Reversible Left Ventricular Dysfunction with Apical Ballooning — A Case Report of Ampulla Cardiomyopathy

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Ampulla cardiomyopathy is a rare cardiac syndrome characterized by transient left ventricular (LV) apical ballooning without significant coronary artery disease. The shape of the left ventricular cavity during the acute phase resembles an ampulla. These patients are usually misdiagnosed as acute myocardial infarction because of similar clinical symptoms, electrocardiogram (ECG) changes and myocardial enzyme elevation. The precise etiology remains unclear. However, the clinical course and prognosis are usually benign. We report a 62-year-old woman with ampulla cardiomyopathy and discussed the possible pathogenesis involved. Catecholamine-related microcirculation dysfunction appears to be the most likely etiology.

Key Words: Ampulla cardiomyopathy • Reversible LV dysfunction • Apical ballooning

INTRODUCTION

An unusual cardiac syndrome has been reported that mimics acute myocardial infarction (AMI) without stenosis of epicardial coronary arteries, and is characterized by reversible left ventricular (LV) dysfunction with apical ballooning.1,2 Patients with this syndrome are generally misdiagnosed as AMI owing to similar symptoms, ECG changes (ST-segment elevation and subsequent giant T-wave inversion), and minimal myocardial enzymatic release. Coronary angiography (CAG) and left ventriculography (LVG) thus are usually performed under the impression of ST-elevation AMI. The LVG reveals hypokinesis or akinesis from the mid to the apical portion and normokinesis of the basal portion during the acute phase. The condition was named ‘ampulla cardiomyopathy’ because the peculiar shape of the LV resembles an ampulla (a round bottom with a narrow neck).3,4 We report a case of reversible LV dysfunction with apical ballooning and discuss its possible pathogenesis.

CASE REPORT

A 62-year-old woman with old cerebral infarction and hypertension was admitted to Chang-Gung Memorial Hospital with acute-onset dyspnea and sustained severe chest pain following a hot bath. ECG displayed ST elevation in leads V1-V4 and Q wave formation in V1-V4, III, and aVf (Figure 1A). Under the impression of ST-elevation AMI, medical treatment, including bolus heparin, aspirin and clopidogrel were prescribed. Emergency cardiac catheterization was performed then. CAG revealed patent epicardial arteries with TIMI 3 flow. LVG confirmed the presence of apical ballooning LV dysfunction with an ejection fraction of 44% (Figures 2A and B). No intraventricular pressure gradient was measured. The level of cardiac troponin-I was < 0.4 ng/mL. Symptoms and ECG changes improved follow-
ing administration of diltiazem (90 mg/day). The follow-up ECGs exhibited giant negative T-wave and QT prolongation, particularly over the precordial leads (Figure 1B). Echocardiography one day after CAG revealed recovery of LV wall-motion abnormalities. The \(^{99m}\)technetium-pyrophosphate images of myocardial infarct scan revealed no evidence of acute myocardial infarction (no hot spot). Viral titers of Coxsackievirus, Cytomegalovirus, Influenza virus, Adenovirus, Epstein-Barr virus, and Herpes virus were checked and found to be negative for viral myocarditis. LVG (Figures 2C, D) performed on the 5th admission day showed normal wall motion of the LV with an ejection fraction of 80%. The ECG changes of ST-segment, T-wave inversion and abnormal Q wave were normalized on the 10th admission day (Figure 1C).

**Figure 1.** Evolutional changes of the ECG. On the first day of chest discomfort, the ECG showed ST segment elevation in leads V1-4, and Q-waves in leads V1-3, III and aVF (A). On day 2, the ECG displayed giant negative T-waves and QT prolongation in the precordial leads (B). On day 10, the R-waves reappeared in leads V1-3, III and aVF, and the ST-segments and T-waves normalized in the precordial leads (C).

This patient is now under regular outpatient clinic follow-up in a stable condition.

**DISCUSSION**

Various reports have described reversible LV dysfunction with symptoms resembling those of AMI but without epicardial coronary artery lesions, even during the acute phase with elevated ST-segment.\(^1\). This type of ventricular dysfunction manifests left ventricular wall motion abnormalities as apical ballooning, which generally normalizes within a few weeks. They are frequently observed in older women, and generally occur following mental or physical stress.\(^1\) Serial ECG changes include initial ST-elevation with reduced R-wave voltage; giant negative T-wave and QT prolongation, and resolution of the T-wave and QT-interval changes accompanied by regrowth of the R-wave.\(^6\) Slight cardiac enzyme elevation is often found, suggesting small myocardial injury and the reversible nature of this disease. The clinical prognosis is good compared to that of AMI.

