JBR–BTR, 2011, 94: 214-216.

TAKOTSUBO CARDIOMYOPATHY OR APICAL BALLOONING: A CASE REPORT AND A SHORT LITERATURE STUDY

T. De Beule[1], L. Ardies[1], M. De Booij[2], E. Janssens[1], P. Vanhoenacker[1]

Takotsubo cardiomyopathy is an important differential diagnosis in patients with an acute coronary syndrome. This syndrome is typically characterized by a reversible contractile dysfunction of the left ventricular myocardium without any obstructive etiological coronary disease. This is not a rare entity in acute settings and a better knowledge of the syndrome could result in a heightened alertness and a higher detection. We present a case of takotsubo showing the importance of cardiac MRI in the differential diagnosis and reviewed the present literature concerning this syndrome and the usefulness of MRI in the detection.

Key-word: Heart, diseases.

Transient left ventricular apical ballooning is an acute reversible heart syndrome also named takotsubo cardiomyopathy. The characteristic appearance consists of a round bottom and narrow neck on the end systolic ventriculogram resulting from a regional wall motion abnormality that extends beyond a single coronary vascular bed (1, 2). This syndrome is defined by the clinical sudden onset of acute chest pain simulating an acute myocardial infarction often induced by a psychologic stressor. However, the major difference between takotsubo and an acute myocardial infarction consists of a lack of significant coronary stenosis in takotsubo. This chest pain is accompanied by ST-T changes on electrocardiography and minimal raise of the cardiac enzyme concentration in the peripheral blood (3). Takotsubo is predominant in women and often triggered by an emotional or physical event (4). The combination of the clinical syndrome comparable to acute myocardial infarction, minimal raise in cardiac enzymes and the reversible apical hypokinesia extending the normal vascular territories should raise suspicion for takotsubo (5). The pathophysiologic mechanism is unkown, but catecholamine excess tends to have a central role. The syndrome is not rare, and heightened awareness could likely lead to a higher reported incidence.

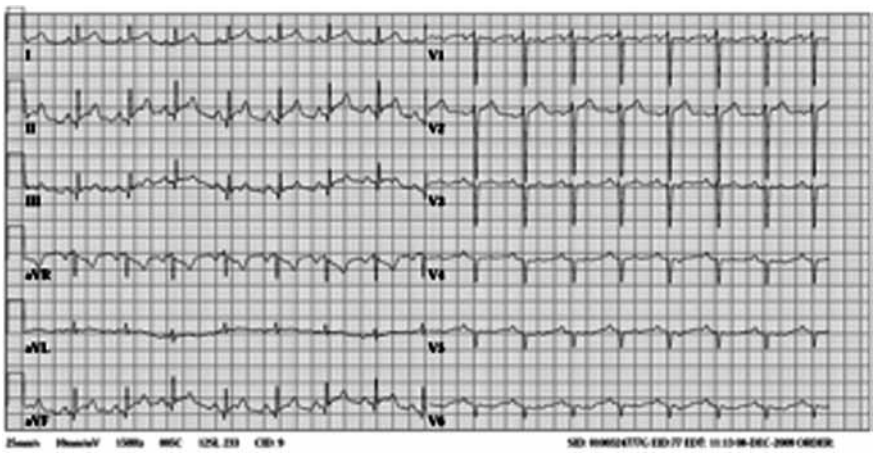

Fig. 1. — ECG on admission shows ST-elevation in I, II and aVF, which is compatible with an inferior infarction.

Case report

A 51-year-old Caucasian women presented at the emergency department with a sudden onset of chest pain radiating to the left jaw and a little shortness of breath. In the past few months she consulted for the same clinical problem with no apparent pathological changes, normal ECG and no coronary stenosis. The chest pain was diagnosed as atypical. She was not known to have diabetes and she didn't smoke. Her familial history did not show any significant cardiac conditions. On presentation at the emergency department, her blood pressure was 100/70 mmHg with a heart rate of 100 bpm. Lung auscultation revealed bibasal rales.

Electrocardiography showed a sinus tachycardia of approximately 100 bpm (PR interval 146 ms, QRS duration 74 ms, QT/QTc 314/400 ms.) and signs of inferolateral ischemia (Fig. 1). The blood analysis showed minor changes in the Troponine T concentration and a slight raise of the CK-MB enzymes. Cardiac ultrasound showed apical hypokinesia. Conventional angiography was preformed, revealing no apparent coronary disease or vasospasm. Left ventriculography showed the same apical hypokinesia as seen on ultrasound.

