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Successful treatment of giant pheochromocytoma-induced Takotsubo syndrome with consequent adrenalectomy conversion

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Short title: Successful treatment of giant pheochromocytoma

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Hypertensive crisis or acute rise in blood pressure (diastolic > 120 mm Hg) in middle-age adults with symptoms of palpitation, flushing, sweating, syncope should remind us about renal or endocrine disorders [1-3]. The only treatment of pheochromocytoma is resection which surgical approach depends on the size of tumour [4].

A 39-year-old woman with a history of two episodes of severe hypertension (180/125 mmHg, treatment with Captoprili 25 mg and Bromazepamum 3 mg were prescribed) was admitted to emergency room due to heart failure. The current episode was the second readmission in a one-month period and she was being investigated for secondary hypertension by her family physician.

On arrival, she reported shortness of breath, heart palpitations, general weakness, headache, sweating and nausea. She was diagnosed with pulmonary oedema, blood pressure was 85/55 mmHg. Also, supraventricular tachycardia 160 BPM was observed, SpO2 - 86%. Lab results upon admission were: troponin I 817.4 ng/l, leucocytosis 11.24*10^9/l, lactoacidosis - pH 7.238, lactate 6.6 mmol/l and glucose level of 17.9 mmol/l.

Computed tomography showed left adrenal heterogeneous enhanced mass measuring 107x93x137 mm (Figure 1A), pectus excavatum (Figure 1B). Chest X-ray revealed pulmonary oedema (Figure 1C). Serum free metanephrine 2.092 nmol/l (URL: < 0,456) and normetanephrine 1171.0 nmol/l (URL: < 1,037) levels (obtained before hospitalization) confirmed the diagnosis. Transthoracic echo was done and showed hipercontractility of basal and apical walls and hypokinesia of mid-ventricular walls of the left ventricle with decreased ejection fraction to 35-40%. The echo was referred as artichoke Takotsubo cardiomyopathy (Figure 1D). Multidisciplinary team of specialists made a decision for surgical removal of pheochromocytoma as soon as hemodynamic stability and full α-blockade will be achieved. Treatment in intensive care unit started with such vasopressor infusions: norepinephrine 0.15 µg/kg/min and dopamine 5 µg/kg/min. After that, intra-aortic balloon
(IABP) insertion was done and urgent renal replacement therapy started. The highest measured level of troponin I and pro-B-type-natriuretic-peptide were present on the second day 15611.2 ng/l; 1746.7 ng/l respectively. IABP was removed on the ninth day of hospitalization. Titration of phenoxybenzamine 20-240 mg was given to the patient on the daily basis. After complete α-blockade was achieved, labetolol infusion was titrated under close supervision in ICU. Labetolol is not recommended for patients with pheochromocytoma as non-selective B-adrenergic blocking drugs may cause severe hypertension. In this case, it was used as selective β1-adrenoreceptor blocking drugs were unavailable [5]. Regardless of tumour size, a surgery of left laparoscopic adrenalectomy was initiated on the seventeenth day of hospitalization, exactly after 10 days of phenoxybenzamine and labetalol treatment.

Initially, anaesthesia was uneventful. Hemodynamic stability was achieved with noradrenaline 0.02 µg/kg/min, until the rupture of giant tumour occurred and massive arterial bleeding started. Laparoscopy was immediately converted to laparotomy thus dosage of vasopressors increased dramatically. The total blood loss of 2.5 litres lead to haemorrhagic shock, transfusion of 1383 ml erythrocyte mass, fresh frozen plasma 1037 ml and thrombocytes 340 ml.

Postoperative period in ICU was successful, no vasoactive drugs were necessary. Prior to patient's discharge, troponin I level decreased to 105 ng/l and BNP 452 ng/l. As cardiac dysfunction was unrelated to coronary ischemia, prognosis for cardiac condition is good [4]. Histological examination confirmed the diagnosis of pheochromocytoma, which was 13 cm in diameter and 450 g in weight. Patient was discharged on the 22nd day of hospitalization when serum free metanephrine and normetanephrine reached normal levels.
REFERENCES


Figure 1. (a) axial computed tomography view with intravenous contrast heterogeneously-enhancing 10 cm left adrenal mass (red arrows); (b) Pectus excavatum (c) Chest X-ray revealed pulmonary oedema; (d) Transthoracic echo demonstrated Takotsubo pattern cardiomyopathy.