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Catecholamine Cardiomyopathy Associated With Paraganglioma Rescued by Percutaneous Cardiopulmonary Support

— Inverted Takotsubo Contractile Pattern —

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A case of catecholamine cardiomyopathy associated with paraganglioma, which was rescued by percutaneous cardiopulmonary support system (PCPS), is presented. Unlike typical apical ballooning, transthoracic echocardiography and left ventriculography revealed severe left ventricular (LV) dysfunction and an abnormal contractile pattern, consisting of akinesis of the basal and midventricular segments and hyperkinesis of the apical segments. Because of the intractable cardiogenic shock, despite conventional treatment, PCPS was performed. The underlying disease was paraganglioma. Catecholamine excess may induce not only transient LV apical ballooning but also atypical LV ballooning without involvement of the LV apex. Early use of PCPS is important for the patient with a catecholamine crisis who is not stabilized by intra-aortic balloon pump as well as infusion of fluid and inotropic agents. (*Circ J* 2007; **71:** 1993 – 1995)

Key Words: Cardiomyopathy; Catecholamine; Paraganglioma; Percutaneous cardiopulmonary support

Catecholamine cardiomyopathy is known to have characteristic transient wall-motion abnormalities involving the left ventricular (LV) apex and midventricle in the absence of coronary artery stenosis. However, a few cases of catecholamine cardiomyopathy without involvement of LV apex have been reported[1–3] and we report a case of catecholamine cardiomyopathy in a patient with paraganglioma presenting as transient LV ballooning that was successfully rescued by a percutaneous cardiopulmonary supporting system (PCPS).

Case Report

A 47-year-old man with no history of cardiac disease was admitted with acute headache, palpitation, and dyspnea. Cerebral computed tomography (CT) scan ruled out intracranial hemorrhage. On physical examination, he was afebrile with blood pressure of 190/97 mmHg and heart rate of 72 beats/min. The heart sounds were regular without gallops, murmurs or rubs. The notable findings of the initial examination were distended jugular veins and moist rales in both lung fields.

Soon after admission, his oxygen saturation dropped to 80% and he developed dyspnea. In addition, plain chest X-ray showed pulmonary congestion and the arterial blood gas analysis was 7.3-25-57-80% (pH-$PaCO_2$-PaO_2-O_2 saturation). Therefore, tracheal intubation was performed for mechanical ventilation. Initial electrocardiography showed normal sinus rhythm and ST depression in II, III, aV_F, and $V_{1–6}$. The creatinine kinase and troponin I levels were elevated. Emergency echocardiography revealed akinesis of the LV basal and midventricular segments and hyperkinesis of the apical area. Ejection fraction calculated by modified Simpson method was 23% (Figs 1A,B). To rule out acute coronary syndrome, emergency coronary angiography was performed, which showed normal coronary arteries. Left ventriculography showed a characteristic regional wall motion abnormality consisting of akinesia in the midventricular and basal segments and apical hyperkinesia (Figs 1C,D). Despite sufficient saline hydration and continuous infusion of inotropic agents, his systolic pressure was unstable. Because of acute cardiac decompensation and inability to maintain normal blood pressure with more than 2 inotropic agents, circulatory support by intra-aortic balloon pump (IABP) was introduced on hospital day 2. Urine output began to decrease after admission to intensive care unit (ICU) with increasing blood urea nitrogen (BUN)/creatinine levels. On hospital day 3, total anuria developed and the BUN level increased to 37.1 mg/dl, at which time continous venovenous hemofiltration was instituted. Because of hypotension refractory to IABP, intravenous fluid and inotropic agents, PCPS was started on hospital day 4.

