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Coronary and Intracerebral Arterial Aneurysms

in a Young Adult with Acute Coronary Syndrome

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A 21-year-old man with no known medical history presented with substernal chest pain. Serial 12-lead electrocardiography showed dynamic ST-segment elevations in the anterolateral leads. Emergent coronary angiography revealed diffuse coronary aneurysmal disease and thrombotic occlusion of the left anterior descending coronary artery. The patient underwent urgent coronary artery bypass grafting. Subsequent imaging showed intracerebral aneurysms that involved his right and left middle cerebral arteries. The incidence, multiple causes, and proposed mechanisms of coronary artery aneurysmal formation are discussed, as is the rare association of these lesions with extracardiac arterial aneurysms.

This association between coronary and extracardiac aneurysms is a phenomenon that warrants further study to determine its prevalence and possible causes. Findings could influence recommendations for further screening of patients diagnosed with coronary aneurysmal disease. ***(Tex Heart Inst J 2012;39(3):380-3)***

Key words: *Acute coronary syndrome; cerebrovascular aneurysms; congenital heart disease; coronary aneurysm/etiology; coronary vessel anomalies; dilatation, pathologic/etiology; intracranial aneurysm; Kawasaki disease*

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Coronary artery aneurysms, or ectasias, are a rare finding on coronary angiography. They are secondary to a variety of multifactorial processes and can be prone to thrombus formation, thromboembolism, vasopasm, dissection, and other sequelae. The incidence of associated extracardiac aneurysms is unknown, and the causal mechanisms are unclear.

Case Report

A 21-year-old man with no known medical history presented at our emergency department with first-time onset of severe substernal chest pain. He denied any history of significant childhood illness, such as Kawasaki disease or rheumatic heart disease. He was not taking any medications at home, and he had no first-degree relatives with any history of premature coronary artery disease or structural heart disease. He denied any history of chest pain, angina, dyspnea on exertion, or other signs of heart failure; in addition, he denied any constitutional symptoms of fever, weight loss, arthralgias, or skin lesions. He used alcohol and tobacco socially but denied any illicit drug use. There was also no travel history.

On physical examination, his temperature was 97.7 °F; pulse rate, 71 beats/min; blood pressure, 152/86 mmHg; respiratory rate, 22 breaths/min; and oxygen saturation, 100% on room air. The patient was alert and oriented but in mild distress from his chest pain. His jugular venous pressure was approximately 5 cm H_2O. His cardiovascular examination was significant only for a positive S_4 at the left sternal border. His lungs were clear on auscultation bilaterally, and no peripheral edema was present.

Serial 12-lead electrocardiography showed dynamic ST-segment elevations in the anterolateral leads that were indicative of an ST-elevation myocardial infarction (Fig. 1). A urine toxicology study was negative, but the troponin level was positive at 0.37 ng/dL. An urgent 2-dimensional transthoracic echocardiogram showed a left ventricular ejection fraction of 0.45, and hypokinesis of the anteroapical, apical septal, and inferoapical aspects of the heart, with no significant wall-motion abnormalities.

The patient was taken emergently to the coronary angiography laboratory, where he was found to have diffuse ectasia of the left anterior descending coronary artery (LAD), with a large proximal aneurysmal segment that was acutely occluded with thrombus (Fig. 2). The right coronary artery was also diffusely ectatic (Fig. 3). At-

tempts to cross the occlusion with a guidewire failed. An intra-aortic balloon pump was placed, and the patient was sent for emergent coronary artery bypass surgery, with a left internal mammary artery graft placed to the LAD (Fig. 4). The patient did well postoperatively, with no complications.

Because of the idiopathic nature of the patient's coronary anatomy, extracardiac vascular imaging was performed. Computed tomographic imaging of the head and neck revealed an aneurysm of the right internal carotid artery that extended into the right middle cerebral artery, and diffuse dilation of a branch of the left middle cerebral artery (Figs. 5 and 6). Computed tomography of the aorta revealed no aneurysms. Autoimmune and vasculitis laboratory markers were negative. Upon more specific history-taking, the patient denied any history of headaches or other neurologic abnormalities. After discussing his management with our neurosurgery and vascular surgery services, we decided to

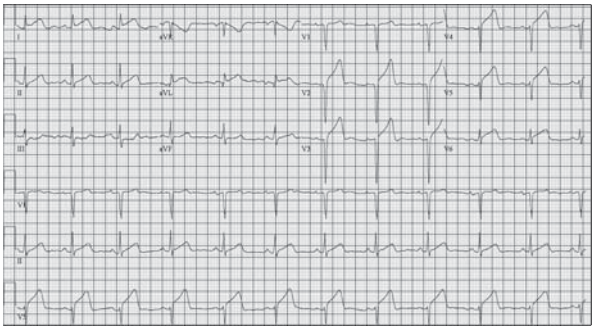

Fig. 1 *A 12-lead electrocardiogram shows diffuse ST-segment elevations in the anterolateral leads.*

