A Rare Association of Systemic Lupus Erythematosus, Morbid Obesity and Takotsubo Syndrome

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The authors report a 68-year-old white female with long-term systemic lupus erythematosus as well as morbid obesity, characterized by very elevated body mass index, who presented a classical picture of acute coronary failure ascribed to Takotsubo syndrome.

Takotsubo syndrome, which has been recently described by Japanese authors, is characterized by a reversible abnormality of the ventricular wall movement, with a morphological aspect similar to a balloon, or more precisely, from a semantic point of view, to an amphora-like octopus trap, at the apical segment levels of the heart and hypercontraction of the basal segments observed during the coronary arteriography with ventriculography, associated with electrocardiographic ST-T segment alterations, similar to an acute myocardial infarction episode, with minimal elevation of cardiac enzymes, affecting preferentially elderly females and being induced by physical or emotional stress[1]. The diagnosis of this entity is reinforced by the almost complete absence of coronary circulation morphostructural alterations. Another aspect that is noteworthy is the rapid duration of the asynergy of the ventricular wall movement, eventually contrasting with the longer duration of the clinical manifestations. The etiology of the disease is not fully known, and the role of coronary microcirculation involvement at a multivascular level (severe expression of microvascular angina), and, more recently, the possibility of the participation of altered adrenergic catecholamine dynamics at myocardial level has been hypothesized[2,3].

The exceptional circumstances of the case reported here are due to the simultaneous presence of two conditions that are potentially harmful to the heart, i.e., systemic lupus erythematosus and morbid obesity, associated with this unusual form of cardiomyopathy induced by acute stress. Pericardial and endocardial alterations, vasculitis of the coronary circulation as well as congenital heart block in neonatal lupus briefly constitute the classical cardiac alterations of this auto-immune disease. The impact of morbid obesity on the cardiovascular system is basically caused by the effect of systemic arterial hypertension, dyslipidemia, diabetes and hypoxemic pulmonary arterial hypertension, being broadly known aspects of the disease in the specialized literature.

Key words

Systemic lupus erythematosus,cardiopatithy obesity, Takotsubo syndrome.

Case Report

The patient was a sixty-eight year-old female patient with long-term systemic lupus erythematosus, who had presented acute symptoms such as joint pain, lassitude, thrombocytopenia and acute alveolitis in the last two years, being the latter symptom confirmed by a spiral CT of thorax of multiple areas of ground glass opacities in the most dependent portions of the lower lobes. Treatment was carried out with methylprednisolone pulse therapy associated with oral cyclophosphamide. As a consequence of long-term corticosteroid use, there was a progressive increase in weight gain, with BMI reaching levels that were compatible with morbid obesity associated with alveolar hypoventilation syndrome (a pickwickian syndrome) as well as diabetic status onset in the subsequent months. When the patient had been discharged from the hospital and was at home after remission of the last crisis,, following intense physical exertion caused by an attempt to rise without help after a fall in one of the rooms, she presented non-specific chest discomfort, cyanosis, dyspnea and acute pulmonary edema. She was transferred to the Intensive Care Unit (ICU), where she was treated with i.v. diuretics and morphine. The hospital admission electrocardiogram was normal (Fig. 1) and the curves of cardiac enzymes and brain natriuretic peptide (BNP) were normal, as shown in Charts 1 and 2. The Doppler echocardiogram (Fig. 2A) showed akinesia and ballooning of the apical portions and hyper-contractility of the basal portions of the left ventricle. The coronary arteriography performed less than 14 hours after hospital admission disclosed coronary artery tree without significant obstructive alterations, as well as classic alterations of basal hypercontraction and dyskinesia in the ventricular free wall shaped as an octopus trap, characterizing the Takotsubo syndrome (Fig. 3).

There was regression of the alterations after three days, with the disappearance of pulmonary edema and the appearance of ventricular repolarization alterations (Fig. 4), as well as normalization of cardiac enzymes, with complete regression ten days after the initial picture started.