Because of the episodic headache, palpitation and fluctuating blood pressure, we suspected a differential diagnosis of pheochromocytoma and performed relevant investigations. CT showed a 7-cm cystic mass in the anterior aspect of the inferior vena cava of the right kidney without distant metastasis (Fig 2). With the use of mechanical support and

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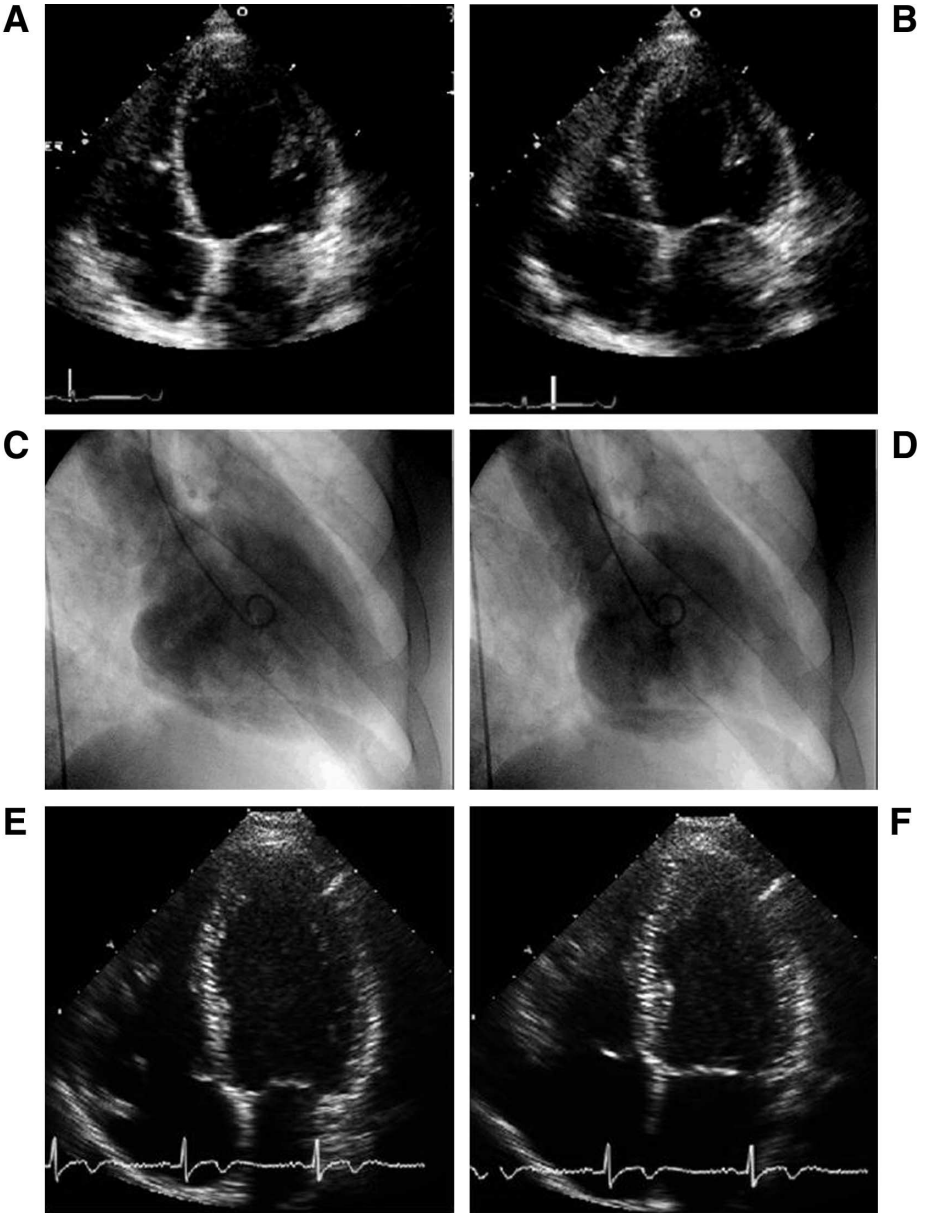

Fig 1. In the acute phase, an apical 4-chamber view shows akinesis of the basal and midventricular segments, with preserved contractility of the apical segments during diastole (A) and systole (B). End-diastolic (C) and end-systolic (D) ventriculograms of the case show akinesia of the basal and middle segments of the left ventricle and normal contractility of the apical segments. On day 18 after surgery, the apical 4-chamber view shows normalized ejection fraction and wall motion during diastole (E) and systole (F).