An assessment of the myocardial viability was done by performing a cardiac MRI. Cine MRI sequences (steady state free precession pulse sequence) revealed circumferential hypokinesis of the apex with normal contraction of the cardiac base (Fig. 2, 3). This image is characteristic for the previous described apical ballooning cardiomyopathy. Perfusion image after intravenous administration of gadolinium showed no apparent delayed enhancement (Fig. 4) which would be suggestive of myocardial infarction.

Our patient improved symptomatically with diuretics and after 4 days no apparent symptoms were noticeable. Control was performed

From: 1. Department of Radiology and Imaging, OLV Ziekenhuis, Aalst, Belgium, 2. Department of Radiology, AMC Hospital, Heerlen, The Netherlands.
Address for correspondence: Dr T. De Beule, M.D., Dept of Radiology, Olv Ziekenhuis, Moorselbaan 164, B-9300 Aalst, Belgium. E-mail: Tom.debeule@gmail.com

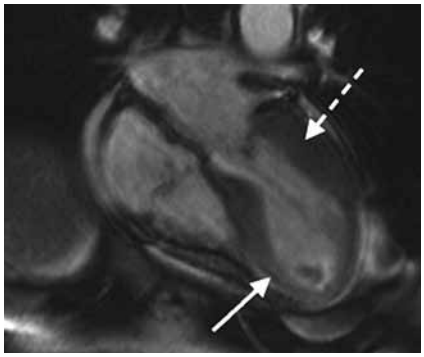

Fig. 2. —Left ventricle in systolic phase of the cardiac cycle which reveals a hypocontractility of the left ventricular apex (arrow) and a normal contraction of the base (striped arrow). Cine MRI sequence with SSFP.

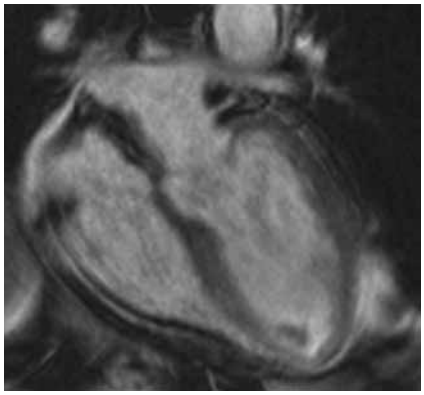

Fig. 3. — A 4-chambre view of the heart (SSFP cine MRI) in the diastolic phase of the cardiac cycle, which shows no diastolic dysfunction.

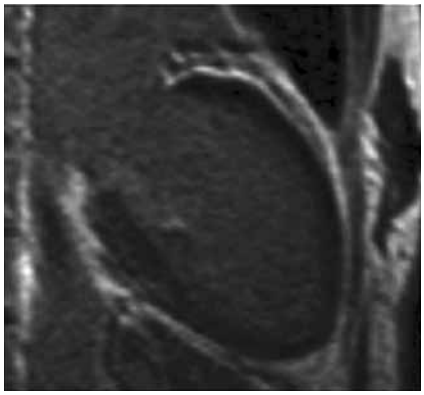

Fig. 4. — Delayed enhancement sequence revealing no enhancement.

which showed improvement of the apical contractility. After one week the patient could be discharged from the hospital.

Discussion

Takotsubo cardiomyopathy is a recently recognized entity which mimics acute coronary syndrome. Transient left ventricular apical ballooning or Takotsubo syndrome has first been reported in the Japanese literature in 1991 (1, 2). This syndrome characterized by sudden onset of chest pain and ecg-changes mimicking an acute myocardial infarction but with a major difference in the regional wall hypokinesia encompassing multiple vascular territories. Most patients present with chest pain or dyspnoea but the clinical presentation can range from these mild symptoms to rare hemodynamic compromise. However the long term prognosis is much better than that of a traditional acute coronary symdrome (6).