The contractile deficit of the ventricular wall disappeared in four days (Fig. 2B) and a myocardial scintigraphy with metaiodobenzylguanidine (MIBG) performed five days later disclosed the compromised cardiac adrenergic

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Received on 06/29/05 • Accepted on 12/08/05

neurotransmission, with a segmental denervation area in the apical and anterior walls of the left ventricle, which subsequently disclosed improvement at the scintigraphy performed on October 15 of the same year, showing that the involvement was restricted to the anterior wall (Figs. 5 and 6).

Discussion

When the current Japanese cardiologic literature is studied, several references of the Takotsubo syndrome are found in the last years, in opposition to the scarcity of reports in the western literature[4-6]. Notwithstanding the apparent benign manifestation of this process, authors eventually mention clinical evolution with acute pulmonary edema, cardiogenic shock, ventricular rupture and death[7]. High concentrations of plasma noradrenaline seem to indicate the possibility of the participation of altered catecholamine dynamics[8]. Other authors believe the genesis of this entity is a diffuse microvascular ischemic insult.

This case report is noteworthy due to the concomitant participation of three entities that are potentially harmful to the heart, i.e., morbid obesity, systemic lupus erythematosus and steroid diabetes, associated with the appearance of the Takotsubo syndrome. The scintigraphy aspects have been the object of current extensive studies. The myocardial scintigraphy utilizing meta-iodine-benzylguanidine (MIBG) with iodine^{123}-labeled metaiodobenzylguanidine performed in patients with myocardiopathies of Takotsubo type has frequently demonstrated a specific pattern of sympathetic-adrenergic hyperactivity with preservation of the coronary flow, which is very suggestive of a neurogenic stunned myocardium with myocardial adrenergic receptor hyperactivity, being the last event in the commencement of this reversible myocardiopathy[9].

It is also worth mentioning the presence of elevated BNP levels during the acute picture, without an unfavorable prognostic significance, and apparent and directly related to the severity of hyperkinesis of the basal segments[10].

The authors present a rare case of association between systemic lupus erythematosus, morbid obesity and Takotsubo cardiomyopathy. Despite the lack of certification or exclusion of coronary spasm through intracoronary infusion of acetylcholine, the evolution findings at the myocardial scintigraphy with iodine^{123}-labeled metaiodobenzylguanidine analyzed together with the clinical, electrocardiographic, echocardiographic and coronary scintigraphy with ventriculography findings allowed the precise diagnostic procedure. It is worth mentioning that iodine^{123}metaiodobenzylguanidine represents a noradrenaline analog and it is actively transported within the noradrenaline granules of the sympathetic nerve terminals by uptake-1.

The decreased MIBG uptake at the initial phase consequently suggests the involvement of the cardiac autonomic nervous system. The cardiomyopathy in this case, also called transient apical dyskinesia, represents a type of acute coronary syndrome that is quite similar to an acute myocardial infarction (AMI) episode due to the clinical and electrocardiographic picture, and has the potential for early complications that are even more significant than those observed in AMI, demonstrated by the presence of acute pulmonary edema in 22%, ventricular arrhythmia in 9% and cardiogenic shock in 15% of the patients, although presenting an excellent middle and long-term prognosis.

The prevalence of the disease in middle-aged or elderly female patients, having as triggering factors psychological stress, unusual physical exertion, worsening of preexisting diseases or the appearance of new ones, either clinical or surgical, is a clinical aspect worth remembering, and which was found in the reported case.

