Two cases of tako-tsubo cardiomyopathy in Caucasians

Pasquale Assennato, Rosa Alfano, Giuseppina Novo, Giovanni Fazio, Rosanna Zito, Dalila Fernandez, Gabriella Carlino, Loredana Sutera, Marco Fazio, Enrico Hoffmann, Salvatore Novo

Department of Cardiology, University of Palermo, Palermo, Italy

Introduction

Tako-tsubo cardiomyopathy is a recently described disease characterized by chest pain, transient left ventricular dysfunction and specific electrocardiographic changes. The disease takes its name from the typical left apical ballooning observed at left ventriculogram. Tako-tsubo cardiomyopathy was first described by Sato in 1990. Since then sporadic cases were reported by Japanese authors, and only a few European publications are available. We describe 2 cases of patients affected by this syndrome.

(ECit Heart J 2005; 6 (7): 614-617)

© 2005 CEPI Srl

Received September 23, 2004; revision received March 2, 2005; accepted March 4, 2005.

Address:
Dr. Giovanni Fazio
Via S. Maria del Gesù, 25
90124 Palermo
E-mail: Giovanni.fazio-aaa@poste.it

Description of cases

Case 1. A 67-year-old female patient was admitted for epigastric pain. She did not show any cardiovascular risk factors. The patient’s clinical history revealed an episode of superficial phlebitis at the right inferior limb, 1 month before admission; moreover, a few days prior to admission she had a car accident described as a very emotionally stressful event. At admission she was in good clinical conditions. The clinical examination was negative.

ECG showed ST-segment elevation in DI and aVL and T-wave inversion in all leads (Fig. 1). Blood tests revealed normal myocardial enzyme levels and an increase in inflammatory indices (erythrocyte sedimentation rate, C-reactive protein, and beta-2-globulins). We did not detect any virus causing myocarditis. An increased level of vanilmandelic acid was detected. During in-hospital stay, ECG showed a progressive deepening of negative T wave in leads that explore the lateral segments of the left ventricle. Echocardiography, at that time, showed a global LV akinesis with basal regional hyperkinesis and an ejection fraction of about 40%. The patient underwent a cardiac catheterization that showed normal coronary arteries and an enlarged left ventricle with global akinesis and basal hyperkinesis (ejection fraction 40%), according to echocardiographic data. Figure 2 shows the characteristic angiographic image known in the literature as “tako-tsubo”. At discharge a treatment with enalapril and diuretics was prescribed.

One month later the patient was readmitted for clinical and instrumental evaluation. The patient was in good clinical conditions. Blood tests confirmed the absence of viral myocarditis. The level of vanilmandelic acid was within normal limits. An echocardiogram showed complete global and segmentary kinesis normalization and an improved cardiac function (ejection fraction 70%).
Case 2. A 69-year-old female patient was admitted for chest pain. She smoked 20 cigarettes/day, and she was obese. The patient has been affected by bilateral hypoacusia for 2 years. Since the last month she was suffering from some episodes of effort dyspnea. It is worth noting that in this patient too, an emotional stress event (the premature death of her husband, occurred only 7 days before admission) was revealed at clinical history. At admission, she was in good clinical conditions.

ECG showed a third degree atrioventricular block with ventricular escape rhythm and heart rate of 30 b/min (Fig. 3). For this reason a pacemaker (DDD mode) was implanted.

During the day of implantation the patient was suffering repeated episodes of dyspnea and chest pain, with profuse perspiration. Pulmonary examination showed basal lung stasis. Blood tests revealed normal myocardial enzyme levels and increased inflammatory indices (erythrocyte sedimentation rate, C-reactive protein, beta2-globulins). We did not detect any virus causing myocarditis. In this case too, we found an increased level of vanilmandelic acid. Echocardiography at that time showed a global LV akinesis with basal regional hyperkinesis (ejection fraction 45%). Cardiac catheterization showed normal coronary arteries and an enlarged left ventricle with global akinesis and basal hyperkinesis and ejection fraction of 45%. Figure 4 shows the angiographic characteristics of tako-tsubo syndrome. At hospital discharge a treatment with ramipril and atenolol was prescribed. One month later the patient was readmitted for a check-up. She was in good conditions. Blood tests confirmed the absence of viral myocarditis. Vanilmandelic acid level was within the normal limits. An ECG showed regular sinus rhythm. Again, echocardiography showed a complete global and segmentary kinesis normalization and an improved cardiac function (ejection fraction 75%).

