Case Report

Left ventricular apical ballooning syndrome in a patient with infundibular stenosis of the right ventricle: A case report

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Summary  A 73-year-old female patient with a past history of right ventricular infundibular stenosis was admitted to our intensive care unit because of right ventricular dysfunction. On the fifth day of hospitalization, she suddenly experienced dyspnea without chest pain despite the improvement of her condition by initial medical treatment. Although electrocardiography revealed no ST-segment elevation, echocardiography and myocardial perfusion using 99mTc-MIBI revealed new development of severe symmetrical akinesia and reduced perfusion of the left ventricular (LV) apex and mid-ventricle. LV apical ballooning syndrome was diagnosed based on the minimal elevation of cardiac enzymes (peak cardiac troponin I 0.18 ng/ml) despite the presence of large regions of focal myocardial damage in the myocardium and the absence of positive ECG diagnosis and urgent coronary angiography. Previous coronary angiography revealed normal coronary arteries and the left anterior descending artery without full irrigation around the apex making apical ballooning. On the 12th day of hospitalization, despite the use of positive inotropic treatment, it was impossible to maintain hemodynamic stability, and the patient died prior to the functional recovery of the left ventricle.

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Introduction

Left ventricular (LV) apical ballooning syndrome (ABS), also known as “Takotsubo” cardiomyopathy, is a cardiac syndrome characterized by transient LV dysfunction with chest discomfort, electrocardiographic changes, and minimal release of myocardial enzymes, collectively mimicking acute myocardial infarction [1–3]. Its prognosis has
generally been reported to be promising [3]. Here, we report a patient with infundibular stenosis of the right ventricle who suffered from LV apical ballooning resembling ‘Takotsubo’ cardiomyopathy and who died in the coronary care unit. This case emphasizes the importance of awareness of this condition as well as the variability of its prognosis in patients admitted with underlying basal cardiac disease in the intensive care unit.

Case report

A 73-year-old woman with a history of colon polyp and benign ovarian tumor was referred to the coronary care unit in our hospital due to progressive leg edema and general fatigue.

One year previously, right-left ventricular pressure study and ventriculography revealed paroxysmal atrial fibrillation and severe right ventricular (RV) infundibular stenosis with a systolic pressure gradient of 105 mmHg between the RV and pulmonary artery (Fig. 1). Surgical intervention to treat the RV infundibular stenosis was rejected by the patient, and a permanent pacemaker was implanted to reduce the pressure gradient between the right atrium and RV in another hospital. Coronary angiography (CAG) revealed normal coronary arteries (Fig. 2), and left ventriculography showed normal contraction with an ejection fraction of 65%.

On admission, the blood pressure was 100/60 mmHg, heart rate was 60 beats per minute (all-pacing rhythm), and temperature was 35.4°C. Her consciousness was normal. A significant ejection murmur was heard in the pulmonary area on auscultation. No abdominal abnormalities were found. Neurological examination revealed no abnormalities. Leg edema and cervical venous distention were recognized. Brain natriuretic peptide level was elevated at 1919 pg/ml. Chest radiography revealed a cardiothoracic ratio of 80% and

**Figure 1** Severe right ventricular infundibular stenosis with markedly high systolic pressure gradient between right ventricular and pulmonary artery based on right-left ventricular pressure study and ventriculography.

Severe dilatation of the pulmonary artery without pulmonary venous congestion and pleural effusion. Electrocardiography (ECG) revealed a complete left bundle branch block pattern with all-pacing rhythm (Fig. 3). Echocardiography revealed severe infundibular stenosis, RV dilatation, severe pulmonary artery dilatation, and LV dislodged by RV (Fig. 4). The LV exhibited mild concentric hypertrophy and normal systolic function without systolic anterior motion and outflow obstruction.

On day 5, she exhibited acute deterioration with signs of LV failure and worsening hypoxemia without overt chest pain. Repeat ECG revealed no ST-segment elevation nor giant negative T wave (Fig. 5). The peak creatine kinase (CK) level, CK-MB level, and cardiac troponin I level were elevated at 2824 U/L, 31U/L, and 0.18 ng/ml, respectively. Plasma epinephrine level, plasma norepinephrine level, plasma dopamine level, and urine vanillylmandelic acid were 0.06 ng/ml (normal range 0–0.17 ng/ml),

**Figure 2** Left anterior descending artery without full irrigation around the apex revealed by previous performed coronary angiography.

**Figure 3** Electrocardiography on admission revealed a complete left bundle branch block pattern with all-pacing rhythm.
2.9 ng/ml (normal range 0.15–0.57 ng/ml), 0.3 ng/ml (normal range 0–0.03 ng/ml), and 6.8 mg/day (normal range 1.4–4.9 mg/day), respectively. Echocardiography demonstrated new development of severe symmetrical akinesia of the LV apex and mid-ventricle with hyperkinesia of the basal segments excluding the septum (Fig. 6). The pressure gradient between the RV and pulmonary artery was 101 mmHg. No LV outflow obstruction was detected. Rest myocardial perfusion imaging using 99mTc-MIBI was performed and revealed significantly decreased uptake at the LV apex and mid-ventricle corresponding to the area of the new LV wall motion abnormality. ABS was diagnosed based on the minimal elevation of cardiac enzymes despite the presence of large regions of focal myocardial damage in the myocardium. Unlike the LV, the RV clearly exhibited global staining without reduced staining of the RV apex (Fig. 7). After the onset of ABS, her general condition remained unchanged until day 11. On day 11, her urine output started to decline with severe general fatigue indicating cardiac output reduction. Her general fatigue and urine output did not improve after the catecholamine administration. Finally, in spite of receiving dopamine (10 µg/kg/min), dobutamine (10 µg/kg/min), and norepinephrine (20 µg/kg/min), the patient died without hemodynamic improvement (Fig. 8). CAG was not performed and mechanical circulatory assist devices were not used in agreement with the patient and her family. She became unresponsive, and died on the 12th day following the admission. Autopsy was not performed.

