Medically Treated Ventricular Septal Perforation Caused by Takotsubo Cardiomyopathy

A Case Report

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Summary

Takotsubo cardiomyopathy is a common disease, but Takotsubo cardiomyopathy complicated by ventricular septal perforation is very rare. We describe the case of a 92-year-old male who had Takotsubo cardiomyopathy complicated by ventricular septal perforation. We treated the patient medically without surgical or catheter interventions. In three weeks, his abnormal wall motion recovered completely, whereas the ventricular septal perforation remained open. He was ambulatory, subsequently discharged, and had been uneventful for one year. In this manuscript, we discuss the importance of non-invasive management for Takotsubo cardiomyopathy complicated by ventricular septal perforation.

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Key words: Non-invasive management, Very elderly, Complications

Takotsubo cardiomyopathy is characterized by a transient left ventricular wall motion abnormality; patients recover within a few weeks.¹⁾ Emotional or physical triggers are thought to contribute to Takotsubo cardiomyopathy's pathophysiology, and the rates of neurologic or psychiatric disorders were higher in Takotsubo Cardiomyopathy than in acute coronary syndrome.²⁾ However, the International Takotsubo Registry, including 1750 patients with Takotsubo cardiomyopathy, reported that 28.5% of Takotsubo cardiomyopathy occurred without evident triggers.²⁾ Takotsubo cardiomyopathy can be a life-threatening disease with several complications, including cardiac rupture, ventricular septal perforation (VSP), heart failure, and arrhythmia.³⁻⁶⁾ Of those complications, ventricular septal perforation is rarely observed in Takotsubo cardiomyopathy and has been managed by open surgeries or catheter interventions.^{4,7-10)} We present a case of Takotsubo cardiomyopathy complicated by VSP. We treated the patient medically without surgical or catheter interventions.

Case Report

A 92-year-old male was transferred to our hospital because of sudden chest pain. He had a history of atrial flutter and atrial fibrillation which were followed by a local doctor. He also had a history of chronic subdural hematoma and prostatic hypertrophy. Aspirin had been prescribed by a local doctor, but he did not use anticoagulants such as warfarin or direct oral anticoagulants. He did

not have any coronary risk factors. His mother had valvular heart disease, and his sister had unspecified heart disease. He did not have a history or an acute episode of a neurologic or psychiatric disorder. He had an attack of herpes zoster one month before admission. His blood pressure was 141/63 mmHg, and his heart rate was 64 beats/minute. Auscultation revealed a systolic heart murmur at apex. The electrocardiogram showed atrial flutter rhythm and ST segment elevation in leads V2-V4 (Figure 1A). A chest X-ray revealed pulmonary congestion with cardiomegaly (Figure 1B). The value of brain natriuretic peptide on admission was 584.1 pg/mL. Although coronary angiography revealed a normal coronary artery (Figures 2A, 2B), left ventriculography showed apical ballooning and a massive shunt from the left to the right ventricle (Figure 2C). Echocardiography also confirmed the left to right shunt at the apex level, and the calculated Qp/Qs was 1.46 (Figure 3). However, since we did not perform right heart catheterization, we could not confirm the Qp/Qs by blood sampling. He had undergone echocardiography eight months before admission, which did not show any shunt diseases such as ventricular septal defect. Furthermore, he underwent resting dual myocardial scintigraphy using ²⁰¹TlCl and ¹²³I-BMIPP, which showed a significant mismatch between ²⁰¹TlCl and ¹²³I-BMIPP (Figure 4A). Therefore, we diagnosed him as Takotsubo cardiomyopathy complicated by VSP.

Since he was very old and his hemodynamic status was preserved, we decided to treat him medically without surgical or catheter interventions. We started intravenous

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Figure 1. **A:** Electrocardiogram at admission. Electrocardiogram (ECG) showed atrial flutter rhythm (heart rate 74/minute), QS pattern in leads II, III, aV_F and ST elevation with terminal inverted T wave in leads V2-V4. **B:** Chest X-ray at admission. Chest X-ray of supine position showed pulmonary congestion with cardiomegaly. **C:** Electrocardiogram at day 5. ECG showed atrial flutter rhythm (heart rate 55/minute), QS pattern in leads II, III, aV_F and ST elevation in leads V2-V4. Giant negative T waves with prolonged QT interval were shown in leads V2-V5. **D:** Chest X-ray at day 5. Chest X-ray of sitting position are shown in panel D. As compared to the chest X-ray at day 1, pulmonary congestion and cardiomegaly were improved. **E:** Electrocardiogram at day 33. ECG showed atrial flutter rhythm (heart rate 47/min), negative T waves in leads V1-V4. As compared to the ECG at day 5, ST elevations in leads V2-V4 were improved. **F:** Chest X-ray at day 33. Chest X-ray of standing position are shown in panel F. As compared to the chest X-ray at day 1 and day 5, pulmonary congestion and cardiomegaly were improved. The cardio-thoracic ratio was 53%.

diuretics followed by oral diuretics. We did not use angiotensin-converting enzyme inhibitors or beta blockers because of hypotension and bradycardia. His abnormal wall motion recovered completely at day 21, whereas the

VSP remained open. Follow-up electrocardiograms and chest X-rays at days 5 and 33 are shown in Figures 1C, 1D, 1E, and 1F. He was ambulatory and discharged at day 35. He underwent a follow-up resting dual myocardial

Figure 2. Coronary angiography and left ventriculography. There was no significant stenosis in the right (A: LAO view) or left coronary artery (B: RAO caudal view, C: RAO cranial view). Squeezing the middle of the left anterior descending artery was observed (Arrow in C). Left ventriculography showed apical ballooning. A massive shunt from the left to the right ventricle was observed (D: systolic phase, E: diastolic phase).

