

Successful treatment of giant pheochromocytoma–induced Takotsubo syndrome with adrenalectomy

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A hypertensive crisis or an acute increase in diastolic blood pressure (>120 mm Hg) in middle-aged adults with symptoms of palpitation, flushing, sweating, and syncope may suggest renal or endocrine disorders.¹⁻³ The only treatment for pheochromocytoma is resection, and the surgical approach depends on the tumor size.⁴

A 39-year-old woman with a history of 2 episodes of severe hypertension (180/125 mm Hg) treated with captopril (25 mg) and bromazepam (3 mg) was admitted to the emergency department due to heart failure. The current episode was the second admission within 1 month, and the patient was investigated for secondary hypertension by her family physician.

On admission, the patient reported shortness of breath, heart palpitations, general weakness, headache, sweating, and nausea. She was diagnosed with pulmonary edema, and her blood pressure was 85/55 mm Hg. Moreover, supraventricular tachycardia (160 bpm) was noted, and oxygen saturation was 86%. Laboratory findings on admission showed leukocytosis (white blood cell count, $11.24 \times 10^9/l$) and lactic acidosis (pH, 7.238). The levels of troponin I were 817.4 ng/l; lactate, 6.6 mmol/l; and glucose, 17.9 mmol/l.

Computed tomography showed a heterogeneously enhancing mass (size, $107 \times 93 \times 137$ mm) in the left adrenal gland (FIGURE 1A) as well as pectus excavatum (FIGURE 1B). Chest X-ray revealed pulmonary edema (FIGURE 1C). The diagnosis was confirmed by serum levels of free metanephrine (2.092 nmol/l; reference range <0.456 nmol/l)

and normetanephrine (1171 nmol/l; reference range <1.037 nmol/l), measured before hospitalization. Transthoracic echocardiography showed hypercontractility of the basal and apical walls as well as hypokinesia of mid-ventricular walls of the left ventricle with a reduced ejection fraction (35%–40%). The echocardiographic findings suggested Takotsubo cardiomyopathy (the inverted anatomic variant, also known as the “artichoke” heart; FIGURE 1D).

A multidisciplinary team decided on a surgical removal of the pheochromocytoma as soon as hemodynamic stability and full α -blockade were achieved. Intensive care unit (ICU) treatment was started with infusions of norepinephrine (0.15 $\mu\text{g}/\text{kg}/\text{min}$) and dopamine (5 $\mu\text{g}/\text{kg}/\text{min}$). Then, an intra-aortic balloon was inserted, and urgent renal replacement therapy was commenced. The highest troponin I and N-terminal fragment of the prohormone brain natriuretic peptide (NT-proBNP) levels were noted on the second day: 15611.2 ng/l and 1746.7 ng/l, respectively. The intra-aortic balloon was removed on day 9 of hospitalization. Phenoxybenzamine at an uptitrated dose of 20 mg to 240 mg was administered on a daily basis. After complete α -blockade was achieved, labetalol infusion was titrated under close supervision at the ICU. Labetalol is not recommended for patients with pheochromocytoma because nonselective β -adrenergic blockers may cause severe hypertension. However, it was used in our patient, as selective

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Received: September 18, 2019.

Revision accepted:

December 30, 2019.

Published online: January 4, 2020.

