

Ampulla Cardiomyopathy (Takotsubo Cardiomyopathy) in A Patient with Diabetic Ketoacidosis : A Case Report

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Abstract

Ampulla cardiomyopathy (Takotsubo cardiomyopathy) is a syndrome, consisting of acute-onset, transient, and abnormal left ventricular wall motion with apical akinesis and basal normokinesis without any detectable coronary lesions. The syndrome has symptoms and signs that are similar to acute myocardial infarction, such as electrocardiogram (ECG) changes (ST-segment elevation and subsequent giant T wave inversion) and abnormal myocardial enzymes. There are many etiologies for this syndrome that have been discussed in the past decade, but the precise etiology remains unclear. The clinical prognosis is usually benign. We reported a 71-year-old woman with ampulla cardiomyopathy precipitated by diabetic ketoacidosis. We propose that catecholamine-related microcirculation dysfunction appears to be the most likely etiology. (J Intern Med Taiwan 2007; 18: 120-124)

Key Words : Ampulla cardiomyopathy, Takotsubo cardiomyopathy

Introduction

In recent years, there have been many reports in which an unusual cardiac syndrome with a clinical presentation that mimics acute myocardial infarction (AMI) was found to have normal coronary arteries and a left ventricle with reversible asynergy¹. Patients with this syndrome are often misdiagnosed as AMI due to similar clinical symptoms and signs, such as ECG changes (ST segment elevation, subsequent giant T-wave inversion and abnormal Q wave) and minimal elevation of cardiac enzymes². In this situation, there is no significant luminal narrowing of the coronary arteries when examined by means of a coronary angiography.

Ventriculography or echocardiography will reveal basal normalkinesis and apical akinesis in the acute phase, which will return to normal appearance within 2-14 days³. This peculiar pattern was originally named Takotsubo's cardiomyopathy by Dr. Sato³. We report a case of ampulla cardiomyopathy in a diabetic ketoacidosis and discuss the possible etiology.

Case Report

A 71-year-old woman with a history of type 2 diabetes mellitus and hypertension was admitted to a hospital due to a sudden onset of mild chest tightness and dyspnea, which lasted for 2 hours. Physical examination revealed a body temperature of 38.5 °C, a blood pressure of 93/55 mmHg, a pulse rate of 131 beats/min and a respiratory rate of 28 breaths/min. Bilateral moist rales were audible during chest auscultation. Chest radiography revealed increasing interstitial thickening over the bilateral lung fields. An EKG displayed ST-segment depression in leads III and aVF beside ST-segment elevation in leads I, aVL, and V₁₋₆ (Fig. 1-A). The laboratory tests administered upon admission showed: white blood cell count (WBC)19000 / μ L; hemoglobin, 14.4 g/dL; platelet count, 266000/mm; CRP, 18.7 mg/dL; Na, 134.4 mmol/L; K, 3 mmol/L ; Cl, 98 mmol/L; BUN,15 mg/dL; Cr, 0.7 mg/dL; Glucose, 432 mg/dL; CPK, 651 U/L (upper normal limit 167 U/L); CK-MB, 33.7 U/L (upper normal limit 24U/L); Troponin-I, 5.077



Figure 1. A: serial electrocardiograms showed ST segment depression in leads III and aV_F besides ST-segment elevation in leads I, aV_L, and V₁₋₆ on the day of onset.
B: following electrocardiograms showed giant negative T-wave and QT prolongation on the second day of hospitalization.

