

Pheochromocytoma-Related Cardiomyopathy Inverted Takotsubo Contractile Pattern

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A 41-year-old woman with no history of cardiac disease or hypertension was admitted to the intensive care unit with acute headache, psychomotor agitation, diaphoresis, nausea, and vomiting. A cerebral computed tomography scan ruled out subarachnoid hemorrhage. The ECG showed sinus tachycardia with ST-segment depression in leads V3–V6, II, III, and aVF. The troponin I level was elevated. The patient continued to have progressive respiratory deterioration, which required mechanical ventilation 24 hours after admission. She also developed 6 episodes of electromechanical dissociation, with circulatory recovery after successful cardiopulmonary resuscitation. Transthoracic echocardiography revealed severe left ventricular dysfunction and a contractile abnormality, consisting of akinesis of the basal and midventricular segments and hyperkinesis of the apical segments (Figure 1 and Movie I in the online Data Supplement). Emergency coronary angiography showed normal coronary arteries (Figure 2). The patient was stabilized after placement of an intra-aortic balloon pump and initiation of dobutamine and noradrenaline infusions, the latter being maintained for 6 days. A computed tomography scan of the abdomen revealed a right adrenal mass, and an α -adrenergic blocker was started, with the presumptive diagnosis of pheochromocytoma. The patient underwent an uncomplicated adrenalectomy on day 16 (Figure 3). Histological and immunohistochemical analyses confirmed the initial diagnosis (Figure 4). Echocardiography repeated on day 14 after surgery showed a left ventricular ejection fraction of 55% (Figure 5 and online Movie II). The patient is alive and remains asymptomatic 2 years after the operation.

Stress-related cardiomyopathy, or Takotsubo cardiomyopathy, is a well-described entity characterized by a typical contractile abnormality consisting of apical and midventric-

ular akinesis or dyskinesis and hyperkinesis of the base. Severe generalized hypokinesis and Takotsubo left ventricular dysfunction have been described in pheochromocytoma-related cardiomyopathy.¹ The physiopathologies of the 2 conditions, stress-induced cardiomyopathy and pheochromocytoma-induced cardiomyopathy, are believed to be similar and mediated by catecholamines.² The patient had severe left ventricular dysfunction with different contractile abnormalities: The basal and midventricular segments were akinetic, whereas the apex showed hyperkinesis. Therefore, there is no unique ventricular dysfunction pattern in catecholamine-related cardiomyopathy.

The mechanism underlying the association between catecholamines and myocardial injury is unknown. Our patient did not show spontaneous coronary vasoconstriction, and coronary flow was normal during cardiac catheterization, which was performed when the patient presented with hemodynamic instability. Therefore, direct myocyte injury was a possible mechanism of myocardial stunning.

Disclosures

None.

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The online-only Data Supplement, which contains 2 movie files, can be found at <http://circ.ahajournals.org/cgi/content/full/113/17/e738/DC1>. Correspondence to Dr Angel Sanchez-Recalde, Unidad de Hemodinámica y Cardiología Intervencionista, Planta 1A Diagonal, Servicio de Cardiología, Hospital Universitario La Paz, Paseo de la Castellana 261, 28046 Madrid, Spain. E-mail asanchezr.hulp@salud.madrid.org (*Circulation.* 2006;113:e738-e739.)

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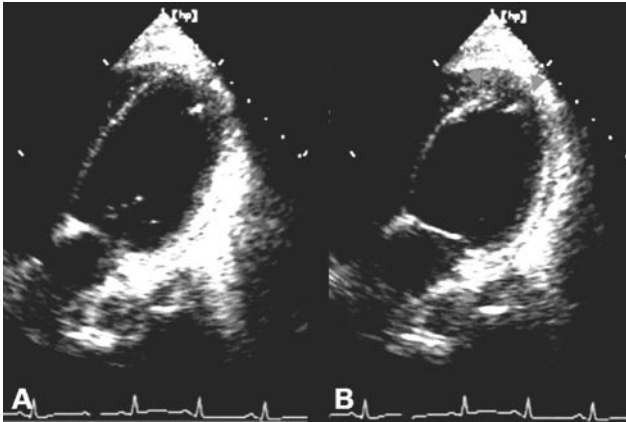


Figure 1. Subcostal long-axis (4-chamber) view, showing akinesis of the basal and midventricular segments, with preserved contractility of the apical segments. A, Diastole. B, Systole.



Figure 2. Coronary angiograms did not show any coronary artery disease.

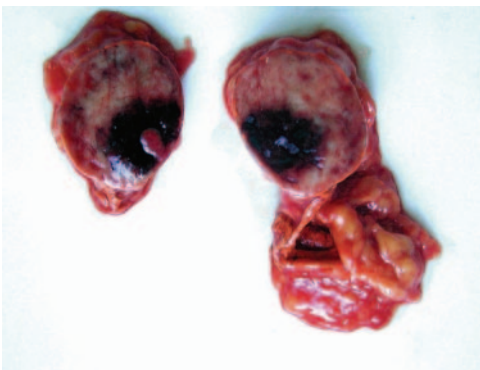


Figure 3. Macroscopic examination of the tumor (2.5×3×2 cm) contained areas of necrosis and hemorrhage within the left adrenal gland.

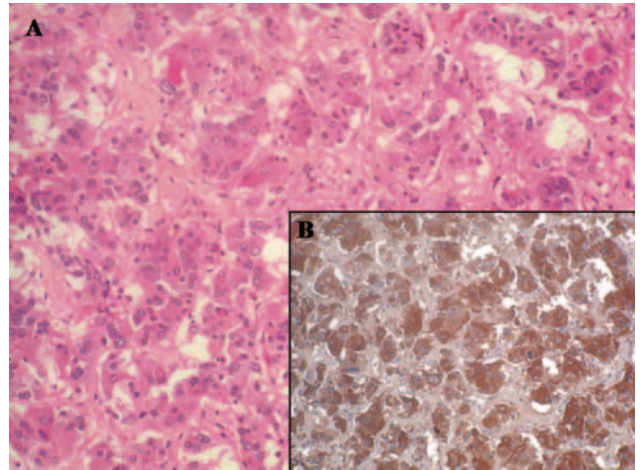


Figure 4. A, Hematoxylin and eosin staining reveals polygonal cells with fibrous tracts between them. B, Chromogranin staining was positive.

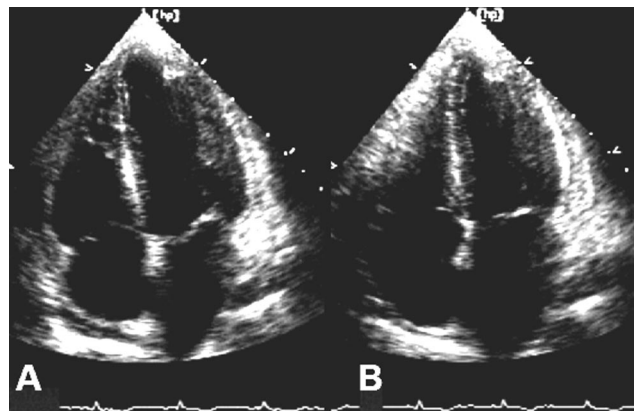


Figure 5. Apical 4-chamber view shows a normal ejection fraction on day 14 after surgery. A, Diastole. B, Systole.

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