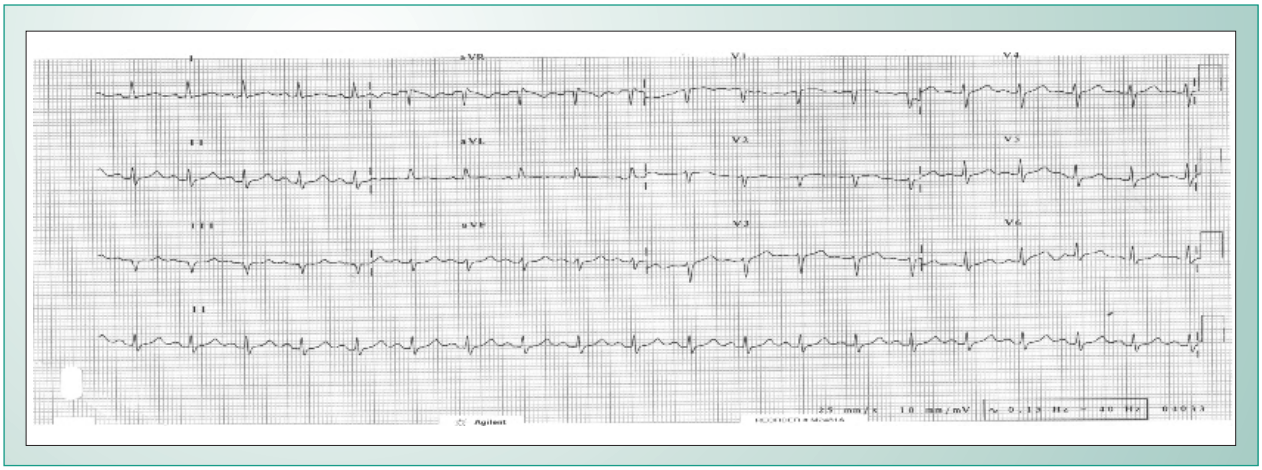


Fig. 1 - *Hospital admission ECG within normal parameters.*

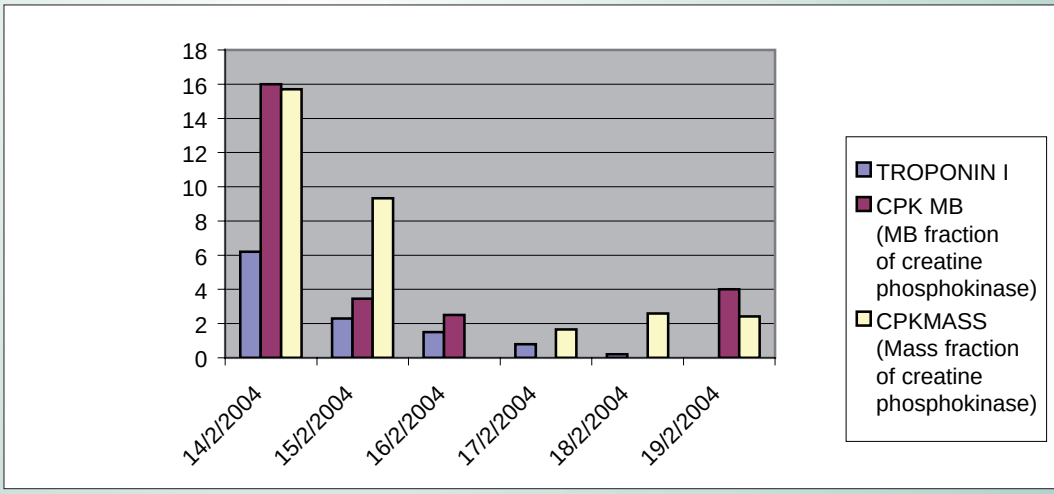


Chart 1 - *Evolution of cardiac enzymes.*

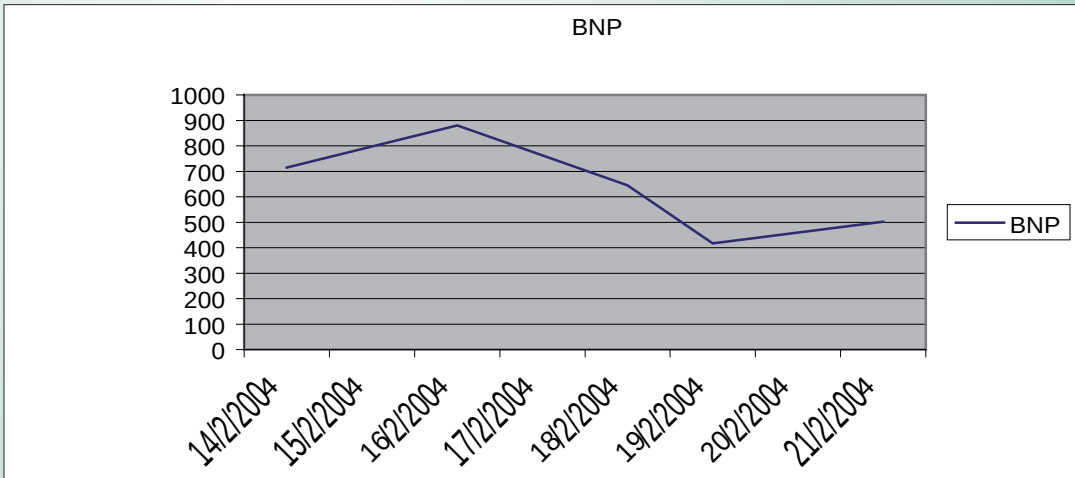


Chart 2 - *BNP curve during hospital stay.*

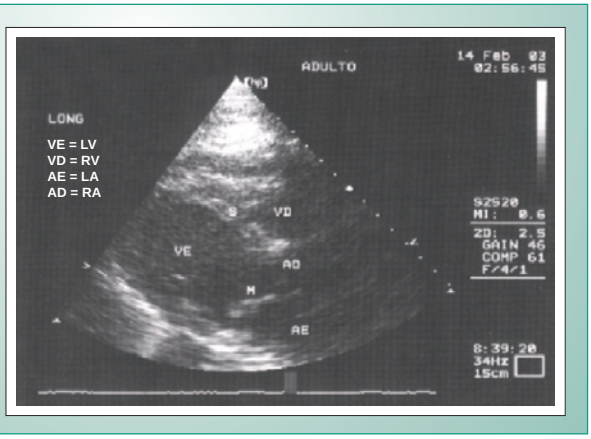


