Case Report

SPONTANEOUS CORONARY ARTERY DISSECTION ASSOCIATED WITH APICAL HYPERTROPHIC CARDIOMYOPATHY

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ABSTRACT
Apical hypertrophic cardiomyopathy (HCM) is a relatively uncommon inherited disease. Spontaneous coronary artery dissection (SCAD) is also uncommonly observed, which often occurs in pregnant or post partum women but is rare in men. This report describes a 38 years old man with apical hypertrophic cardiomyopathy who developed SCAD leading to acute inferior myocardial infarction. After emergent appendectomy operation at another hospital, he was immediately transferred to the Cardiology Department of our hospital due to acute myocardial infarction. He emergently underwent coronary angiography which showed a long dissection involving the right coronary. He underwent an emergent CABG with cardiopulmonary bypass. Postoperative recovery was uneventful and he was discharged. According to our knowledge, no case of spontaneous coronary artery dissection associated with apical hypertrophic cardiomyopathy unrelated to postpartum period or oral contraceptive use has been reported so far.

KEY WORDS: Apical Hypertrophic Cardiomyopathy, Coronary Artery Dissection.

INTRODUCTION
Hypertrophic cardiomyopathy (HCM) is a familial cardiac disease caused by mutations in encoding protein components of the cardiac sarcomere. The phenotypic features of HCM may develop at any age from infancy to adulthood, and are characterized by a great heterogeneity in the extent, magnitude, and distribution of left ventricular (LV) hypertrophy. Apical HCM is a relatively rare morphological expression of the disease (0.5% of patients), in which LV wall thickening is confined to the most distal portion of the ventricle, below the papillary muscle level. This form of HCM is more frequently sporadic, but may also be encountered in the context of clinical screening of HCM families with more common patterns of distribution of LV hypertrophy.

Spontaneous coronary artery dissection (SCAD) is a rare cause of myocardial infarction. It is an uncommon disease, which often occurs...
in pregnant or postpartum females but is rare in males. Cases with SCAD are usually encountered by physician as a sudden death or acute coronary syndrome. These cases have no additional risk factor for coronary artery diseases. About 80% of reported cases are female and associated with pregnancy and/or oral contraceptive. The diagnosis of SCAD should be strongly considered in any patient who presents with symptoms suggestive of acute myocardial ischemia. The diagnosis can be established by urgent coronary angiography which is also needed to determine the appropriate therapeutic approach. In addition, medical therapy and revascularization procedures using PTCA or coronary bypass graft surgery may be required. According to our knowledge, no case of spontaneous coronary artery dissection associated with apical hypertrophic cardiomyopathy unrelated to postpartum period or oral contraceptive use has been reported so far.

**CASE REPORT**

A 38 years old male patient without any previous medical history or coronary risk factors (nondiabetic, normotensive, nonsmoking and normal lipids level) was emergently operated on due to acute appendicitis. During operation, ST elevation in DII and DIII derivations suggesting inferior myocardial infarction was noted.

After successful emergent appendectomy operation at another hospital, he was immediately transferred to the Cardiology Department of our hospital. There was no family history of connective tissue disorders or recent trauma. Physical examination revealed a heart rate of 125 bpm and blood pressure of 80/60 mm Hg. On auscultation, the first and second heart sounds were normal but there was apical systolic murmur (2/6). Typical ST segment elevations were present in inferior derivations at electrocardiographic examination (Fig-1). Echocardiography demonstrated marked apical hypertrophy and inferior wall hypokinesis with ejection fraction of approximately 50%. There were signs of moderate mitral regurgitation. Then, patient was transferred to angiography unite for primer percutaneous coronary intervention. Coronary angiography showed a long dissection involving the dominant right coronary artery between its ostium and distal portion. There was no evidence of any atherosclerotic luminal narrowing or obstruction (Figure-2). He has remained unstable with evidence of ongoing ischemia and hemodynamic compromise. He underwent an emergent CABG with cardiopulmonary bypass. Free right internal mammary artery graft (RIMA) for right coronary artery bypass was used. Postoperative recovery was uneventful and he was discharged home in a stable condition. The patient resumed normal daily activities and remained symptom free nine months after surgery. A repeat coronary angiography was performed approximately 9 months later due to chest pain and nonspecific ST changes. It revealed open RIMA graft and no recurrent dissections in the same and other coronary arteries.

**DISCUSSION**

Apical HCM is LV wall thickening is confined to the most distal portion of the ventricle, below the papillary muscle level. This form of HCM is more frequently sporadic. The clinical course of apical HCM appears to be generally benign, without severe symptoms or major cardiac events. Indeed, a particularly low annual cardiovascular mortality rate (0.1%) was
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reported during a mean follow-up of 9 years in a survey of the largest available cohort of patients with apical HCM (a total of 105 patients). In our case it was sporadic. His parents and brothers were normal at electrocardiographic and echocardiographic examinations. Spontaneous coronary artery dissection is an unusual cause of myocardial ischemia with complex pathophysiology. Fewer than 200 cases of SCAD have been reported in the literature, and the majority (80%) has occurred in young women during the peripartum period or in association with oral contraceptive use.

The clinical presentation of this syndrome relates to the extent and rate of dissection as well as the degree of myocardial ischemia. Patients may present with chronic stable angina, acute coronary syndromes, myocardial infarction, cardiogenic shock, sudden cardiac death, or pericardial tamponade. Sudden death may occur in up to 50% of cases, particularly in those with left main coronary artery (LMCA) dissection. Myocardial infarction was present in our case. The left anterior descending coronary artery is the artery most often involved in SCAD (in about 75% of cases whereas LMCA 24% and multivessel dissection occur in 40% of cases). Most patients with LMCA dissection sustain myocardial infarction (vs. only 50% of patients with RCA dissection). Other series RCA dissection was 17% cases. In our case SCAD was present at RCA.

Atherosclerosis is one of the main risk factors for SCAD. Hering and co-authors reported atherosclerotic plaque rupture in 35 of 42 SCAD patients. In this patient, atherosclerotic coronary artery stenosis was not seen on coronary angiography. The treatment options for SCAD include medical therapy and revascularization procedures using percutaneous coronary intervention or coronary artery bypass graft surgery. The decision to intervene surgically or per catheter depends on the clinical presentation, the patient’s hemodynamic state, the site and extent of the dissection, and the number of vessels involved. Stable patients with spontaneous CAD are limited to a single coronary artery dissection, usually have a favorable long-term outcome and are candidates for conservative medical management. Thrombolytic therapy is controversial. Thrombolysis might dissolve the compressing intramural clot but might also expand the hematoma and thereby proliferate the dissection. Stenting seems to be an attractive
option, for it can yield very good long-term results in symptomatic patients who have localized dissections. Coronary artery bypass grafting should be performed in patients with main stem or multivessel involvement or in those with hemodynamic instability. Angioplasty and stenting may be preferred in single-vessel dissection. CABG should be performed in patients who have LMCA or multivessel dissection or who have undergone stenting without success. We preferred CABG for treatment of our patient, because RCA was dominant, coronary dissection was long starting from RCA ostium to distal segment and stenting was not considered to be a viable option.

CONCLUSION

The diagnosis of SCAD should be considered in any patient with apical hypertrophic cardiomyopathy who presents with symptoms suggesting acute myocardial ischemia, particularly in young males without other risk factors of SCAD such as atherosclerosis, hypertension, illicit drug use, heavy exercise, Marfan syndrome or other collagen disease.

REFERENCES


