Case Report

Reversible Posterior Leukoencephalopathy Syndrome and Takotsubo Cardiomyopathy: The Role of Echocardiographic Monitoring in the ICU

John Papanikolaou, Maria Tsirantonaki, George Koukoulitsios, Dimitrios Papageorgiou, Christina Mandila, Dimitrios Karakitsos, Andreas Karabinis

Intensive Care Unit, General Hospital of Athens "G. Gennimatas", Athens, Greece

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We report a case of a female patient with acute renal failure due to *polyarteritis nodosa*. Her clinical course was initially complicated by an unusual form of hypertensive encephalopathy called reversible posterior leukoencephalopathy syndrome (RPLS). Soon afterwards she developed cardiogenic shock; she was intubated and admitted to our ICU. Echocardiography and pertinent laboratory data were suggestive of takotsubo cardiomyopathy (TTC), a rare form of stress-induced, reversible cardiac dysfunction. We hypothesized that TTC was pathophysiologically linked to RPLS, presumably through an overstimulation of the sympathetic nervous system. Both RPLS and TTC turned out to be totally reversible conditions, and intensive echocardiographic monitoring was of great importance in order to optimize the hemodynamic support in our patient.

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Address:
John Papanikolaou

80, 17th November 1973 St.
15562 Holargos
Athens, Greece
e-mail:
y_papanikolaou@hotmail.com

Reversible posterior leukoencephalopathy syndrome (RPLS) has recently been described as a specific form of hypertensive encephalopathy that is strongly associated with renal disease, vasculitis, eclampsia and immunosuppressive treatment.[1,2] It is characterized by typical transient clinical (headache, altered mental status, abnormal visual perception and seizures) and neuro-radiological findings (white matter abnormalities on magnetic resonance imaging [MRI], mainly located in the parietal-occipital areas).[3,4]

It is well known that the central nervous and cardiovascular systems are closely related, and a recent example is takotsubo cardiomyopathy. The latter is a recently described clinical entity, presenting as transient myocardial dysfunction due to tremendous emotional and/or physical stress. Sympathetic nervous system overstimulation seems to participate in the development of this response.[5]

We present a very rare case in which acute renal failure led to RPLS and takotsubo cardiomyopathy. The role of ultrasound monitoring in the diagnosis and the therapeutic interventions is also thoroughly analyzed.

Case presentation

A 47-year-old postmenopausal white woman was admitted to the Nephrology Department of our Hospital, presenting acute renal failure (urea 28 mg/dl, creatinine 3.5 mg/dl) and excessive hypertension (200/120 mmHg). She had been suffering from abdominal pain and fever for about two months; these symptoms were attributed to possible cholecystitis and a cholecystectomy had been performed a few days before the admission. On the second day of hospitalization, the patient started to complain of severe headache. Soon, she presented three episodes of total visual loss (the third one persisted for 24 hours), while in the meantime she manifested two epi-

sodes of witnessed seizures. MRI revealed hyperintense signal areas in the posterior brain regions (occipital-parietal white matter), which were typical of RPLS (Figure 1a). The patient was put on intravenous nitrates and anticonvulsive therapy. A few hours later, she complained of precordial discomfort and exhibited dyspnea, diaphoresis, hypotension and systemic hypoperfusion (oliguria, altered mental status). A chest X-ray showed severe pulmonary congestion (Figure 1b) and the diagnosis of cardiogenic shock was established. The patient was intubated and mechanically ventilated, put on inotropic support and transferred to the ICU. The initial electrocardiogram (Figure 1c) did not reveal any specific findings compatible with acute coronary syndrome. Echocardiography showed global left ventricular hypokinesia, especially localized in the median portions and apex (Figure 1d), with severe diastolic dysfunction (restrictive left ventricular filling pattern) (Figures 1e & 1f). The combination of almost normal cardiac enzymes (troponin I just slightly elevated) and the severe myocardial dysfunction, diagnosed by continuous echocardiographic monitoring, was more compatible with the diagnosis of takotsubo cardiomyopathy rather than myocardial infarction. As the patient showed severe hemodynamic instability and multi-organ failure, a conservative therapeutic strategy was adopted, and the patient did not undergo coronary angiography during the acute phase of the disease. Acute respiratory failure was managed with mechanical ventilation techniques (pressure-regulated volume control ventilation), while inotropes (dobutamine) and vasoactive agents (norepinephrine and vasopressin) were used to support tissue hypoperfusion. Acute re-