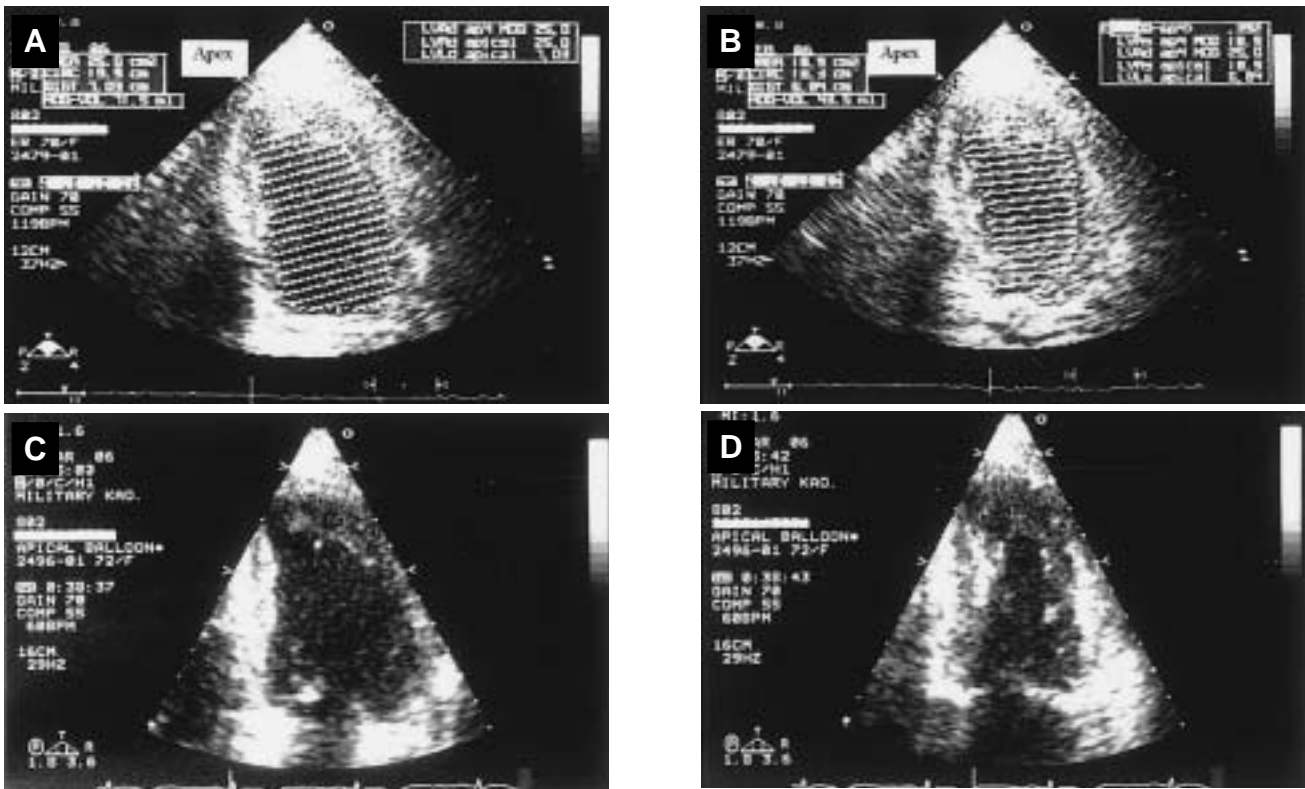


Figure 2. A and B: two-dimensional echocardiogram showed akinesis of anterior wall, apex distal septum and distal lateral wall of the left ventricle on the day of onset. (EF=39 %).

C and D: the second echocardiogram showed recovery from LV wall-motion abnormalities two weeks after admission. (EF=63%)

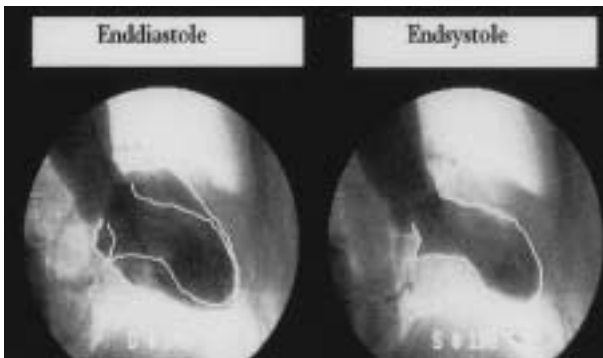


Figure 3. Left ventriculogram showed akinesis around the apical area on the 8th day of hospitalization. (EF=58%)

ng/dL (upper normal limit 0.04 ng/dL). Arterial blood gases were as follows: pH, 7.376; PCO_2 , 31.1 mmHg; PO_2 , 76 mmHg; HCO_3^- , 17.8 mEq/L; O_2SAT , 93.5%. In addition, serum ketone body and pyuria were also detected. Under the impression of acute myocardial infarction and diabetic ketoacidosis precipitated by urinary tract infection (UTI), hospital admission was arranged.