Fig. 2A - *Akinesia and ballooning of the apical portions and hypercontractility of the basal portions of the left ventricle.*
M - *mitral valve;* S - *interventricular septum;* LV - *left ventricle;* RV - *right ventricle;* LA - *left aorta;* RA - *right aorta.*

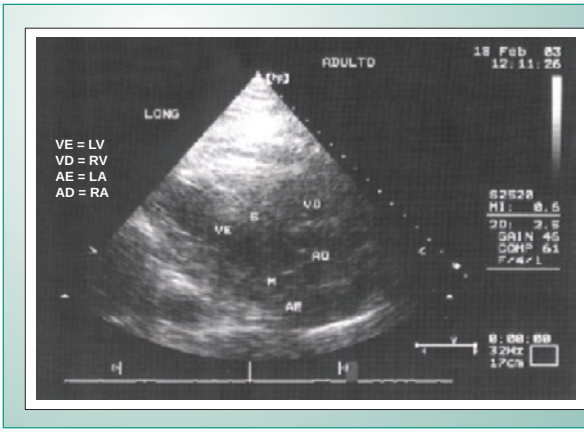
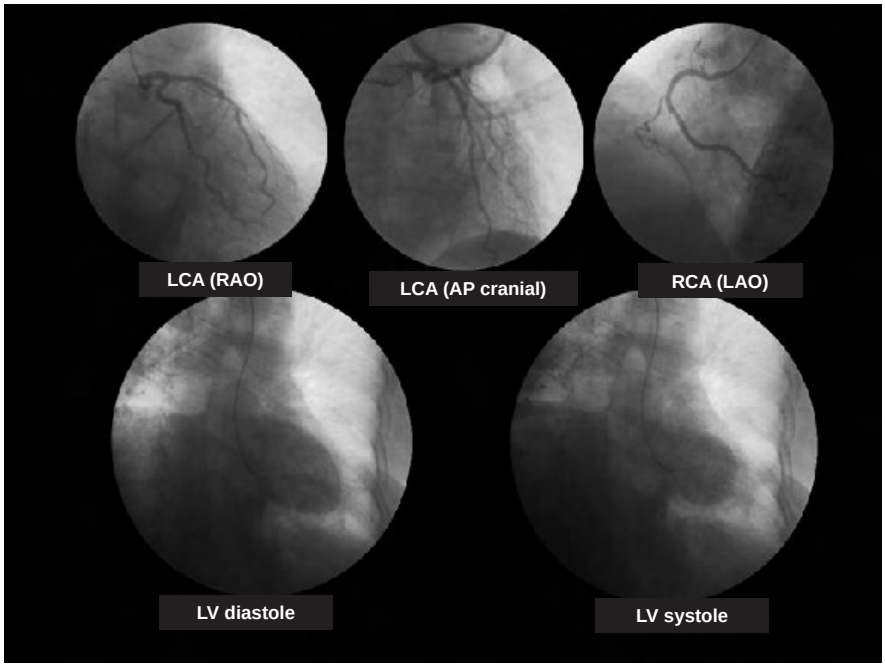