Figure 3. Echocardiogram. Transthoracic echocardiography confirmed the shunt from the left to the right ventricle. An arrow indicates the ventricular septal perforation. The calculated Qp/Qs was 1.46.

scintigraphy using ^{201}Tl and ^{123}I -BMIPP at day 48, which showed complete recovery both in ^{201}Tl and ^{123}I -BMIPP (Figure 4B). He had been uneventful since he was discharged from our hospital, but died due to pneumonia at other facility just one year after the onset of Takotsubo cardiomyopathy. Because we could not access the details of his death at the other facility, it was not clear whether his pneumonia was accompanied by heart failure.

Discussion

We described a case of Takotsubo cardiomyopathy complicated by VSP. The patient was ambulatory and discharged without surgical or catheter intervention. While Takotsubo cardiomyopathy is frequently observed in the current clinical setting, Takotsubo cardiomyopathy complicated by VSP is rare. In fact, only eight cases have been reported in the literature (Table). Of those eight patients, four underwent surgery and two underwent catheter intervention. Of the two patients who underwent catheter intervention, one patient underwent surgery following catheter intervention. The other two patients did not undergo surgery or catheter intervention and died. Our case is the first report of a patient who was successfully treated without surgical or catheter interventions.

The difference between post-infarction ventricular septal rupture (VSR) and Takotsubo cardiomyopathy complicated by VSP should be discussed. Post-infarction VSR usually requires emergent or urgent surgery for in-hospital survival because the clinical outcome is poor in post-infarction VSR without surgical interventions.¹¹⁾ Since post-infarction VSR usually occurs after broad anterior myocardial infarction, left ventricular systolic function, such as ejection fraction, is significantly impaired.¹²⁾ Moreover, the impaired left ventricular systolic function would not fully recover even after surgical or catheter interventions.^{13,14)} On the other hand, the impaired left ventricular systolic function in Takotsubo cardiomyopathy

Figure 4. Dual myocardial scintigraphy using $^{201}\text{TlCl}$ and ^{123}I -BMIPP. Dual myocardial scintigraphy at day 5 showed significant mismatch between $^{201}\text{TlCl}$ and ^{123}I -BMIPP (A), whereas dual myocardial scintigraphy at day 48 did not show any mismatch (B).

Table. Summary of Literature Cases with Takotsubo Cardiomyopathy Complicated by Ventricular Septal Perforation

Case	Reference No.	Age	Gender	LVEF (%)	Qp/Qs	Assisted circulation	Invasive treatment	Outcomes
1	7)	71	Female	25	NA	IABP	Surgical	Survival
2	8)	73	Female	49	2.96	-	Surgical	Survival
3	1)	84	Female	NA	1.97	IABP	None	Death
4	5)	81	Female	67	2.82	-	None	Death
5	4)	79	Female	41	NA	-	Intervention*	Survival
6	6)	73	Male	NA	1.9	-	Surgical	Survival
7	9)	84	Female	50	NA	IABP	Intervention* → Surgical	Survival
8	10)	73	Female	58	NA	ECMO	Surgical	Death

LVEF indicates left ventricular ejection fraction; Qp/Qs, pulmonary blood flow/systemic flow ratio; IABP, intra aortic balloon pumping; ECMO, extracorporeal membrane oxygenation; NA, not available; and Intervention*, amplatzer septal occluder.

would fully recover without surgical or catheter interventions in a few weeks.¹⁾ In fact, the cardiac scintigraphy results clearly revealed recovery of the left ventricular systolic function in the present case. If the patient with Takotsubo cardiomyopathy complicated by VSP can survive without surgical interventions for several days, the hemodynamics would gradually improve. Only a shunt disease (VSP) remains after a few weeks, and the risk of surgical interventions would be lower in patients with normal left ventricular systolic function than in those with decreased left ventricular systolic function. Therefore, the non-surgical approach toward Takotsubo cardiomyopathy complicated by VSP may be considered as an initial option if the hemodynamics is maintained. Then, we can evaluate the severity of shunt disease and decide whether elective surgery is indicated.

In our case, we chose non-invasive medical therapy rather than surgery, taking into account the patient's advanced age. Patients with Takotsubo cardiomyopathy complicated by VSP may opt to postpone choosing between medical or surgical therapy until the left ventricular systolic function improves. The risk of surgery would be greater in patients with decreased systolic function with VSP than in patients with preserved systolic function with VSP. Nevertheless, we should mention the possibility of survival without surgery because all surviving patients in the literature received surgery or catheter intervention.

In conclusion, Takotsubo cardiomyopathy complicated by VSP is a very rare disease. Unlike post-infarction VSR, the left ventricular systolic function would improve in Takotsubo cardiomyopathy complicated by VSR. If possible, choosing medical or surgical therapy can be postponed until the left ventricular systolic function improves.

Disclosures

Consent: Written informed consent was obtained from patient for publication of this case report.

Conflicts of interest: The authors declare no conflicts of interest in association with the present case report.

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