Atypical Takotsubo Cardiomyopathy

Cardiomyopathie de stress : Tako-Tsubo inversé

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Résumé
La cardiomyopathie de stress ou Takotsubo ou ballonisation apicale est une cardiomyopathie consistant en une sidération myocardique qui touche essentiellement les segments apicaux du ventricule gauche survenant après un stress émotionnel. Nous rapportons le cas d’une patiente ayant présenté une cardiomyopathie de stress atypique où l’altération touchait essentiellement les segments médio-ventriculaires. Différentes explorations (échocardiographie, imagerie par résonance magnétique et coronarographie) ainsi que l’évolution ont permis de retenir le diagnostic.

Summary
Stress-induced cardiomyopathy or Takotsubo cardiomyopathy is characterized by a transient left ventricular dysfunction, usually followed by a complete resolution. It is precipitated by severe stress. While there is apical hypokinesis and ballooning with basal hyperkinesis in the majority of cases, a small proportion of patients present with basal or mid-ventricular dysfunction also called ‘atypical or reverse or inverted’ Takotsubo cardiomyopathy.
We report the case of a stress-induced cardiomyopathy with a mid-ventricular takotsubo profile. We highlight the importance of several exams (trans-thoracic echocardiography, cardiac catheterization and cardiac magnetic resonance) for the diagnosis.

Mots-clés
Takotsubo; Cardiomyopathie de stress; Echocardiographie; Dysfonction médio-ventriculaire

Keywords
Takotsubo, stress-induced cardiomyopathy, mid-ventricular dysfunction

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INTRODUCTION

The stress-induced cardiomyopathy, or Takotsubo cardiomyopathy (TTC), is characterized by a reversible ventricular dysfunction and mimics acute coronary syndrome with similar symptoms ranging from isolated chest discomfort to, rarely, cardiogenic shock, in the absence of coronary stenosis [1]. The mechanism of TTC is still debated, although it seems often triggered by emotional or physical stress. We report the case of a woman admitted for an atypical mid-ventricular TTC.

CASE REPORT

A 68-year-old woman with a prior history of hypertension was admitted to our hospital for sudden chest pain and dyspnea. On the day of presentation, the patient reported an important physical and emotional stress due to a fight with a neighbor, after which, she began to develop neck pain, which progressed to prolonged chest pain associated with mild shortness of breath. On examination, the patient appeared quite well but was very anxious. The heart rate was 70/minute and the blood pressure was 132/85 mm Hg. The first electrocardiogram (ECG) recording in the emergency room showed sinus rhythm with abnormal repolarisation in anterior leads. The blood tests revealed an elevated cardiac troponin I level (6.2ng/mL). The patient was initially treated for a non-ST-segment elevation myocardial infarction (NSTEMI). She received morphine, aspirin, clopidogrel, enoxaparin and nitrates and was taken urgently to the cardiac catheterization unit. But, the coronary angiography was negative for obstructive coronary disease with no significant stenoses, occlusion nor dissection (Figure 1). The echocardiogram revealed a moderate reduced overall left ventricular ejection fraction (LVEF) of 0.40, with basal hypokinesis, midventricular segmental anteroseptal and lateral akinesis and apical hyperkinesis (Figure 2). An analysis of LV global longitudinal peak systolic strain showed normal apical strain with reduced strain in the mid and basal segments of the anterior, lateral and septal segments (Figure 2).

Figure 1: Coronary angiography: The absence of obstructive coronary artery disease

Figure 2: Echocardiographic analysis at admission: (a) Left ventricle (LV) dysfunction (b) LV global longitudinal peak systolic strain (GLPS) shows normal apical strain and reduced strain in the mid and basal segments of the anterior, lateral and septal LV walls.
The cardiac magnetic resonance imaging (MRI) confirmed basal segment, mid-anterior, and anteroseptal segmental hypokinesis with the remaining segments noted to be hyperkinetic. The right ventricular function was normal. A pattern of myocardial edema by T2-weighted imaging in the midventricular segments with no late gadolinium enhancement confirmed the diagnosis of TTC with a midventricular pattern (Figure 3).