Fig. 2B - *Complete regression of the described alterations.*
M - *mitral valve;* S - *interventricular septum;* LV - *left ventricle;* RV - *right ventricle;* LA - *left aorta;* RA - *right aorta.*



LCA- Left coronary artery; RAO- Right anterior oblique view; RCA- Right coronary artery; LAO- left anterior oblique view; AP - anteroposterior; LV - left ventricle.

Fig. 3 - *Coronary scintigraphy disclosing slight parietal alterations and left ventriculography with akinesia and ballooning of the apical portion and hypercontractility of the basal portions of the left ventricle.*

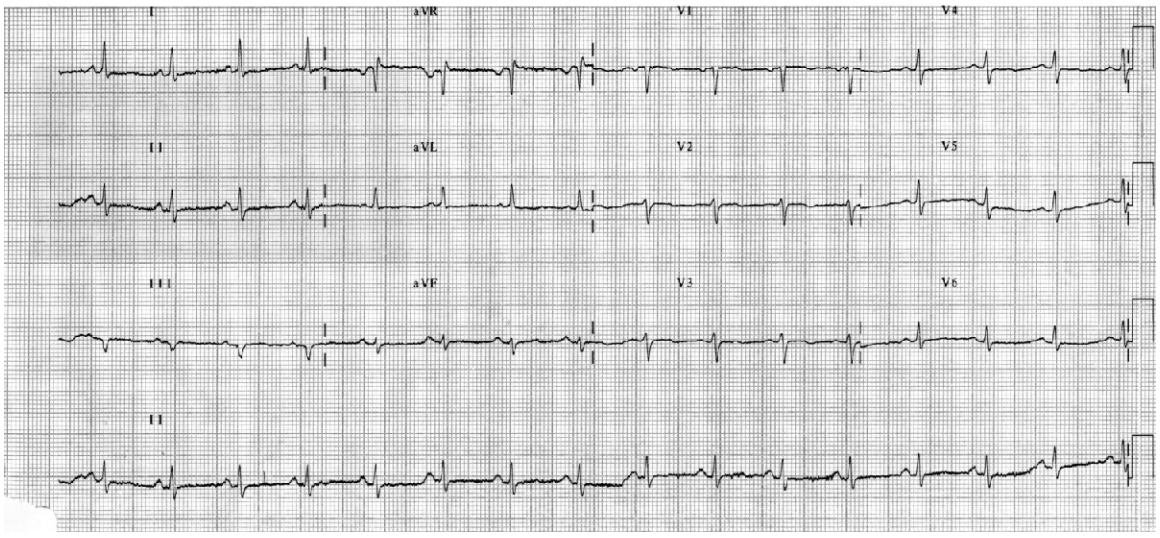


Fig. 4 - *ECG performed on the 4th day of hospital admission, showing diffuse alterations of ventricular repolarization.*