Kardiol Pol. 2020; 78 (3): 249-250

doi:10.33963/KP.15122

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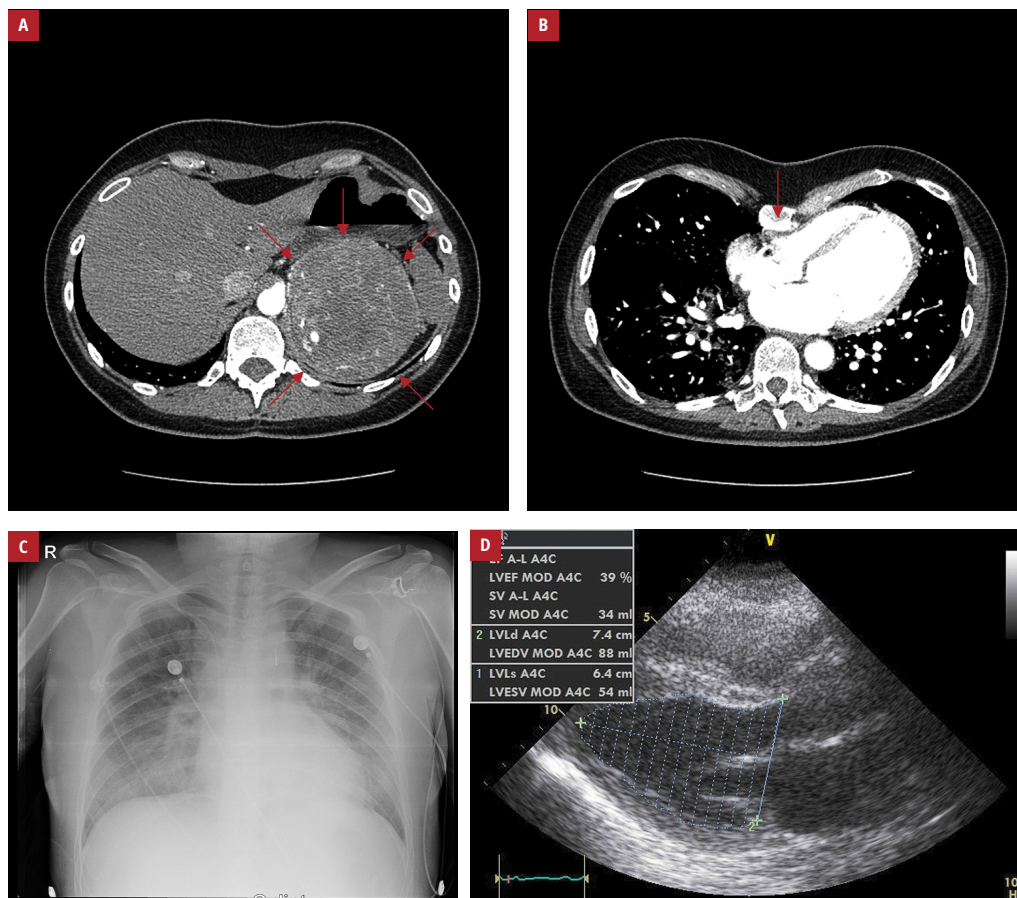


FIGURE 1 **A** – contrast-enhanced computed tomography scan showing a heterogenous left adrenal mass of 10 cm in diameter (arrows), an axial view; **B** – contrast-enhanced computed tomography scan showing the pectus excavatum (arrow); **C** – chest X-ray showing pulmonary edema; **D** – transthoracic echocardiography demonstrating Takotsubo cardiomyopathy

β_1 -adrenoreceptor blockers were unavailable.⁵ Regardless of the tumor size, left laparoscopic adrenalectomy was performed on day 17 of hospitalization, exactly after 10 days of phenoxybenzamine and labetalol treatment.

Initially, anesthesia was successful. Hemodynamic stability was achieved with noradrenaline (0.02 $\mu\text{g}/\text{kg}/\text{min}$), until the rupture of a giant tumor and massive arterial bleeding occurred. Laparoscopy was immediately switched to laparotomy, and thus the dose of vasopressors was dramatically increased. The total blood loss of 2.5 liters lead to hemorrhagic shock, and transfusion of red blood cells (1383 ml), fresh frozen plasma (1037 ml), and platelets (340 ml) was performed.

The postoperative period at the ICU was uneventful, and no vasoactive drugs were necessary. Prior to discharge, troponin I levels decreased to 105 ng/l, and NT-proBNP, to 452 ng/l. As cardiac dysfunction was unrelated to myocardial ischemia, the cardiac prognosis is good.⁴ Histologic examination confirmed the diagnosis of pheochromocytoma, which was 13 cm in diameter and 450 g in weight. The patient was discharged on day 22 of hospitalization after serum free metanephrine and normetanephrine levels had normalized.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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HOW TO CITE Kontrimaviciute E, Valeviciene N, Motiejuniene AB, et al. Successful treatment of giant pheochromocytoma-induced Takotsubo syndrome with adrenalectomy. *Kardiologia Pol.* 2020; 78: 249-250. doi:10.33963/KP.15122

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