Takotsubo Cardiomyopathy: The Broken Heart Syndrome

Chris Gray, MD, FRCPC and Richard Townley, MD, FRCPC

Charlotte's chest pain

Charlotte, 45, presents to the ED with an hour of substernal chest pain. Her electrocardiogram reveals diffuse ST-T abnormalities. Her chest pain is not relieved with sublingual nitroglycerine; in fact, she has transient hypotension after the administration of this medication. Her pain and ECG abnormalities persist despite being given acetylsalicylic acid (ASA), supplemental oxygen and morphine.

Her only risk factor for coronary disease is a family history of premature coronary disease. Her past medical history is unremarkable. She has recently been told that her mother, who was just placed in a nursing home, has metastatic colon cancer. Charlotte was quite upset by this news and initially thought that her chest pain was stress related.

Charlotte is taken to the cardiac catheterization laboratory. Her coronary angiogram demonstrates normal coronaries. Her left ventriculogram (Figure 1 and Figure 2) shows basal hyperkinesis with apical dyskinesis or apical ballooning. Charlotte becomes profoundly hypotensive with intravenous nitroglycerine, but eventually stabilizes with medical therapy.

What’s your CardioCase diagnosis?

See page 22 for Charlotte’s follow-up.
CardioCase discussion

What is the Diagnosis?

Stress induced cardiomyopathy is characterized by an acute onset of reversible left ventricular (LV) apical ballooning (during systole), with chest pain, electrocardiographic changes and no, or minimal, elevation of cardiac enzymes in the absence of significant coronary artery disease. This disorder is also referred to as:

- Broken heart syndrome,
- Transient LV apical ballooning and
takotsubo cardiomyopathy.

Takotsubo cardiomyopathy

Takotsubo cardiomyopathy was initially described in Japan, but has subsequently been observed in Europe, the US and Canada. This condition tends to affect postmenopausal women, but may affect premenopausal women and men. Patients usually present with features mimicking acute myocardial infarction (MI), including chest pain, dyspnea, hypotension or isolated ECG abnormalities. Coronary angiography, by definition, does not show any critical lesions.

Pathophysiology

The exact mechanism of this disorder is not well understood. This condition is usually precipitated by significant intense emotional or physical stress, such as the death of a relative, domestic abuse, severe medical illness, natural disasters and catastrophic medical diagnoses.

Catecholamines may play an important role in the genesis of this disorder, as illustrated by a small case series showing significantly increased plasma epinephrin and norepinephrine in these patients. These patients also tend to be prone to coronary vasospasm, possibly induced by excess catecholamines.

More on Charlotte

While in the hospital Charlotte had transient hypotension treated with inotropes which worsened her condition. The diagnosis of left ventricular (LV) outflow tract obstruction was made clinically and was confirmed by an echocardiograph. She stabilized quickly with appropriate medical therapy. By the time she was discharged (sixth day), her LV function was much improved.

At her one month follow-up appointment, her LV function was normal as confirmed by an echocardiograph.

In summary, it is most likely a non-ischemic, metabolic syndrome caused by stress-induced activation of cardiac adrenoreceptors in the absence of ischemia and reperfusion.

Presentation

Most patients present with signs and symptoms similar to an acute MI. ECG and cardiac biomarkers are also usually abnormal. The most common ECG abnormality is ST segment elevation. Also seen are ST depression, T wave abnormalities, a prolonged QT interval and rarely, pathologic Q waves. Cardiac biomarkers (troponin and creatine kinase) are often only mildly elevated.

Coronary angiography, by definition, reveals no hemodynamically significant atherosclerotic lesions. Left ventriculography or echocardiography reveal typical apical ballooning of the distal half to two thirds of the LV walls.

Occasionally LV outflow tract (OT) obstruction is seen and is felt to be secondary to LV basal hyperkinesis.

About the authors...

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Diagnosis

The diagnosis of stress induced cardiomyopathy is usually made after initiation of treatment for an acute coronary syndrome, either at the time of LV function assessment or at the coronary angiography. Certain authors have also proposed the absence of other diagnoses known to cause ECG abnormalities and LV dysfunction as a diagnostic criteria for Takotsubo (e.g., hypertrophic cardiomyopathy, intracranial hemorrhage/significant recent head trauma, pheochromocytoma, myocarditis, etc.).1,2,5

Treatment

Stress induced cardiomyopathy is, by definition, a self-limiting disorder, treated with supportive therapy. Care must be taken in the subset of these patients with LVOT obstruction so as not to worsen the gradient with inotropes, volume depletion or overaggressive vasodilation. Specific treatments for patients with LVOT obstruction are similar to patients with hypertrophic cardiomyopathy with obstruction and include beta-blockers, fluid resuscitation, and pure alpha agonists.1,3,6

Prognosis

The prognosis is excellent for patients with this disorder. Hospital mortality rates range from 0% to 8%. LV systolic function typically returns to normal within one to four weeks. Stress induced cardiomyopathy can recur, and the role of long term therapy is unclear.1,3,4,5

Characteristics of stress induced cardiomyopathy

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<th>Patients affected</th>
<th>Usually post-menopausal women</th>
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<td>ECG changes</td>
<td>ST elevation of diffuse ST-T abnormalities</td>
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<td>Presenting features</td>
<td>Chest pain, dyspnea, hypotension</td>
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<td>Cardiac enzymes</td>
<td>No rise or minimal abnormality</td>
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<td>Coronary angiography</td>
<td>No significant coronary artery disease</td>
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<td>LV wall motion abnormality</td>
<td>Apical dyskinesis or apical ballooning during systole</td>
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<td>Precipitating factors</td>
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LV: left ventricle

References