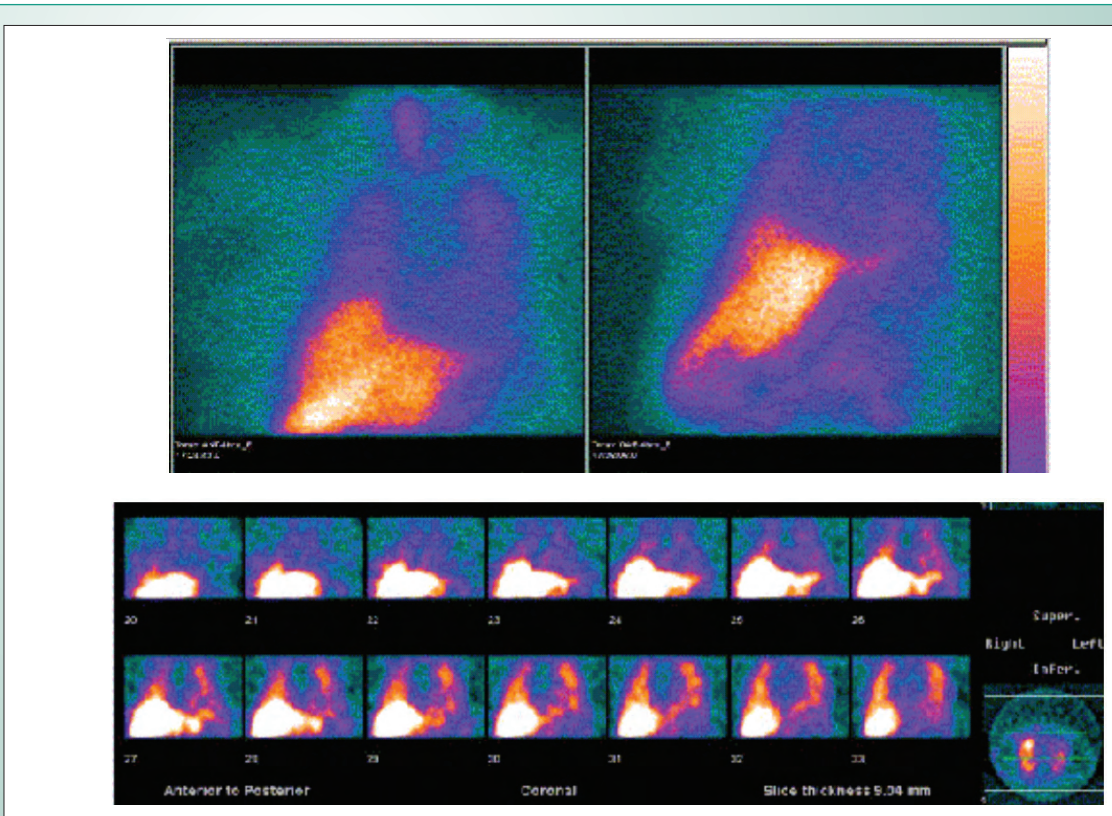


Fig. 5 - *The planar images (Feb./2003) with 123I-MIBG, at the anterior and left anterior oblique views, show the activity of the radiotracer almost restricted to the liver. The accentuated decrease in MIBG in the heart corresponds to a significant involvement of cardiac adrenergic neurotransmission.*
Spect performed in February/2003 showing intense activity in the liver and pulmonary fields. The heart is little visualized.

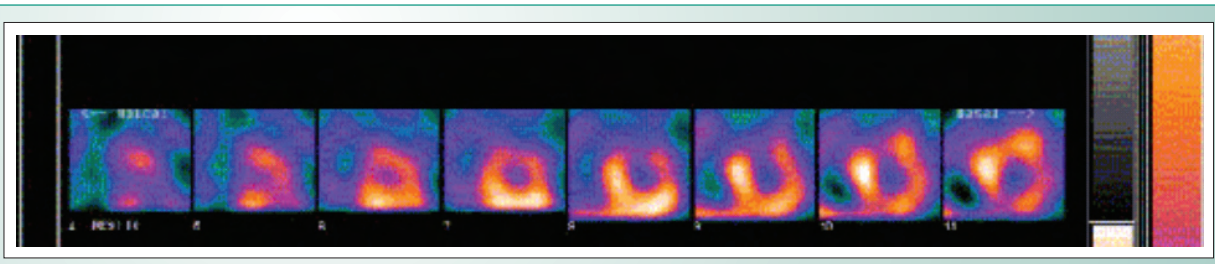


Fig. 6 - *Spect performed in October/2003 showing low uptake of the radiotracer in the apical and anterior segments of the heart, although there is an improvement when compared to the previous one.*

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