

MORNING REPORT

Status Update: Heart Broken

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A 62-year-old female is transferred to our hospital from an outside facility for management of a subarachnoid hemorrhage (SAH) diagnosed by computed tomography (CT) scan of the head. She is afebrile, hemodynamically stable with intact consciousness, and has essentially normal blood tests on presentation. Her past medical history includes depression. She is a physically active home-maker, denies ever smoking, and does not report a family history of cardiovascular disease.

Non-traumatic/spontaneous hemorrhages are frequently seen, usually occurring in the setting of a ruptured cerebral aneurysm or arteriovenous malformation (AVM). About 80% of cases of SAH result from ruptured berry (circle of Willis) saccular aneurysms. Subarachnoid hemorrhage may be seen commonly in head trauma. The history and neurologic examination are essential in the diagnosis and clinical staging (such as the Hunt and Hess staging system) of SAH. The diagnosis is confirmed radiologically by computed tomography (CT) scan without contrast. In the case of a high clinical suspicion of SAH, a negative CT scan is followed with lumbar puncture. Non-contrast CT followed by CT angiography (CTA) of the brain can rule out SAH with greater than 99% sensitivity.¹

These patients are usually managed in the intensive care unit, with careful attention to blood pressure management. A major focus is to prevent cerebral vasospasm-related complications. Surgical treatment to prevent re-bleeding consists of clipping or endovascular treatment by

coiling of the offending berry aneurysm.

On physical examination, her vital signs include a temperature of 38° C, heart rate 90 beats/minute, blood pressure of 150/90 mm Hg, and oxygen saturation 99% on room air. Neurological examination reveals meningeal signs with no focal neurological deficits and intact mentation. The examination of all other organ systems is normal.

She undergoes emergency cerebral angiography and coiling of a ruptured anterior communicating artery (ACOM) aneurysm followed the next day by elective coiling of an un-ruptured second ACOM aneurysm. Post procedure, the patient is placed on vasopressor support to circumvent cerebral vasospasm with norepinephrine, dobutamine, and vasopressin. An echocardiogram is done showing normal contractility and left ventricular ejection fraction.

Ten days later, the patient is severely hypotensive and unresponsive to vasopressor support. Cardiac enzymes are elevated with no electrocardiographic changes noted. The patient's neurological status worsens, and she requires intubation with mechanical ventilation. An echocardiogram shows severely impaired left ventricular function with an ejection fraction of 10%, apical akinesis with ballooning, and basal wall sparing.

Knowledge of the cerebral auto-regulatory mechanism is important post SAH and is vital in its management. In the early 1900s, Cushing described the cardiac effects of subarachnoid hemorrhage, including

elevated blood pressure and bradycardia.² Later with the advent of the EKG, Byer et al. described the ST segment and T wave changes in the EKG.³ Autopsy studies have shown varying degrees of myocardial necrosis and subendocardial hemorrhage. Cardiac dysfunction is a known complication of subarachnoid hemorrhage, and several variants of this dysfunction have been described in the literature ranging from descriptions such as Takotsubo cardiomyopathy (broken heart syndrome) to neurogenic stunned myocardium.⁴ The differences between the two entities relate to the variations noted in echocardiographic findings. In the neurogenic stunned myocardium, the cardiac apex is usually spared, and there is basal hypokinesis; however, in Takotsubo cardiomyopathy (TCM), there is apical akinesis and basal sparing.⁵

Takotsubo cardiomyopathy is a transient cardiac dysfunction that involves left ventricular apical dysfunction resembling acute coronary syndrome. TCM was first described in Japan in 1990. Patients often present with chest pain and may have ST-segment elevation on electrocardiogram and elevated cardiac enzyme levels consistent with a myocardial infarction. However, when the patient undergoes cardiac angiography, left ventricular apical ballooning is present, and there is no significant coronary artery obstruction. On this association, the Mayo criteria were developed for the diagnosis of Takotsubo cardiomyopathy based on available literature.⁶ They include:

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- *Transient hypokinesia, dyskinesia, or akinesia of the left ventricular mid-segments, with or without apical involvement (The regional wall motion abnormalities extend beyond a single epicardial vascular distribution, and a stressful trigger is often, but not always, present.);*
- *Absence of obstructive coronary disease or angiographic evidence of acute plaque rupture;*
- *New electrocardiographic abnormalities (either ST-segment elevation and/or T-wave inversion) or modest elevation in cardiac troponin levels; and*
- *Absence of pheochromocytoma or myocarditis.*

After hemodynamic stabilization, the patient undergoes coronary angiography demonstrating the absence of significant coronary artery disease. After clinical improvement in hemodynamics, the patient is placed on an angiotensin-converting enzyme (ACE) inhibitor and beta-blocker. Four weeks later, repeat echocardiography demonstrates improvement in left ventricular ejection fraction, representing a spontaneous improvement in her cardiac contractility.

The prognosis for Takotsubo cardiomyopathy is usually favorable, if appropriate supportive care is provided during the acute phase. It is widely accepted that neurohormonal and catecholamine-induced myocardial toxicity is most likely to be the basis of both Takotsubo cardiomyopathy and neurogenic stunned myocardium. Other possible etiologies include multi-vessel coronary vaso-

spasm, impaired cardiac microvascular function, impaired myocardial fatty acid metabolism, and even acute coronary syndrome with spontaneous reperfusion with associated reperfusion injury. The apical portions of the left ventricle have the highest concentration of sympathetic innervation found in the heart and may explain why excess catecholamines seem to selectively affect its function.

A significant emotional or physical stressor typically precedes the development of TCM. Nearly 90% of reported cases involve postmenopausal women. The reversibility of left ventricular dysfunction with time and conservative medical management as described in this case is characteristic of TCM and reflects the norm that these patients do indeed have an excellent prognosis for full recovery. Mortality from TCM is estimated to be less than 3%. Non-mortality-related complications to anticipate include the following:

- *Left heart failure with and without pulmonary edema,*
- *Cardiogenic shock,*
- *Left ventricular outflow obstruction,*
- *Mitral regurgitation,*
- *Ventricular arrhythmias,*
- *Left ventricular mural thrombus formation, and*
- *Left ventricular free-wall rupture.*

No medical therapies have been studied specifically for TCM; however, it is common practice to prescribe ACE inhibitors or angiotensin receptor blockers at least until left

ventricular function is restored. Beta blockers are also indicated and may be useful in the long term. Other standard outpatient post myocardial infarction medications, such as statins, aspirin, and clopidogrel, are of unknown benefit. Until the emergence of large randomized controlled trials on this interesting and relatively novel condition, it is important for clinicians to anticipate TCM in high-risk populations and recognize and aggressively manage it, given its excellent prognosis.

Learning Points

- Takotsubo cardiomyopathy often mimics acute coronary syndrome in patients (usually post-menopausal females) with emotional/physical stressors, which can include medical problems such as intracranial catastrophes.
- TCM often presents with chest pain, ST segment elevation, and elevation in cardiac enzymes in the absence of demonstrable coronary artery disease on cardiac catheterization.
- The characteristic echocardiographic appearance includes left ventricular apical akinesia/hypokinesia (ballooning) with basal sparing.
- Management is supportive, with hemodynamic support in the acute phase and the use of ACE inhibition and beta blockade.
- In most cases, prognosis is excellent with complete reversibility of cardiac dysfunction.

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