A series of medical treatments, including heparin, aspirin, isosobide dinitrate, insulin and empirical antibiotics were prescribed. Urine culture grew *Escherichia coli*. On the second day of hospitalization, the follow-up ECG exhibited giant negative T-waves and QT prolongation (Fig. 1-B). A two-dimensional echocardiogram showed akinesis of the anterior wall, apex, distal septum, and distal lateral wall besides hypokinesis of the mid to distal inferior wall and posterior wall (Fig. 2-A,B). Chest tightness and dyspnea were relieved in the days following the above medical treatment. Cardiac catheterization was not performed until the 8th hospital day due to patient reluctance. Coronary angiography revealed patent epicardial arteries with TIMI 3 flow. Left ventriculogram showed apical ballooning akinesis (Fig. 3). The estimated ejection fraction (LVEF) was 58%. Detected plasma level of catecholamine was 1024

pg/mL (upper normal limit 827 pg/ml) on the same day. The second echocardiogram showed recovery from the LV wall-motion abnormalities two weeks after admission (Fig. 2-C,D).

Presently, the patient is undergoing regular outpatient clinic follow-up and is in stable condition.

Discussion

Many reports have described reversible LV dysfunction with clinical symptoms resembling those of acute myocardial infarction but without having evidence of epicardial coronary lesions. Most of these cases have been concentrated in Japan. It accounts for about 1 to 2 % of all patients presenting with apparent manifestation of acute myocardial infarction⁴. This ventricular dysfunction, named ampulla cardiomyopathy, manifests LV wall motion abnormalities, as apical ballooning, which is often relieved within 2 weeks⁵. Abnormal findings in ECGs include initial ST-elevation with low R wave voltage, a giant negative T-wave, and QT prolongation⁶. Deep negative T waves may be seen during the course of recovery¹. A mild elevation of cardiac enzymes is often found, which is indicative of a small myocardial injury and the reversible nature of this disease.

Triggering factors for this condition, which have been discussed, included systemic disorders (e.g. subarachnoid hemorrhage, epileptic attacks, exacerbation of bronchial asthma, Guillain-Barre Syndrome, crush syndrome, or an abdominal operation) and a history including an emotional and/or physical trauma (sudden accident, death of a family member, unusual exercise and vigorous excitation)^{2,3}. However, the triggering event for our patient was diabetic ketoacidosis. As we know, this is the first case in which ampulla cardiomyopathy was precipitated by diabetic ketoacidosis. The etiology of this syndrome is still unclear but may be related to epicardial multi-vessel coronary spasm, microcirculation disturbance, catecholamine overload or acute myocarditis⁷.

Although a coronary spasm provocation test was

not arranged in our case to exclude epicardial spasm, simultaneous multi-vessel spasm is very rare. Besides, it has been difficult to directly assess microvascular function until now.

Another possible mechanism is catecholamine overload which was thought to be related to emotional stress-induced ampulla cardiomyopathy⁸. In these cases, elevated levels of plasma catecholamines were detected on the initial hospitalization and even on the ninth hospitalization day. To our knowledge, the DKA results from relative or absolute insulin deficiency combined with counter-regulatory hormone excess, such as catecholamine excesses. In our case, we think catecholamines in the DKA may play an important role on the pathogenesis of nonemotional stress-induced ampulla cardiomyopathy. Review of Yoshihiro et al, suggests elevation of catecholamines may be the primary cause of takotsubo cardiomyopathy, or a result of the condition, but is not absolute.

