattern was similar to that of acute myocardial infarction, but the coronary artery was normal, and the lesion was pot-shaped on left ventriculography! At the beginning, the pathology was considered to be abnormal wall movement caused by blood flow insufficiency because of multivessel coronary spasm, ie, conduit vessel-derived ischemic stunned myocardium. However, recent studies proposed that a microcirculatory disorder, myocardial dis-

Fig 6. Pathology. Top: relatively new myocardial infarction with only nuclear loss was present in the ventricular septum. Bottom: an old infarct lesion with hemosiderin deposition was present in the root of the papillary muscle.

order directly as a result of catecholamine, and autonomic nerve rupture-associated neurogenic stunned the myocardium,¹⁻⁷ but no cause has been established. Recent studies on stress-induced LV wall movement disorder have elucidated the importance of c-fos expression in not only the

Fig 7. Left ventriculography after surgery. Left ventricular wall movement was normal.

heart but also the brain, particularly the lateral septal and lateral ventricular subthalamic nuclei considered to be involved in cardiac autonomic control, and its inhibition by estradiol⁸. The prognosis of takotsubo cardiomyopathy was considered to be good at the beginning, but cases with cardiorrhexis^{9,10} and ventricular septal perforation¹¹ have been reported, although the incidence is rare, indicating that severe complications or sudden cardiac death might be induced.

This patient showed all clinical features of takotsubo cardiomyopathy. The features are: (1) The incidence is high in elderly females; (2) the symptoms at onset were similar to those of acute myocardial infarction; (3) the LV wall movement was reduced in the central to apical region, but enhanced in the base; (4) there was no coronary arterial lesion; (5) the abnormal wall movement was improved within several weeks; (6) creatine kinase was within the normal range or only slightly elevated; (7) ECG changes (ST segment elevation and negative T-waves); and (8) marked mental stress was present at the onset,¹⁻⁷ based on which takotsubo cardiomyopathy was diagnosed. Regarding ECG changes, the ST segment was elevated on V₂₋₅, no reciprocal changes was noted, and negative T waves appeared on V₂₋₅ 2 weeks after the onset without abnormal Q waves. The negative T waves on V₂₋₄ were normalized 1 month after the onset. Normalization of the abnormal wall movement was confirmed 1 month after the onset by left ventriculography and echocardiography.

Protracted coronary spasm and thrombus are considered to be involved in the pathology of myocardial infarction with a normal coronary artery, and such cases have been reported.¹²⁻¹⁴ In this patient, a necrotizing heart muscle lesion was present in the perforated region on pathological examination. Thus, it cannot be ruled out that coronary spasm induced myocardial infarction, and led to ventricular septal perforation. No coronary spasm induction test was performed. However, myocardial necrosis in the ruptured region on pathological examination in takotsubo cardiomyopathy complicated by cardiorrhexis has been reported.¹⁰ In addition, several cases of takotsubo cardiomyopathy with myocardial necrosis on pathological examination have been reported,^{2,15,16} and the findings in our case also supported these reports. To our knowledge, there has been no reported case of takotsubo cardiomyopathy complicated by ventricular septal perforation and cured by surgical treatment. This case suggested that takotsubo cardiomyopathy might be complicated by ventricular septal perforation, which should be kept in mind.

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