Discussion

Diagnosis of ABS without ST segment elevation and CAG

In our case, ABS was diagnosed based on the minimal elevation of cardiac enzymes despite the presence of large regions of focal akinesia in the myocardium and the absence of positive ECG diagnosis and CAG. Moreover, based on the myocardial perfusion imaging with 99mTc-MIBI, a typical distribution matching ABS was observed. On the other hand, her left anterior descending artery did not irrigate around the apex by previous CAG. Acute coronary syndrome with left anterior descending artery single vessel disease could not cause the severe reduced tracer uptake and severe symmetrical akinesia at the LV apex and mid-ventricle in her coronary perfusion.

The diagnostic criteria for ABS do not include neurohumoral activation. However, it has been reported that ABS can be induced by neurohumoral activation [4], and elevation of catecholamine levels might indicate the occurrence of ABS in the present case. The patient did not show the typical five characteristics of pheochromocytoma; namely, severe...
hypertension, hypermetabolism, hyperglycemia, headache, and hyperhidrosis before ABS development. Moreover, we looked for adrenal tumor by abdominal computed tomography, but we did not detect any mass lesion indicating pheochromocytoma.

**ABS complicating another basal cardiac disease**

On day 7 after the onset of ABS, the patient died suddenly without any thromboembolic events, ventricular fibrillation, and cardiac rupture. In patients with ABS, basal clinical

**Figure 6** New development of severe symmetrical akinesia of the left ventricular apex and mid-ventricle indicating left ventricular apical ballooning as determined by echocardiography.

**Fig. 7** Significantly decreased uptake at the left ventricular apex and mid-ventricle indicating left ventricular apical ballooning based on rest myocardial perfusion imaging using $^{99m}$Tc-MIBI.
condition, especially basal cardiac disease and low cardiac function, may correlate with poor clinical outcome such as cardiac death. In the present case, the RV dysfunction induced by infundibular stenosis comprised a basal organic heart disease. RV pressure overload is associated with abnormal LV systolic and diastolic function [5,6]. Therefore, the additional reduction of ABS-induced cardiac output resulted in decompensated heart failure, and finally lethal deterioration of LV systolic function. Thus, our patient probably died from decompensated heart failure without RV involvement, suggesting that the combination of her underlying basal cardiac disease and ABS rather than severe ABS might have been the direct cause of cardiac death.

It has been reported that in-hospital prognosis is generally good after the appropriate treatment of the acute complication [3]. In a series of 88 patients, only one patient died during hospitalization, and the mortality rate was 2% during a mean follow-up of more than a year. A good clinical outcome may be expected for ABS when the wall motion abnormality is transient and full functional recovery occurs after the acute phase. However, the LV systolic dysfunction caused by ABS, even if it is transient, makes monitoring of patients with ABS essential, especially those with other basal cardiac disease who cannot endure circulatory collapse during the acute phase of ABS. On the other hand, we should discuss the therapeutic strategy for severe RV infundibular stenosis and ABS. Positive inotropic treatment for LV dysfunction could cause a marked reduction of RV output, leading to a hemodynamic catastrophe. Basically, we should determine how to manage adequate catecholamine administration to treat hemodynamic collapse depending on the hemodynamic derangements based on the catheter study. However, we could not perform the pressure study without the patient’s consent. Furthermore, RV pacing discontinuation for hemodynamic improvement might be considered rather than catecholamine administration.

Mental stress and ABS

The etiology of ABS is still unknown. Collapse or deformity of LV chamber induced by RV pressure overload or hypoperfusion of the pulmonary circulation possibly led to ABS development in our patient. Interestingly, Park et al. reported that ABS is relatively frequent in the critically ill patients who are admitted to medical intensive care units without manifestation of cardiac disease [7]. Factors triggering ABS include emotional and physical stress [3,7]. Conscious patients may feel quite uncomfortable in the intensive care unit whether or not they have cardiac disease. Moreover, immobilization alone causes physical stress [8]. Patients with a poor prognosis will experience mental stress due to the fear of death. Taken together, it is not unreasonable for ABS to occur in the intensive or coronary care unit after the admission due to another cardiac disease, as in the present case.

Conclusion

In the coronary or intensive care unit, in which patients may experience mental or physical stress, ABS can occasionally develop after the admission.
In patients with underlying basal cardiac disease, the occurrence of ABS may unfortunately be fatal, although the prognosis of ABS is usually satisfactory. Since the transient LV systolic dysfunction caused by ABS probably varies widely in severity, it is important to monitor ABS patients with underlying basal cardiac disease and avoid excessive optimism regarding the prognosis of ABS.

References


