Inverted Takotsubo cardiomyopathy due to pheochromocytoma

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A 52-year-old woman with history of weight loss and hypertension was referred to our cardiology department with acute typical chest pain, shortness of breath, hypertensive crisis (180/120 mmHg) and headache. The ECG showed sinus tachycardia with ST-segment depression in precordial leads V4–V6. Troponin I level as well as creatin kinase were elevated to 4.06 μg/L (normal <0.01 μg/L) and 266 U/L (normal 38–157 U/L), respectively. Because of these findings and ongoing chest pain the patient underwent emergency coronary angiography that excluded obstructive coronary artery disease (Panels A and B). However, ventriculography revealed a severely depressed left ventricular ejection fraction (LVEF 20%) with akinetic basal and midventricular segments (Panels C and D, arrows) and a hyperkinetic apex (arrowheads), findings consistent with an inverted Takotsubo cardiomyopathy. Twenty-four hours later a transthoracic echocardiography demonstrated an improvement in LVEF (40%). Cardiac magnetic resonance, taken 3 days after admission, revealed a normalized LVEF of 60% without wall motion abnormalities. Given the clinical history, the normal result of the coronary angiography, and the rapid normalization of the heart function, a pheochromocytoma was suspected. Serum total Metanephrine was 51 nmol/L (normal 2.03–4.16 nmol/L) and free Metanephrine was 5 nmol/L (normal 0.06–0.61 nmol/L). Abdominal computer tomography revealed a right adrenal mass. After pretreatment with alpha and beta blockade the patient underwent an uncomplicated adrenalectomy (Panel E). Histological and immunohistochemical staining confirmed the diagnosis of pheochromocytoma (Panels F and G). The postoperative course was uneventful.

Panels A and B. Shows normal right and left coronary arteries.
Panels C and D. End-systolic (C) and end-diastolic (D) left ventriculography showed akinesia of the basal and midventricular segments (arrows) and apical hypercontractility (arrowheads).
Panel E. Right adrenal gland. The tumour (4 x 3 x 2.5 cm³) contains areas of necrosis and haemorrhage (arrows).
Panel F. Hematoxylin and eosin staining reveals polygonal cells with fibrous tracts.
Panel G. Positive chromogranin staining.