Apical Ballooning Syndrome in Polymyositis Following Placement of Pericardial Drainage Catheter

Jackson J. Liang, DO, Kalkidan G. Bishu, MD and Nandan S. Anavekar, MBBCh*

Abstract - Transient left ventricular apical ballooning syndrome (Takotsubo cardiomyopathy) nearly always occurs following psychological or physical stress. We present a case of apical ballooning syndrome following pericardiocentesis for pericardial effusion related to polymyositis. This case illustrates a rare presentation of apical ballooning syndrome and we provide a brief review of the literature on the pathogenesis and management of this uncommon condition.

Keywords — apical ballooning syndrome; pericardiocentesis, polymyositis, stress cardiomyopathy, Takotsubo cardiomyopathy

Cite this article as: Liang JJ, Bishu KG and Anavekar NS. Apical ballooning syndrome in polymyositis following placement of pericardial drainage catheter. JCVD 2014;2(3):174-176.

I. INTRODUCTION

Transient left ventricular (LV) apical ballooning syndrome (ABS), also called Takotsubo cardiomyopathy or “broken heart syndrome” is characterized by reversible systolic function mostly affecting the apical segments of the LV in the absence of significant coronary artery disease. It nearly always occurs following psychological or physical stress. The clinical picture of ABS often mimics that of acute coronary syndrome (ACS) as patients may present similarly with chest pain, dyspnea, electrocardiogram (ECG) abnormalities, and cardiac biomarker elevation.

We present a case of ABS following pericardiocentesis for pericardial effusion related to polymyositis, which illustrates an uncommon presentation of transient LV ABS.

II. CASE DESCRIPTION

A 56 year old postmenopausal woman was diagnosed with polymyositis after presenting with proximal upper and lower extremity weakness and elevated creatinine kinase with positive antinuclear and anti-Jo-1 antibodies. She also exhibited symptoms of Raynaud’s phenomenon with transient blue discoloration of her hands, exacerbated by cold. She was treated with prednisone and mycophenolate mofetil with improvement in her weakness initially. She subsequently presented for a rheumatology outpatient appointment complaining of worsening dyspnea on exertion, wheezing, lower extremity edema, and weight gain. Her heart rate was 100 beats per minute and her blood pressure was 120/82 mm Hg. Jugular venous pulsations were elevated 5-6 cm. Bibasilar crackles were heard on pulmonary auscultation. Heart sounds were decreased across the precordium. No murmurs, gallops, or rubs were detected on auscultation. She had 2+ pitting pre-tibial edema extending to the calves bilaterally.

Transthoracic echocardiogram (TTE) demonstrated normal LV ejection fraction (69%) and a moderate-sized circumferential pericardial effusion with findings concerning for early tamponade physiology on mitral inflow Doppler studies with evidence of reduced inspiratory collapse of a dilated inferior vena cava. She was admitted to the hospital for pericardiocentesis.

Pericardiocentesis was successfully performed with drainage of 275 ml serous fluid. A 65-cm 6-French pigtail catheter was left in the pericardial space for continued drainage of fluid. Overnight she had severe pleuritic chest pain. Severe anxiety, stress, and discomfort kept her awake all night and she was intermittently hypertensive and tachycardic secondary to pain. The pericardial catheter was removed with complete resolution of pain. Cardiac MRI (Fig. 1, a and b; Supplemental Movie 1) was performed primarily to evaluate for pericardial inflammation and incidentally demonstrated new regional wall abnormalities with severe mid and apical hypokinesis of both ventricles and hyperdynamic basal contractile function. Ejection fraction was depressed at 39%, down from 69% on echocardiogram less than 48 hours prior to hospital admission. The pericardium was mildly thickened with circumferential pericardial enhancement consistent with pericarditis, but there...
was no myocardial edema or abnormal myocardial delayed enhancement to suggest myocarditis.

Figure 1. Initial MRI 4-chamber end-diastolic (a) and end-systolic (b) images demonstrate apical ballooning with preserved systolic basal contraction. Follow-up cardiac MRI images at end-diastole (c) and end-systole (d) one week later reveal resolution of LV apical dysfunction.

She underwent coronary angiogram (Fig. 2, a and b) which demonstrated no evidence of atherosclerotic coronary artery disease, suggesting that her abrupt in-hospital drop of left ventricular function was most likely due to LV ABS.

Figure 2. Diagnostic coronary angiogram showing normal coronary arteries. (a) Right anterior oblique view of the left main, left circumflex and left anterior descending coronary arteries. (b) Right anterior oblique view of a small non-dominant right coronary artery

In addition to continuing steroids and mycophenolate mofetil for polymyositis, she was started on lisinopril and metoprolol for the cardiomyopathy. Her dyspnea improved dramatically following the pericardiocentesis. She remained clinically and hemodynamically stable over the next two days and was discharged home.

She returned for follow-up one week after discharge and noted that her exertional dyspnea had improved substantially. Blood pressure was 100/61 mmHg and her pulse was 72 beats per minute and regular. Jugular venous pulsations were no longer elevated. Lung auscultation was clear. She had no murmurs, gallops, or rubs. She did have trace pretibial pitting edema bilaterally. Followup cardiac MRI (Fig. 1, c and d; Supplemental Movie 2) that day demonstrated normalization of her LV function (ejection fraction improved to 65%).

