"Grandma's" and substernal chest pain. An educational in Tako-tsubo cardiomyopathy.

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Learning objectives

To illustrate the typical imaging findings in Tako-tsubo cardiomyopathy (TC). This educational will discuss three different cases of TC. We report one case showing typical reversible midventricular and apical akinesia, a second case with myocardial edema in the acute setting and lastly we report a case with left ventricular (LV) thrombus. Additionally, in two of the cases late gadolinium enhancement (LE) was found.
Background

TC is known under a variety of different names including 'transient left ventricular apical ballooning syndrome', 'stress induced cardiomyopathy', or more informal as 'the broken heart syndrome'. It was first recognized in Japan in 1990. Subsequently, it was increasingly reported upon around the world. The clinical presentation is similar to acute coronary syndrome (ACS) and includes substernal chest pain and/or dyspnea. Most often the syndrome is accompanied by electrocardiogram (ECG) abnormalities and positive cardiac biomarkers. TC is typically identified by imaging showing transient LV dysfunction in a non-coronary distribution pattern. These abnormalities are often triggered by acute emotional or physical stress. Furthermore, no significant coronary artery stenosis is found in these patients. TC predominantly affects postmenopausal women. The precise incidence is unknown but it was estimated that 2% of patients presenting with suspected ACS have TC. The exact cause remains unresolved but it is hypothesized that an excessive catecholamine release may play a significant role in the transient myocardial dysfunction. The treatment of TC is usually supportive and includes standard care for congestive heart failure. Despite the favorable prognosis there are complications associated with TC, including cardiogenic shock, ventricular arrhythmias, mitral valve dysfunction, LV rupture and mural thrombus.
Findings and procedure details

TC is identified by transient LV dysfunction in a non-coronary distribution pattern with typical 'apical ballooning'. The latter refers to mid and distal akinesis of the LV with normal or hyperdynamic basal contraction. This akinesis causes the shape of the LV to change, which thereby resembles the shape of a Japanese octopus trap (fig 1). Therefore the syndrome is called "Tako-tsubo" cardiomyopathy.

Until recently the absence of LE was thought to be typical in TC. However, several studies challenged this by reporting transient LE in TC patients. The signal intensity of LE is much lower than that seen in myocardial infarction or myocarditis. It is thought that LE in TC is associated with increased LV wall stress and mass. Furthermore LE may even be a prognostic determinant of complications such as heart failure and severe arrhythmias.

Case 1:

A 63 year old woman presented to the emergency room with severe substernal chest pain, which started after resuscitation of her husband. Both blood tests and ECG showed abnormalities which could indicate ischemic heart disease. Because an ACS was suspected, subsequently a coronary angiography (CAG) was performed showing no significant coronary artery stenosis. In addition, a LV angiogram was made, which showed ballooning of the mid segments of the LV and normal/hyperdynamic basal and apical segments (fig 2). A cardiac magnetic resonance (MR) study performed the next day confirmed normal basal and apical contraction of the LV and akinesis of the mid segments predominantly in the anteroseptal segments (fig 3). Furthermore, there was mild dilatation of the LV (fig 4). No LE was present. In this case resuscitation of her husband was the emotional trigger that probably caused the TC. No complications were observed and follow-up cardiac MR several months later showed normalized LV function.

Case 2:

A 62 year old woman presented herself to a cardiologist in a local hospital in the vicinity. She suffered from substernal chest pain and blurred vision. At presentation she suffered from severe socially related emotional stress. Her ECG showed signs of ischemia of the inferoposterolateral LV wall and lab tests showed mildly elevated cardiac enzymes. Under the suspicion of ACS she was transferred to our hospital for CAG. The CAG showed normal coronary arteries.

The next day a cardiac MR showed a dilated LV with thinning of the LV wall and typical "apical ballooning" (fig 5). T2 weighted images showed diffuse hyperintense signal in the myocardium from midwall down to the apex (fig 6). Mild midwall LE was seen apical most
significantly in the septal segments (fig 7). In this case the probable emotional trigger for TC was her ongoing relational stress. No complications were observed. Follow-up cardiac MR three months later showed a normalized LV function and absence of LE.

**Case 3:**

A 74 year old woman, at presentation post-ictal which rendered communication impossible.

She suffered from epilepsy due to multiple meningeomas of which one was removed surgically several years ago. Her ECG and lab tests showed mild signs of ACS and subsequently a CAG was performed. The CAG showed no significant coronary artery stenosis. Additionally a LV angiography was made, which showed the typical "apical ballooning" (fig 8). Because of her neurological history a computed tomography (CT) of the brain was obtained which revealed the pre-existing meningeomas, post-surgical changes in the frontal area and no signs of intracranial bleeding or ischemia. A cardiac MR and a MR of the brain were made two days later. The MR of the heart confirmed normal basal contraction of the LV and akinesia of the mid and distal segments (fig 9). Mild LE of the myocardium was seen in a mid-wall distribution, in the septal and frontal wall of the mid ventricle down to the apex (fig 10). Furthermore, an apical thrombus was seen in the LV (fig 9 and 10). The brain MR showed several small areas with restricted diffusion, representing recent lacunar infarctions as thrombo-embolic complications of the LV thrombus (fig 11 and 12). In this case a seizure was the probable physiological trigger that caused TC. This has previously been reported in a small number of cases. Follow-up cardiac MR two months later showed a normalized LV function and absence of apical thrombus or LE.
Fig. 1: An example of a "tako tsubo": Japanese octopus trap.

Fig. 2: LV angiography showing normal/hyperdynamic basal and apical segments en hypokinetic mid segments.

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**Fig. 3:** 2 chamber long axis cine gradient echo MR movie, showing normal basal and apical contraction of the LV and akinesis of the mid segments.

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Fig. 4: Midventricular short axis cine gradient echo MR image, in late diastole, showing LV dilatation and anteroseptal thinning of the LV wall.

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Fig. 5: 2 chamber long axis cine gradient echo MR movie, showing typical "apical balloonning".

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Fig. 6: Midventricular short axis T2 weighted MR image, showing myocardial edema infero-septal.

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**Fig. 7:** 4 chamber long axis MR image after administration of gadolinium, showing midwall LE apico-septal.

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Fig. 8: LV angiography showing normal/hyperdynamic basal segments and hypokinetic mid and apical segments.

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Fig. 9: 2 chamber long axis cine gradient echo MR movie, showing normal basal LV contractions and akinesis of mid and distal segments. Also note LV thrombus.

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**Fig. 10:** 2 chamber long axis MR image after administration of gadolinium, showing LE antero-apical. Note LV thrombus.

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Fig. 11: MR of the brain, showing a small left parietal lacunar infarct. A diffusion weighted (b 1000) MR image, showing a hyperintense lesion in the left parietal lobe.

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Fig. 12: MR of the brain, showing a small left parietal lacunar infarct. The apparent diffusion coefficient (ADC) map MR image showing hypointensity in the same area confirming restricted diffusion.

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Conclusion

Consider TC in postmenopausal women who present with symptoms of ACS. MR imaging typically shows transient LV dysfunction in a non-coronary distribution pattern. Despite the favorable prognosis serious complications associated with LV dysfunction such as LV thrombus may occur. Low blood flow in the apex of the LV resulting from the apical ballooning is thought to be the main cause of thrombus formation. The clinical relevance of LV thrombus consists mainly of the potential threat of thrombo-embolic events. LV thrombus can be present at initial presentation or occur at a later time during disease. For this reason all patients with TC should be evaluated for the presence of a LV thrombus. Moreover, in TC transient myocardial LE may occur.
References


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