Overactivation of the cardiac catecholamine receptors may have been suggested as the cause in these patients with a normal level of serum epinephrine. Due to the above reasons, an adrenoceptor blockade may have the effect of preventing this syndrome⁹.

Furthermore, a high-dosage steroid infusion such as methylprednisolone, was prescribed for takotsubo cardiomyopathy to try to reduce the secretion of norepinephrine or the production of cytokines as recommended on the opinion of Sato et al¹⁰.

Although the precise cause of takotsubo cardiomyopathy is still unclear, we propose that catecholamine-related microcirculation dysfunction might be the primary cause. Further investigation of the pathogenesis is necessary, which will in turn clinically provide optimal therapeutic strategy. Although this cardiomyopathy has a good prognosis relative to acute myocardial infarction, it should be regarded as a possible cause of sudden cardiac death in patients without obvious heart disease. Adequate hemodynamic support will reverse LV dysfunction without long-term adverse sequelae.

References

1. Kawai S, Suzuki H, Yamaguchi H, et al. Ampulla cardiomyopathy ('Takotusbo' cardiomyopathy) reversible left ventricular dysfunction with ST elevation. *Jpn Circ J* 2000; 64: 156-9.
2. Kyuma M, Tsuchihashi K, Shinshi Y, et al. Effect of intravenous propranolol on left ventricular apical ballooning without coronary artery stenosis (ampulla cardiomyopathy) - three cases. *Circ J* 2002; 66: 1181-4.
3. Owa M, Aizawa K, Urasawa N, et al. Emotional stress-induced 'ampulla cardiomyopathy'-discrepancy between the metabolic and sympathetic innervation imaging performed during the recovery course. *Jpn Circ J* 2001; 65: 349-52.
4. Akashi YJ, Nakazawa K, Sakakibara M, et al. The clinical features of takotsubo cardiomyopathy. *QJM* 2003; 96: 563-73.
5. Hiroyuki T, Yoshiaki H, Jun S, et al. Transient increase in wall thickness of the left ventricular apex during recovery from ampulla cardiomyopathy. *Ann of Nucl Med* 2003; 17: 707-10.
6. Riyo O, Yoshikazu H, Takefumi T, et al. Specific findings of the standard 12-lead ECG in patients with 'Takotusbo' cardiomyopathy: comparison with the findings of acute anterior myocardial infarction. *Circ J* 2003; 67: 687-90.
7. Nishikawa S, Ito K, Adachi Y, et al. Ampulla ('Takotsubo') cardiomyopathy of both ventricles-evaluation of microcirculation disturbance using 99mTc-tetrofosmin myocardial single photon emission computed tomography and doppler guide wire. *Circ J* 2004; 68: 1076-80.
8. Wittstein IS, Thiemann DR, Lima JAC, et al. Neurohumoral features of myocardial stunning due to sudden emotional stress. *N Engl J Med* 2005; 352: 539-48.
9. Akashi YJ. Reversible ventricular dysfunction takotsubo (ampulla-shaped) cardiomyopathy. *Intern Med* 2005; 44: 175-6.
10. Sato T, Hagiwara K, Nishikido A, et al. Takotsubo (ampulla-shaped) cardiomyopathy associated with microscopic polyangiitis. *Intern Med* 2005; 44: 251-5.

壺腹形心肌病變發生於酮酸中毒病人：個案報告

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摘要

壺腹形心肌病變是一種徵候群，包括急性及短暫性左心室功能異常，其特徵為心尖收縮無力，心臟底部收縮正常，但卻無任何冠狀動脈阻塞之病兆。這種徵候群之症狀及徵象與急性心肌梗塞相似，例如：心電圖之變化(ST段上升和T波倒置)和異常心肌酵素。許多造成此徵候群之病因在過去十年來已被廣泛探討，但真正的致病原因仍然不明。臨床之預後通常良好。我們報告一個由酮酸中毒所引發壺腹型心肌病變於七十一歲女性之個案。我們認為兒茶酚胺所引發微小循環功能異常為最可能之致病原因。