Mitral Valve Prolapse and Giant Coronary Artery Aneurysms

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Abstract

The majority of coronary artery aneurysms are atherosclerotic in origin. Atherosclerotic coronary artery disease (CAD) is the predominant cause in adults. Other causes include Kawasaki's disease, Marfan's syndrome, Behçet's disease, and use of stents. Their size and clinical manifestations are variable. Giant coronary aneurysms, measuring ≥8 mm in diameter, are rare entities. Multivessel coronary artery involvement is still rare. Our case was a 32-year-old lady with an incidental finding of multiple giant coronary artery aneurysms in the evaluation for cardiomyopathy. (*Iranian Heart Journal 2015; 16(1): 46-49*)

Keywords:  ■ Mitral valve prolapse; ■ Coronary aneurysm; ■ Cardiomyopathies

Introduction

A 32-year-old lady presented with progressive dyspnea having exacerbated over the previous 2 months. Echocardiography revealed severe prolapse of the anterior mitral valve leaflet and severe regurgitation. Because of moderate left ventricular dysfunction on echocardiography (EF=35-40%), cardiac magnetic resonance imaging (MRI) was recommended to rule out cardiomyopathy. Cardiac MRI using a 1.5-Tesla Avante Siemens device with gadolinium-based contrast agent (Magnevist) revealed myocardial late gadolinium enhancement in the left coronary territory and a cystic mass in the epicardial aspect of the heart. Subsequently, coronary angiography was recommended for the patient (Figures 1-3).

Figure 1. Phase sensitive inversion recovery (PSIR) sequences, revealing late gadolinium enhancement in the left circumflex artery territory
Coronary angiography showed multiple large coronary aneurysms in the left anterior descending artery (LAD) and right coronary artery (RCA), suggesting Kawasaki's syndrome.

Cardiac computed tomography (CT) angiography with a 256-slice dual-source multi-detector CT with retrospective gating and current tube modulation also revealed multiple large coronary aneurysms.
The patient underwent successful coronary artery bypass grafting, aneurysmectomy, and mitral valve replacement.

**Discussion**

Coronary dilation that exceeds the diameter of the normal adjacent segments by 1.5 times is defined as a coronary artery aneurysm. The majority of coronary artery aneurysms are atherosclerotic in origin. Their size and clinical manifestations are, however, variable. Coronary aneurysms have frequently been reported to have varied incidence and etiology. Atherosclerotic CAD is the predominant cause in adults. Other causes include Kawasaki’s disease, Marfan’s syndrome, Behçet’s disease, and use of stents. Giant coronary aneurysms, measuring ≥8 mm in diameter, are rare entities. Multivessel coronary artery involvement is still rare. Some of the complications recorded to occur during the course of the disease include myocardial infarction due to thrombosis and distal embolization. Kawasaki’s disease (mucocutaneous lymph node syndrome) is a pathological process seen mostly in children, but it has been reported in adults as well. The late manifestations of the disease are multiple coronary artery aneurysms. Coronary artery aneurysms develop in only 10-20% of patients, and about 50% of them may regress in 1-2 years. Aneurysms that occur during the early phase of Kawasaki’s disease are known to involve the proximal segments of the major coronary arteries. Treatment should ideally be provided within 10 days of symptom onset to reduce the risk of coronary artery complications. The standard of care for treatment is intravenous immunoglobulin plus Aspirin; nevertheless, adding corticosteroids may provide additional benefits for high-risk patients. Some patients do not respond to intravenous immunoglobulin and require additional therapy. Part II of this continuing medical education article will focus on the complications of Kawasaki’s disease and potential treatment options.

The most common sites of the aneurysms are the proximal LAD and proximal RCA, followed by the left main coronary artery, LCX, and finally the distal RCA. The aneurysms are classified as small (<5mm diameter), medium (5-8mm diameter), and giant (>8mm diameter). All patients with Kawasaki’s disease should undergo echocardiography on diagnosis and 6-8 weeks thereafter. Those with giant aneurysms may
require coronary angiography to identify stenotic lesions. Young adult patients with coronary artery aneurysms who have no history of atherosclerotic disease should be questioned about any childhood illnesses resembling Kawasaki's disease.⁶

References

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