Discussion

Reversible or quite reversible ventricular dysfunction usually related to myocardial stunning is often observed in ischemic heart disease. This dysfunction is accompanied by a coronary event16 and by an increase in specific myocardial enzymes. In our cases creatine kinase, creatine kinase-MB and troponine levels did not increase. Moreover, although in both patients, echocar-
diographic and angiographic regions of akinesis were observed, they did not correspond to a specific coronary-related territory and coronary angiogram was normal. According to these findings we rejected the diagnosis of ischemic heart disease in our patients.

Another cause of transient LV dysfunction\(^{17}\) is myocarditis. Only a myocardial biopsy allowed us to exclude clearly the diagnosis of infective myocarditis. However, the extent of akinesis, the blood tests not showing a viral infection, the disease that was clearly related to an emotional stress (car accident in the first case and husband death in the second) and the very fast recovery, characteristics absolutely similar to those reported by authors who published reports concerning the tako-tsubo disease, convinced us to consider this latter diagnosis. Subarachnoid hemorrhage\(^{18,20}\), pheochromocytoma\(^{21,22}\), Guillain-Barré syndrome\(^{23}\) and pneumothorax\(^{24}\) were also reported as possible causes of diffuse reversible ventricular dysfunction. However, these disorders were not demonstrated in our cases.

Moreover, according to the literature, in our patients cardiomyopathy occurred in elderly women, with negative anamnesis for cardiovascular diseases and the first symptom was a chest pain episode, following an emotional stress. At the beginning, ECG showed ST elevation in all leads and after that, T waves become negative\(^{25}\). Imaging tests, such as echocardiography and angiography, showed a large akinetic area, that did not correspond to a specific coronary artery-related region, and hypercontraction of the basal segment\(^{4}\). Coronary angiography showed epicardial artery uninjured.

This reversible asynchrony is typical of tako-tsubo cardiomyopathy and in fact it gave the name to the disease.

In 2001, Owa et al.\(^{26}\) proposed a role of the autonomic nervous system and catecholamine in the pathogenesis of tako-tsubo cardiomyopathy. In 2002, Kyuma et al.\(^{25}\) reported an improvement in LV function after treatment with beta-blockers (propranolol).

In our patients we found an increase in vanilmandelic acid urine concentration, during in-hospital stay and normalization at one-month follow-up. An increase in catecholamine levels in our patients may be due to the emotional stress experienced some days earlier\(^{24,27,29}\).

As for beta-blocker therapy, only the first patient received atenolol, but both showed an improvement in LV kinesis. By demonstrating the role of catecholamine in

---

**Figure 3.** Case 2 electrocardiogram.

**Figure 4.** Angiographic case 2 images showing left apical ballooning typical of tako-tsubo: proto-systolic (A) and end-systolic (B) frames.
the disease etiology and considering the scintigraphic findings of patients described in some literature’s reports, probably it should be more correct to classify the tako-tsubo syndrome as a toxic myocarditis instead of a cardiomyopathy.

Regarding possible disease-related complications, Akashi et al. reported a case in which they found some episodes of polymorphic ventricular tachycardia; in our second patient we found a third degree atrioventricular block. For this reason aDDD mode pacemaker was implanted. When clinical and echocardiographic patterns had been normalized, Holter ECG did not show any pacemaker activation.

This finding consolidates the idea that this disease is a toxic myocarditis.

In conclusion, the tako-tsubo cardiomyopathy was first described in 1990 to explain transient clinical, ECG and echocardiographic myocardial abnormalities. Nowadays, although only a few cases have been described in the literature, we believe that this condition is not so rare and many diagnoses are missed since it is still largely unknown. We hope that the description of these 2 cases may give an even small contribution to the characterization of this disease.

References