III. DISCUSSION

Instances of transient LV apical ballooning syndrome have been reported to occur with connective tissue diseases including systemic lupus erythematosus and systemic sclerosis. To our knowledge, we describe the first case of ABS in association with pericarditis in the setting of polymyositis and only the second reported case complicating pericardiocentesis. ABS was thought to be provoked by post-procedural pain in our patient and this case demonstrates the importance of adequate control of pain and anxiety following procedures such as pericardiocentesis.

Transient LV ABS, also known as stress-induced cardiomyopathy, Takotsubo Syndrome, or “Broken Heart Syndrome”, is a rare cause of transient LV dysfunction that is often preceded by a psychological or physical stress in two thirds of patients. Commonly reported emotional stressors include the death or severe illness/injury of loved one or pet, public speaking, car accidents, or surprise parties. Physical stresses including pain, surgeries, procedures, and cardiac stress tests may also induce ABS. As seen in our patient, ABS has a predilection for postmenopausal women.

1-2% of patients who are initially diagnosed with acute coronary syndrome are later found to have transient LV ABS. The clinical presentation is quite similar to ACS, with most patients presenting with chest pain and dyspnea. ECG abnormalities and cardiac biomarker elevation are also frequently observed. TTE or cardiac MRI typically reveals transient apical LV ballooning with contractile dysfunction in “classic” transient LV ABS. Mid-ventricular or basal (reverse) variants have been described as well. Coronary angiography shows normal coronary arteries or only minimal atherosclerosis.

The precise pathophysiologic mechanisms of ABS have yet to be fully elucidated. The adrenal-cardiac axis has been increasingly recognized to be an important factor in the development of heart failure and may be contributory to the pathogenesis of ABS. Exaggerated sympathetic stimulation has been postulated to play a role and plasma catecholamine levels are two to threefold higher in patients with ABS than those with acute myocardial infarction. The adrenal glands produce catecholamines, which are released in response to sympathetic nervous system stimulation to increase cardiac function. However, when catecholamine levels remain chronically elevated, receptor desensitization may occur. At a cellular level, beta-adrenergic receptor desensitization occurs by a process which involves proteins including G-protein coupled receptor kinases (GRK) and beta-arrestins, both of which have been associated with cardiac dysfunction.
beta-adrenergic receptors are phosphorylated by GRKs, translocated, and subsequently internalized after binding to beta-arrestins. Betagate-Siryk, et al. demonstrated in a mouse model that beta-arrestin 1 is detrimental to cardiac function and exacerbates heart failure after myocardial infarction. Beta-arrestin 1 prevents the inhibition of catecholamine release from the adrenal glands, enhances the secretion of aldosterone, and may be contributory to the pathogenesis of ABS. Additionally, it inhibits the inotropic effects of cardiac beta-1 adrenergic receptors resulting in decreased cardiac function. Additionally, diffuse coronary microvascular dysfunction is postulated to possibly play a role. As previously proposed by Melchiorre, et al. in their case description of a patient with ABS and Raynaud’s phenomenon, vasospasm of the coronary microvasculature (so called “myocardial Raynaud’s phenomenon”) may result in transient myocardial ischemia and subsequent systolic dysfunction.

The degree of initial LV dysfunction is not predictive of increased mortality. LV function generally resolves within days to weeks after initial presentation, although complications such as acute left heart failure, pulmonary edema, dynamic intraventricular obstruction, cardiogenic shock, ventricular arrhythmias, LV free-wall rupture, LV thrombus formation, and even death may occur. Mainstay of treatment in ABS is supportive care until LV function recovers. Due to its similarity at presentation, patients should initially be treated for suspected ACS with aspirin, beta-blockers, and intravenous heparin. Optimal management for the acute treatment of ABS, once differentiated from myocardial infarction, is not yet clear. Beta blockers in theory should be beneficial as patients are hyperadrenergic. Angiotensin converting enzyme inhibitors and diuretics are reasonable to give in the acute phase, although there is little data proving long-term benefit. Transient LV ABS can also cause LV outflow tract obstruction (LVOT) due to LV basal hyperkinesis. In subjects with evidence of LVOT obstruction, it may be prudent to avoid inotropic agents, angiotensin converting enzyme inhibitors and angiotensin receptor blockers to prevent worsening of LVOT obstruction.

IV. CONCLUSIONS

ABS should be considered in the differential diagnosis of chest pain, especially when symptoms develop following a psychological or physical stressor. Pericardiocentesis may provoke ABS, particularly when it causes excessive pain and anxiety. Previously associated with rheumatic conditions such as lupus and systemic sclerosis, ABS may also be more likely to develop in patients with polymyositis, particularly when Raynaud’s phenomenon coexists.

V. ACKNOWLEDGMENTS

The authors report no conflicts of interest.

VI. SUPPLEMENTAL FILE LEGENDS

Movie 1: Initial cardiac MRI short-axis views demonstrating LV apical akinesia with preserved basal contraction.

Movie 2: Follow-up cardiac MRI short-axis views demonstrating resolution of LV apical akinesia.

References


