

CLINICAL VIGNETTE

Catecholamine-induced secondary Takotsubo syndrome in a patient with pheochromocytoma and synchronous papillary renal cell carcinoma

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A 61-year-old man with a history of hypertension and type 2 diabetes was admitted to the hospital because of acute chest pain and severe dyspnea.

On admission, his blood pressure was 153/103 mm Hg. Initial electrocardiography showed sinus tachycardia at 120 bpm with a significant ST-segment elevation and abnormal Q waves in anterolateral leads. Serum levels of troponin I, creatine kinase, and MB isoenzyme were 10.3 ng/ml (reference range, 0–0.1 ng/ml), 248 U/l (reference range, 0–171 U/l), 17 U/l (reference range, 0–24 U/l), respectively. Urgent coronary angiography showed no significant coronary stenosis. Left ventriculography showed systolic midventricular and apical dyskinesia with basal hypercontraction and reduced left ventricular ejection fraction (LVEF) of 30% (FIGURE 1A and 1B; Supplementary material, Video S1). Transthoracic echocardiography performed 2 days later was remarkable for hypokinesia of the apical and midventricular segments and LVEF of 45% to 50%.

During rehabilitation, the patient experienced an episode of transient loss of consciousness with concomitant tachycardia and refractory, severe hypertension. He was pale and sweaty and complained of occipital headache. His blood pressure was 230 to 270 / 120 to 130 mm Hg, and pulse rate was 120 to 130 bpm. Electrocardiography showed no signs of myocardial ischemia.

The 24-hour urine metanephrine levels were as follows: metanephrine, 10 247.7 µg (reference range, 52–341 µg); normetanephrine, 5395.4 µg (reference range, 88–440 µg); and 3-methoxytyramine, 622.2 µg (reference range, 0–220 µg). The patient's 24-hour urine catecholamine levels were: epinephrine, 416 µg (reference range, 0–20 µg); norepinephrine, 1413.7 µg (reference range, 15–80 µg); and dopamine, 130 µg (reference range, 65–400 µg).

Abdominal magnetic resonance imaging revealed a large (5.6 × 5.3 × 5.3 cm), well-defined, heterogeneous mass in the left suprarenal area, which was suggestive of pheochromocytoma (FIGURE 1C and 1D). Furthermore, in the hilar region of the right kidney, we found an oval (3.6 × 2 × 3 cm), well-circumscribed, solid tumor with heterogeneous T1- and T2-weighted signal intensity (FIGURE 1D).

No clinical evidence of multiple endocrine neoplasia syndromes nor signs of von Hippel–Lindau disease or neurofibromatosis were detected.

An uneventful left laparoscopic adrenalectomy was performed, and histological examination confirmed the diagnosis of pheochromocytoma (Pheochromocytoma of the Adrenal gland Scaled Score [PASS], 4). Subsequently, the patient underwent successful radical right nephrectomy. Histological findings were consistent with type 2 papillary renal cell carcinoma (RCC) (Fuhrman grade 2) locally invading renal

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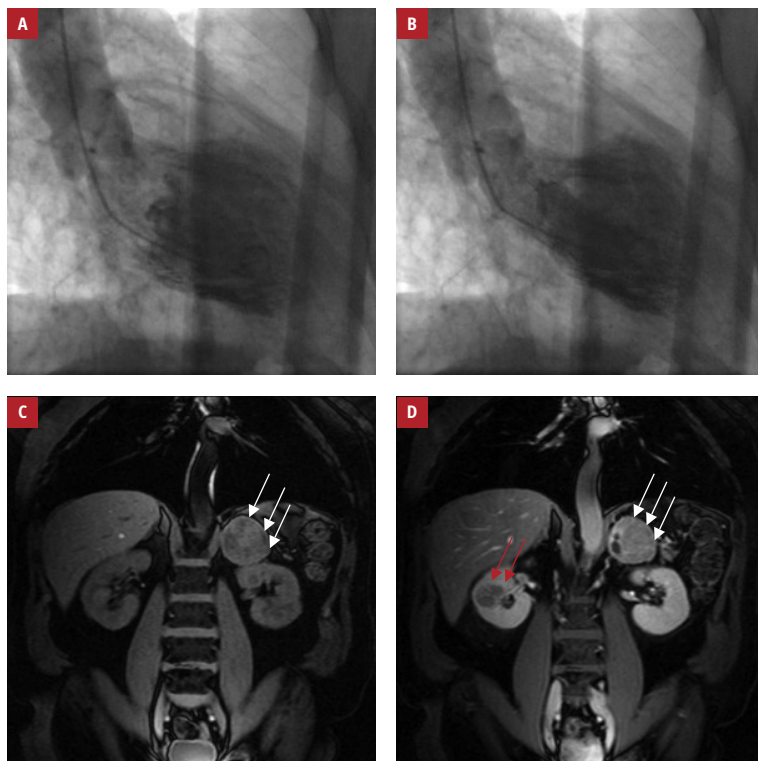


FIGURE 1 Imaging of a patient with pheochromocytoma and papillary renal cell carcinoma: **A, B** – left ventriculography showing systolic midventricular and apical dyskinesia with basal hypercontraction; **C, D** – abdominal magnetic resonance imaging revealing a large (5.6×5.3×5.3 cm), well-defined, heterogeneous mass in the left suprarenal area (white arrows) and an oval (3.6×2×3 cm), well-circumscribed, solid tumor with heterogeneous contrast enhancement in the hilar region of the right kidney (red arrows)

sinus fat (pT3aN0M0). After surgery, the patient recovered completely. Left ventricular contractility further improved and LVEF increased to 60%. At 5-year follow-up, he remained asymptomatic and normotensive, showing no recurrence of cardiovascular symptoms or neoplastic disease.

Here, we presented an extremely rare case of coexistence of 2 uncommon tumors: adrenal pheochromocytoma and contralateral type 2 papillary RCC in a patient with reversible, catecholamine-induced Takotsubo syndrome.

Pheochromocytoma is called “the great imitator” or “the great masquerader” owing to its wide variety of clinical presentations.¹⁻³ Catecholamine-induced secondary Takotsubo syndrome is a rare (incidence <3%) manifestation of pheochromocytoma.¹ The synchronous occurrence of at least 2 primary tumors of different histological types is also an infrequent finding.^{4,5} In a series of 550 patients who underwent radical nephrectomy, 80 (15%) had concomitant renal and adrenal lesions, mainly clear cell RCC and adrenal adenoma/hyperplasia.⁵ No case of synchronous pheochromocytoma and papillary RCC was found in that study.⁵

In conclusion, we believe that the exclusion of pheochromocytoma should be part of

the routine management of all patients with Takotsubo syndrome, especially those with sustained/paroxysmal hypertension and palpitations, headaches, and excessive/abnormal sweating.

SUPPLEMENTARY MATERIAL

Supplementary material is available at www.mp.pl/kardiologiapolska.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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