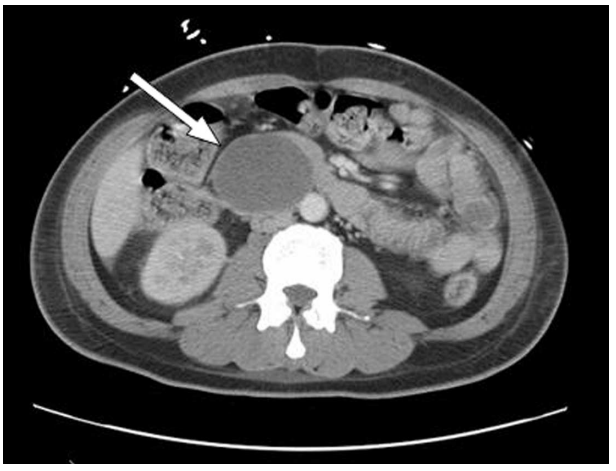

Fig 2. Computed tomography shows a 7-cm cystic mass (arrow) with a thick wall in the anterior aspect of the inferior vena cava.

α- and β-adrenergic blockers, cardiac function was normalized dramatically and his vital signs became stable. He was weaned from the IABP and PCPS on the 10th hospital day. On the 15th day, mass excision was performed and The Histological and immunohistochemical analyses confirmed paraganglioma or extraadrenal pheochromocytoma.

Serum and urinary catecholamines were markedly high initially and became almost normal after the operation. Follow-up echocardiography revealed dramatically normalized cardiac function and chamber dimensions (Figs 1E,F). He was discharged in good condition.

Discussion

The underlying mechanisms of transient LV ballooning syndrome remain unknown. Although several theories have been proposed, including epicardial coronary arterial spasm and microvascular coronary spasm, catecholamine-mediated myocardial stunning may be the central cause of this syn-

drome. Catecholamines are known to decrease the viability of myocytes through cyclic AMP-mediated calcium overload of the cell[4] Catecholamines are also known to generate oxygen-derived free radicals and in an animal model antioxidants attenuated myocyte injury. Because free radicals can interrupt sodium and calcium transport, increased transsarcolemmal calcium influx and cellular calcium overload may result in myocyte dysfunction[5,6] Patients with transient LV ballooning syndrome showed elevated levels of plasma catecholamines, suggesting exaggerated sympathetic activation[7]

Catecholamine cardiomyopathy with takotsubo contractile pattern has been described in pheochromocytoma-induced cardiomyopathy. As a possible explanation for the distinctive contractile pattern in catecholamine cardiomyopathy, there is evidence that the apical myocardium has enhanced responsiveness to sympathetic stimulation, making it more vulnerable to a sudden catecholamine crisis[8,9] Two cases of pheochromocytoma accompanied by inverted takotsubo contractile pattern were reported recently[2,3] The actual ventricular dysfunction patterns in catecholamine-mediated cardiomyopathy reported to date are heterogeneous;[10] so catecholamine-induced myocardial dysfunction may cause both typical and atypical transient LV ballooning

Cardiogenic shock because of a catecholamine crisis is a less commonly recognized manifestation of catecholamine-secreting tumors. Because conventional treatment, including pharmacological and mechanical management (ie, IABP), was not effective in the present patient, PCPS was initiated to maintain systemic circulation. To our knowledge, this is the first case of atypical transient LV ballooning that has been successfully rescued by PCPS and it emphasizes the importance of early aggressive treatment, including mechanical support, to improve the clinical outcome in catecholamine-mediated cardiomyopathy.

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