The patient was eventually discharged from the hospital with the diagnosis of stress-induced midventricular cardiomyopathy. She was placed on a regimen of aspirin, low dose β-blocker, and an angiotensin-converting enzyme (ACE) inhibitor. At a follow-up visit four weeks later, the patient remained symptom-free and repeat echocardiogram showed complete resolution of the wall-motion abnormalities, together with an estimated LV ejection fraction of 0.55, an ameliorated global longitudinal LV strain, and the restitution of the midventricular segmental strain (Figure 4).

**Figure 3:** Cardiac MRI: A pattern of myocardial edema by T2-weighted imaging in the midventricular segments with no late gadolinium enhancement confirmed the diagnosis of Takotsubo cardiomyopathy with an inverted pattern

**Figure 4:** Echocardiographic analysis during the follow-up (1-month later): (a) complete resolution of the wall-motion abnormalities and estimated left ventricle (LV) ejection fraction of 55% (b) LV global longitudinal peak systolic strain (GLPS) shows the restitution of the mid ventricular segmental strain.
DISCUSSION

We report a rare case of atypical TTC involving only the basal and mid-ventricular segments and occurring in a 68-year-old woman after an important physical and emotional stress. The TTC, also known as stress-induced cardiomyopathy and transient left ventricular apical ballooning cardiomyopathy or “broken heart syndrome”, was first described in the Japanese literature in 1990 as a syndrome of reversible LV dysfunction with apical ballooning and the appearance of a Japanese octopus trap or Tako-tsubo in Japanese [2].

The TTC is a rare acute cardiac syndrome occurring predominantly in post-menopausal women and often associated with sudden important emotional or physical stress [3, 4]. It is characterized by the rapid development of severe transient LV dysfunction involving the apical segments in the absence of angiographic coronary stenosis [5]. Since the first description of this syndrome, different atypical patterns of left ventricles involvement have been described (reverse type, midventricular type, and focal type) which represent about 20% of all cases [6].

The atypical TTC has different characteristics than typical TTC, including younger age of onset, more frequent ST-segment depression, higher prevalence of neurologic diseases and less pronounced reduction in left ventricular ejection fraction [6]. The troponin levels are higher compared to other patterns, which is the consequence of the larger muscle region involved in reverse TTC compared to apex alone, but, on the other hand, the brain natriuretic peptide values are more elevated in apical and midventricular patterns, which is clinically translated by more severe symptoms and higher NYHA functional class [1, 6].

The pathophysiological basis of the myocardial dysfunction is multifactorial. The adrenergic storm is one of the most recognized mechanisms of TTC, where excessive circulating epinephrine induces multiple coronary spasm, microvascular dysfunction, negative inotropic effect due to anomaly in the intracellular calcium metabolism, and myocardial damage [7].

The reason of the distribution of myocardial stunning is not yet well understood. The distribution of adrenergic receptors seems to play an important role and it was hypothesized that areas with a higher density of adrenergic receptors may determine the area of hypokinesis. This phenomenon explains that typical TTC occurs more often in older patients where adrenoreceptors density in the apex is reduced because of hormonal change, while inverted variant occurs in younger patients [1].

The present case is original because of the atypical location of wall motion abnormalities occurred in a post-menopausal woman. It illustrates the potential usefulness of several exams such as trans-thoracic echocardiography (TTE), cardiac MRI and coronary angiography for the diagnosis. The Cardiac MRI can be helpful as a second-line imaging modality in patients with suspected TTC, it might be useful to confirm the presence of viable myocardium in the akinetic regions [8]. It can also detect LV dysfunction in a non-coronary regional distribution pattern, myocardial edema in the segments with wall-motion abnormalities, and the absence of high-signal areas. Several MRI findings are useful for differential diagnosis, including intense delayed subendocardial or transmural hyperenhancement in acute myocardial infarction, patchy hyperenhancement in myocarditis, and edema in TTC [8]. The prognosis of TTC is good [9]. The short-term treatment of TTC involves supportive care. Further treatment usually involves standard heart-failure therapy, such as ACE inhibitors and β-blockers [9]. After recovery, ACE inhibitors can be discontinued when the LV returns to normal [9].

CONCLUSION

We report a case of an atypical TTC after an emotional stress in a post-menopausal woman. The basal and mid-ventricular LV involvement may be the first presentation of TTC. Echocardiography and cardiac MRI should be performed in patients with acute chest pain, biological cardiomyocytes necrosis and normal coronary angiogram to track stress-induced cardiomyopathy.

REFERENCES