Apical Ballooning Syndrome
An Important Differential Diagnosis of Acute Myocardial Infarction
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Case presentation: A 60-year-old woman presented to the emergency department 2 hours after the onset of severe retrosternal chest pain that started soon after she was told that her son had died in a car accident. A 12-lead ECG demonstrated ST-elevation in the precordial leads (Figure 1), and the plasma troponin T level was elevated at 0.07 ng/mL. A diagnosis of acute ST-elevation myocardial infarction was made, and the patient was admitted for emergency coronary angiography, which revealed normal coronary arteries. The left ventriculogram showed severe systolic dysfunction involving the mid and apical segments (Data Supplement Movie I).

Recognition of Clinical Syndrome
Physicians have long been aware of the possible association between stress and cardiovascular events. Awareness has increased of a distinct cardiac syndrome that was originally described in the Japanese population and was called Takotsubo cardiomyopathy, named after the octopus-trapping pot with a round bottom and narrow neck that resembles the left ventriculogram during systole in these patients. Other names used to describe the condition include apical ballooning syndrome (ABS), broken heart syndrome, and stress or ampulla cardiomyopathy. The precise incidence of ABS is unknown, but it may account for 1% to 2% of patients who present with an acute myocardial infarction.3

Clinical Features
The majority of patients have a clinical presentation that is indistinguishable from an acute coronary syndrome. Most present with chest pain at rest, although some patients have dyspnea alone as their initial presenting symptom. Rarely, patients present with syncope or an out-of-hospital cardiac arrest.4 ABS appears to occur almost exclusively in postmenopausal women; however, a few cases have been reported in younger women and males.4 The patients are usually hemodynamically stable, but clinical findings of mild-to-moderate congestive heart failure commonly coexist. In a minority of patients, hypotension may occur from reduced stroke volume and occasionally from dynamic left ventricular outflow tract obstruction. Cardiogenic shock may be a rare complication. A unique feature of ABS is the occurrence of a preceding emotionally or physically stressful event in approximately two thirds of patients. Importantly, such a trigger is not observed in all individuals, and its absence does not exclude the diagnosis.

Diagnosis
The most frequent finding on the admission ECG is mild ST-segment elevation, which occurs in approximately 50% to 60% of patients. It is typically present in the precordial leads, but the ECG can be normal or can show nonspecific T-wave abnormality or major ST elevation across the precordial and limb leads. In general, the 12-lead ECG alone is not helpful in differentiating ABS from an ST-elevation myocardial infarction.5 Characteristic evolutionary changes that occur over 2 to 3 days include resolution of the ST-segment elevation and subsequent development of diffuse and often deep T-wave inversion that involves most leads (Figure 2). New pathological Q waves may be seen occasionally, and there is frequent prolongation of the corrected QT interval. Most, if not all,
patients have a small early increase in cardiac biomarkers, especially when cardiac troponins are measured. Trans-thoracic echocardiography can detect the regional wall-motion abnormality, but the diagnosis is frequently made in the cardiac catheterization laboratory because the patients are initially suspected of suffering from an acute coronary syndrome and are referred for urgent or emergency coronary angiography. Patients with ABS typically do not have obstructive coronary artery disease. The left ventriculogram shows characteristic regional wall-motion abnormalities involving the mid and usually the apical segments. There is sparing of the basal systolic function, and the wall-motion abnormality extends beyond the distribution of any 1 single coronary artery (Data Supplement Movie I). Apical sparing variants have also been described (Data Supplement Movie II).6 Cardiac magnetic resonance imaging may be helpful in excluding myocardial infarction, because delayed gadolinium hyperenhancement is not a feature of ABS.7

On the basis of our experience, we have previously proposed criteria for making a clinical diagnosis of ABS, and all 4 criteria must be present. The Table illustrates a modified version of the criteria that incorporates contemporary knowledge about the syndrome.4 The diagnosis of ABS is most likely to be made at institutions where primary percutaneous coronary intervention and an early invasive strategy are practiced for the management of acute coronary syndromes. The absence of obstructive coronary artery disease and the characteristic regional wall-motion abnormality are likely to lead to the diagnosis. Diagnosis of ABS in patients who present at hospitals without cardiac catheterization laboratories requires a high index of suspicion. The diagnosis should be considered in a postmenopausal woman who presents with cardiac symptoms that are temporally related to an emotional or physical stressor, with positive cardiac biomarkers or an abnormal ECG. Establishing the diagnosis is particularly important if fibrinolytic therapy is being considered for a presumed diagnosis of ST-

Figure 1. Twelve-lead ECG demonstrating ST-segment elevation in precordial leads.

Figure 2. Twelve-lead ECG demonstrating resolution of ST-segment elevation and development of diffuse T-wave inversion.