The precise etiologic basis of transient LV apical ballooning is unclear. Several possible mechanisms explain this phenomenon, including epicardial coronary spasm, microvascular spasm or dysfunction, and direct myocardial damage. The patient described here displayed no coronary artery stenosis during the acute stage with elevated ST-segment and LV wall motion abnormalities. Although coronary spasm provocation test was not performed to exclude the possibility of epicardial spasm, simultaneous multivessel spasm is rare. A previous investigation also demonstrated coronary spasm could be induced in only 21% of these patients.\(^1\) Multiple epicardial vasospasm or left main coronary artery spasm may be triggering factors, but are not main factors.

Another possible mechanism is myocardial ischemia resulting from microvascular spasm or dysfunction. Direct assessment of microvascular function has been difficult until now. Ako et al. reported that the coronary flow reserve measured by Doppler guide wire was significantly reduced in patients with ampulla cardiomyopathy, suggesting microvascular dysfunction.\(^5\) The syndrome of angina or angina-like chest pain with a normal coronary angiogram is frequently referred to as syndrome X.
Some evidence indicates that syndrome X results from reduced vasodilator reserve in the microcirculation.\textsuperscript{7} Syndrome X, resembling ampulla cardiomyopathy, is often observed in women. These two clinical entities share several common characteristics, and possibly have the same pathogenesis.

ECG changes, such as depressed or elevated ST segments, QT prolongation and T wave abnormalities, similar to those observed in patients with myocardial ischemia, are observed in 49-100% of patients following subarachnoid hemorrhage.\textsuperscript{8} Some of these patients may present with reversible LV dysfunction.\textsuperscript{8} The observation that beta-receptor blockade reverses ECG changes, and prevents myocardial necrotic lesion suggests that such cardiac abnormalities are caused by increased sympathetic nervous activity with subsequent elevation in levels of circulating catecholamines.\textsuperscript{9,11,12} The released catecholamine could damage myocardial cells either by constricting the myocardial microcirculation or by a direct toxic effect. Previous studies have stated that the apex contains higher density of adrenoreceptor,\textsuperscript{10} which may contribute to its vulnerability to catecholamine toxicity.\textsuperscript{11,12} Kyuma et al. applied intravenous propranolol in three patients with ampulla cardiomyopathy, and found improvement in ECG and LV wall motion abnormalities in the two patients with intraventricular pressure gradient, but no improvement in the patient without intraventricular pressure gradients.\textsuperscript{4} Beta-blocker could be useful in the presence of intraventricular pressure gradient, or in preventing catecholamine cardiotoxicity before the occurrence of heart damage. No intraventricular pressure gradient was found in the present patient. We treated her with calcium channel block instead of beta-blocker, and rapid recovery of LV wall-motion abnormalities was

\textbf{Figure 2.} Acute ballooning and subsequent normalization of the left ventricle. On day 1 of chest discomfort, left ventriculography in the right anterior oblique view showed dilatation and akinesis of the mid and apical portions of the left ventricle (A: systolic phase; B: diastolic phase). On day 5, the sizes and motions of the left ventricle became normal (C: systolic phase; D: diastolic phase).
noted. Experience from a few case studies is insufficient
evidence to conclude that calcium channel blocker is
helpful in patients with ampulla cardiomyopathy, be-
cause the prognosis of this disease entity is relatively
good and the LV dysfunction may recover even without
medication.

Although the precise cause of ampulla cardiomyopathy
remains unclear, we postulate that catecholamine-related
microcirculation dysfunction might be the primary cause.
Further investigations are necessary to clarify the
pathogenesis, which may provide the optimal therapeu-
tic strategy for these patients. Although it is usually
clinically harmless, the transient reversible LV asynergy
can result in cardiogenic shock and mortality.\textsuperscript{11,12} It should
be put into the differential diagnosis of acute coronary
syndrome. Aggressive medical treatment with adequate
hemodynamic support achieves reversal of LV dysfunc-
tion without long-term adverse sequelae.

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壺腹狀心肌病變是一種罕見的疾病，其特徵是短暫的左心室頂部氣球狀突起，卻無明顯的冠狀動脈疾病，在急性期左心室的形狀類似壺腹。這些病人常被誤診為急性心肌梗塞因為兩者有相似的臨床症狀、心電圖變化和心臟酵素升高。雖然此種疾病確切的成因還不清楚，但臨床表現和預後通常不錯。我們報告一位 62 歲壺腹狀心肌病變的女性個案並探討其可能的機制。和 Catecholamine 有關的微循環功能不全是最可能的原因。

關鍵詞：壺腹狀心肌病變、可逆性左心室功能不全、頂部氣球狀突起。