Takotsubo is a Japanese word referring to a trap with a round bottom and a narrow neck used for catching octopuses in Japan. The LV morphological features resemble this takotsubo trap (7). The morphology is a typical result of the apical hypokinesia which causes a ballooning of the apex. Other suggested names are 'transient ventricular ballooning syndrome,' 'left ventricular apical ballooning,' 'stress-induced cardiomyopathy,' 'ampulla cardiomyopathy,' and 'broken heart syndrome.' Recently a new atypical form of takotsubo has been described (8). This variant consists of a hypokinesia of the base or middle segment of the myocardium with normal contractility of the apex.

In general, a prevalence of 1,2-2,0% among patients with acute coronary syndrome has been reported (9, 10). Most patients tend to be female and post-menopausal. The age ranges from the first decade until the ninth decade (5). The typical clinical presentation of takotsubo, mimicking an acute myocardial infarction, results in an underestimation of the disease. With a raised awareness for the syndrome and an implementation in the differential diagnosis of an acute coronary syndrome, a higher prevalence is suspected.

In most of the cases patients present with chest pain (70-90%) or with less common symptoms such as dyspnea or pulmonary edema. A striking presentation can be cardiac arrest or cardiac shock, but these remain very rare (5, 11). The electrocardiogram at presentation shows ST-segment elevation in one third of the patients, most commonly in the anterior leads (10). T-wave inversion and non-specific ST-elevation can also be recognized. Pathological Q-waves are present in almost 40% of all patients. In serial electrocardiograms a prolongation of QT interval and a T-wave inversion is commonly seen (12). However, torsade the pointes is rarely reported despite the prolongation in QT-interval (13).

Cardiac biomarkers of myonecrosis are generally slightly elevated at the time of presentation. This is almost always the case for troponin. Elevation of NT-pro BNP is almost always seen, resembling the extend of the disease. When a more extensive part of the myocardium is akinetic, a larger drop in ventricle output can be expected and the atrium will suffer from a heavier afterload. The concentration can be correlated with the outcome of the disease.

Coronary angiography is frequently normal or reveals only mild abnormalities. These patients generally do not have obstructive or instable coronary lesions (6). Almost all patients recover fully and the compromised LV-systolic function improves rapidly over a short period of time.

The underlying cause of takotsubo remains unkown. However the available studies show that the disease results from extreme emotional and/or physical stress combined with a strong predominance in postmenopausal women (14). A few pathophysiological mechanisms have been proposed. Almost all patients presenting with Takotsubo do not have relevant coronary artery obstruction. Therefore, a concept of epimyocardial spasm was proposed. However, in takotsubo there is no regional wall motion abnormality corresponding to vascular territories. In a study, Gianni et al. found that only few patients experienced multivessel spasms even after using provocative tests (15) Concepts like microvascular disturbance and catecholamine-trigged myocyte injury have been proposed (16).

An important imaging modality to differentiate the clinical presentation of takostubo with other aetiologies, such as myocardial apical infarction, or myocarditis is cardiac MRI (17). Delayed hyperenhancement is known to detect myocardial fibrosis and myocardial inflammation and therefore it is typically used for

examining the extent of myocardial infraction or myocarditis. Myocardial fibrosis develops after a myocardial infarction and the delayed enhancement (DE) area is typically distributed corresponding to a vascular territory. Another important differential diagnosis, myocarditis, can be ruled out in this way by using DE. Patchy areas of delayed enhancement can be seen in 88% of the patients with proven myocarditis (17). Delayed enhancement is virtually absent in takotsubo cardiomyopathy. However, the lack of hyper enhancement itself is not sufficient for diagnosing takotsubo, because in a minority of patients with infarction no apparent enhancement is seen.

Second, Cine MRI sequences (steady state free precession pulse sequence) can provide an insight in the typical hypokinesia/akinesia of the myocardium and have the advantage of detecting major complications such as thrombus or aneurysmal dilatation of the ventricular apex.

Conclusion

Takotsubo cardiomyopathy is an important differential diagnosis of an acute coronary syndrome. Characterized by normal or near-normal coronary arteries and regional wall abnormalities extending beyond the vascular territories, this syndrome generally has a favorable prognosis. MRI can help in the differential diagnosis between other etiologies of an acute coronary syndrome. Myocarditis and acute myocardial infarction can be ruled out by using the delayed enhancement sequence. The cine sequences can confirm the regional wall motion abnormality specific to takotsubo.

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