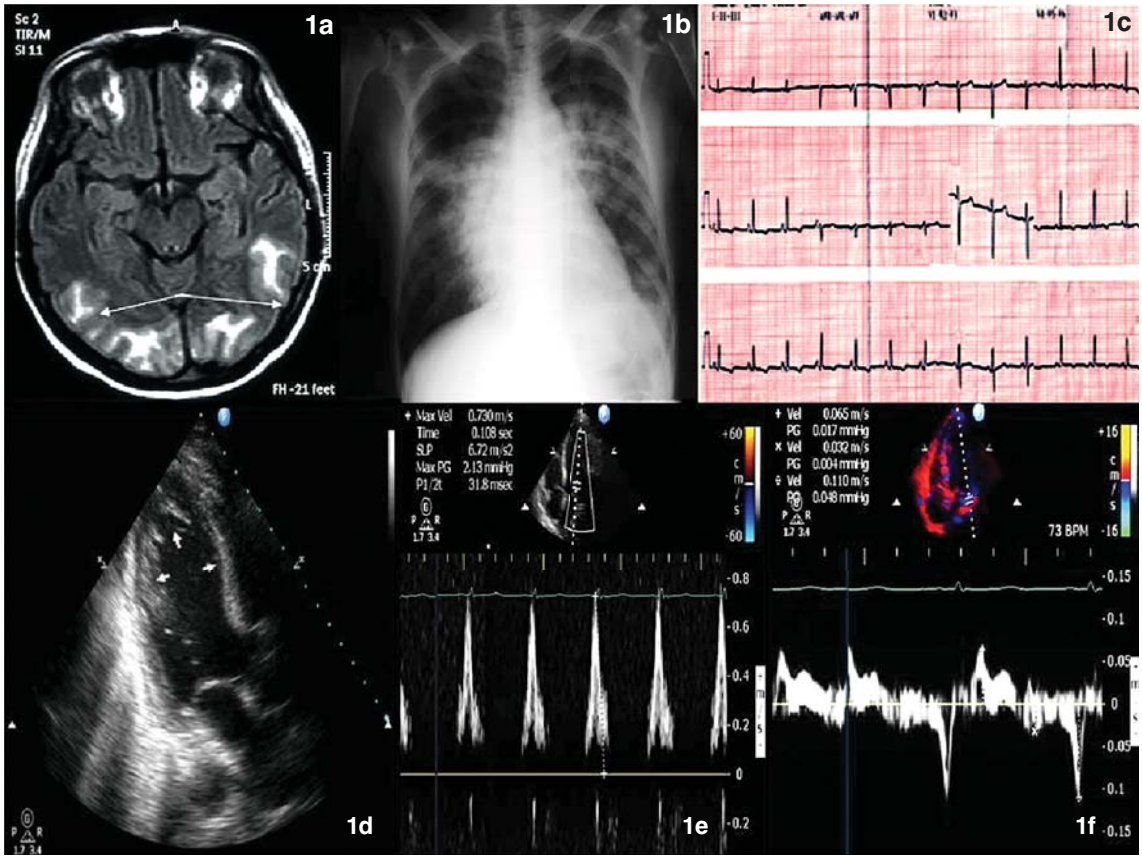

Figure 1. a: Magnetic resonance imaging showed hyper-intense lesions in white matter of both parietal-occipital regions, which are typical of reversible posterior leukoencephalopathy syndrome (white arrows). b: The portable chest X-ray upon admission shows acute pulmonary edema. c: The ECG showed a rhythm of about 75 beats/min, pulmonary P waves in leads II, III, aVF, and negative T waves in the inferior and lateral wall recordings. d: A modified apical long-axis systolic frame manifests significant left ventricular apical ballooning (arrowheads). e, f: Pulsed wave transmitral flow Doppler signal (E/A >2, EDT 110 ms) and tissue Doppler imaging of the lateral mitral annulus (Em ≤Am, E/Em 25) show evidence of severe diastolic left ventricular dysfunction (restrictive filling pattern).

nal failure and volume overload were dealt with by continuous renal substitution therapy (continuous venous-venous hemodiafiltration) and the left ventricular filling pressures were optimized after repeated and careful echocardiographic evaluation of the systolic and diastolic heart function.

The patient's clinical status (cardiovascular and neurological) improved dramatically thereafter. Repeated cardiac ultrasound scans also showed an impressive improvement in systolic as well as in diastolic cardiac function. After a week of hospitalization her cardiac function was totally restored. At that point, the biopsy specimens taken from the gallbladder during cholecystectomy turned out to be positive for *polyarteritis nodosa*.

Discussion

Severe hypertension *per se* represents a high-loading condition that may dynamically affect cardiac performance.[6] However, in this specific case we assumed that RPLS as a result of uncontrolled hypertension, together with vasculitis-induced acute renal failure, was the principal pathophysiological mechanism for the development of takotsubo cardiomyopathy (presumably through catecholamine hyper-secretion). In this case, continuous ultrasound monitoring was of major importance for the evaluation of the patient's cardiovascular status and arriving at the correct diagnosis and therapy.

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