Proposed Mayo Clinic Criteria for the Clinical Diagnosis of ABS

1. Transient hypokinesis, akinesis, or dyskinesis of the left ventricular mid segments with or without apical involvement. The regional wall-motion abnormalities extend beyond a single epicardial vascular distribution.*

2. Absence of obstructive coronary disease or angiographic evidence of acute plaque rupture.†

3. New ECG abnormalities (either ST-segment elevation and/or T-wave inversion) or elevated cardiac troponin.

4. Absence of:
   - recent significant head trauma
   - intracranial bleeding
   - pheochromocytoma
   - myocarditis
   - hypertrophic cardiomyopathy

*There are rare exceptions to these criteria, such as those patients in whom the regional wall-motion abnormality is limited to a single coronary territory.

†It is possible that a patient with obstructive coronary atherosclerosis may also develop ABS. However, this is very rare in our experience and in the published literature, perhaps because such cases are misdiagnosed as an acute coronary syndrome.

In both of the above circumstances, the diagnosis of ABS should be made with caution, and a clear stressful precipitating trigger must be sought.
appropriate myocardial infarction. Inappropriate administration of fibrinolytics to a patient with ABS may lead to harm, and it would be appropriate to transfer a patient suspected of having ABS for emergency coronary angiography.

Management

The optimal management of ABS has not been established, but supportive therapy invariably leads to spontaneous recovery. Because the presentation in these patients is indistinguishable from an acute coronary syndrome, initial management should be directed toward the treatment of myocardial ischemia, with continuous ECG monitoring and administration of aspirin, intravenous heparin, and β-blockers. Once the diagnosis of ABS has been made, aspirin can be discontinued unless there is coexisting coronary atherosclerosis. β-Blocker therapy may be continued, if tolerated, especially because an excess of catecholamines may play a role in precipitating the conditions. Diuretics are effective for the treatment of congestive heart failure. The right ventricle is also involved in approximately one third of cases, and these patients are sicker and more likely to develop congestive heart failure (Data Supplement Movie III). On rare occasions, when there is significant hypotension, it is important to exclude dynamic left ventricular outflow tract obstruction with echocardiography. If present, a cautious trial of β-blockers may help by reducing the hypercontractility of the base of the left ventricle. Alternatively, an infusion of phenylephrine may be effective by increasing the afterload and left ventricular cavity size. Inotropes would be contraindicated in this situation. In contrast, cardiogenic shock owing to pump failure is treated with standard therapies that include inotropes and intra-aortic balloon counterpulsation. Anticoagulation should be considered in cases of severe left ventricular systolic dysfunction to prevent thromboembolism until there has been recovery.

Prognosis

Patients with ABS generally have a good prognosis in the absence of significant underlying comorbid conditions. The systolic dysfunction and the regional wall-motion abnormalities are transient and resolve completely within a matter of days to a few weeks. This is such a uniform finding that an alternative diagnosis should be considered in patients in whom the cardiomyopathy does not resolve. Typically, assessment of the ejection fraction should be performed at approximately 4 to 6 weeks after discharge from the hospital to document recovery of function. At this time, the ECG usually shows complete resolution, although T-wave inversion may persist for a longer period of time. In-hospital mortality from ABS is very low and unlikely to be greater than 1% to 2%. The recurrence rate of ABS is no more than 10%. In our practice, in the absence of contraindications, we recommend chronic β-blocker therapy, with the aim of reducing the likelihood of a recurrent episode. Annual clinical follow-up is advisable because the natural history of ABS remains unknown.

Pathophysiology

The pathophysiology of ABS is poorly understood. Several mechanisms for the reversible cardiomyopathy have been proposed, including multivessel epicardial spasm, coronary microvascular spasm, catecholamine-induced myocardial stunning, and myocarditis. Multivessel epicardial coronary artery spasm has not been observed in any of the patients at our institution, but it is possible that the routine administration of nitrates for ischemic chest pain may obscure its presence. Microvascular dysfunction is present in at least two thirds of patients at the time of presentation, and its severity correlates with the magnitude of troponin elevation and ECG abnormalities. It remains to be established whether impairment in microvascular function is the primary mechanism for the injury or is an epiphenomenon. Catecholamines may play a role in triggering the syndrome.

Case Conclusion

The left ventricular end-diastolic pressure was 30 mm Hg, and the patient was markedly hypotensive. An intra-aortic balloon was inserted. Her symptoms and hypotension resolved over 48 hours. She was discharged on the fourth day with a prescription for an ACE inhibitor and β-blocker. An echocardiogram 4 weeks later revealed normal left ventricular function. The ACE inhibitor was discontinued, and she remains well 1 year after her admission.

Disclosures

None.

References

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