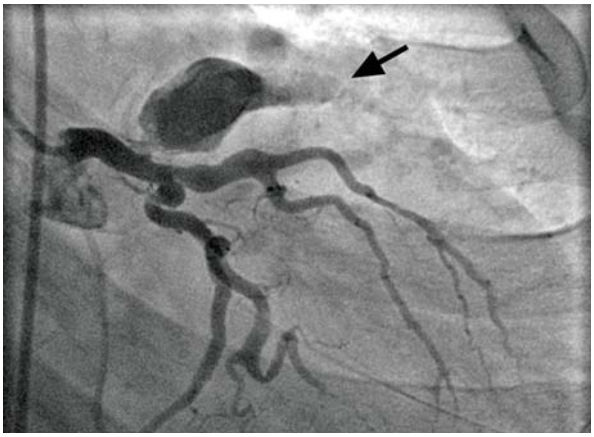

Fig. 2 *Coronary angiogram of the left main coronary artery (right anterior oblique caudal view) shows acute thrombotic occlusion (arrow) of the proximal left anterior descending coronary artery with severe aneurysmal disease proximal to the occlusion. There is also diffuse ectasia of the left circumflex coronary artery.*

Click here for real-time motion image: Fig. 2.

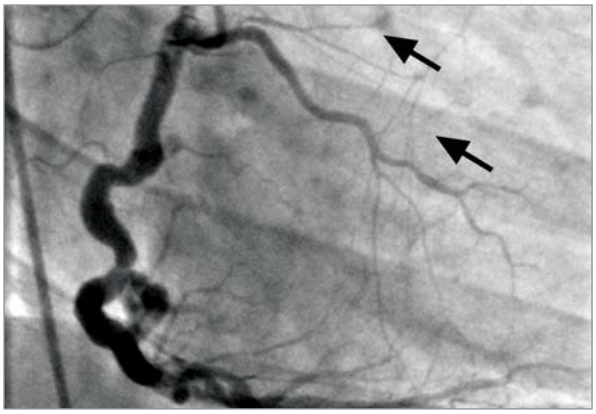

Fig. 3 *Coronary angiogram of the right coronary artery (right anterior oblique view) shows diffuse ectactic disease with no obstructive coronary artery disease. Right-to-left collateral vessels (arrows) extend from the conus branch and the posterior descending artery to the mid-left anterior descending coronary artery septal branches.*

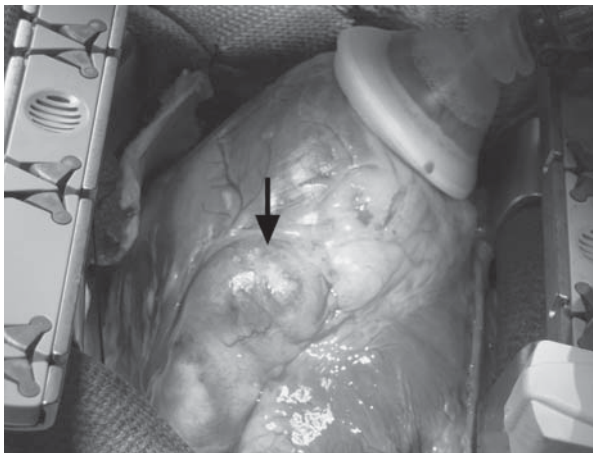

Fig. 4 *Intraoperative photograph of the heart shows a large, aneurysmal, and thrombosed proximal left anterior descending coronary artery (arrow), 3 × 6 cm.*

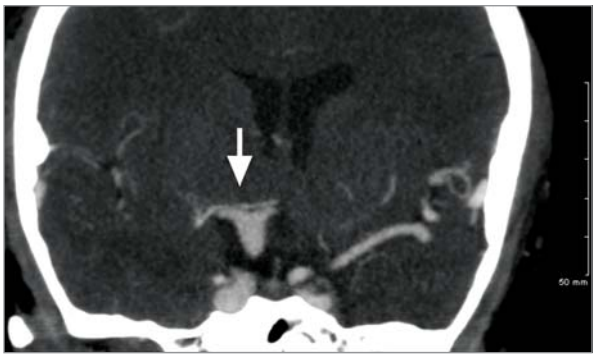

Fig. 5 *Computed tomographic angiogram of the head shows fusiform enlargement of the right internal carotid artery, measuring 7.7 mm in diameter (arrow), with extension into the M1 and A1 segments of the right middle cerebral artery. The distal right A1 segment is approximately 6 mm in diameter. There was also diffuse dilation of a branch of the left middle cerebral artery (not shown).*

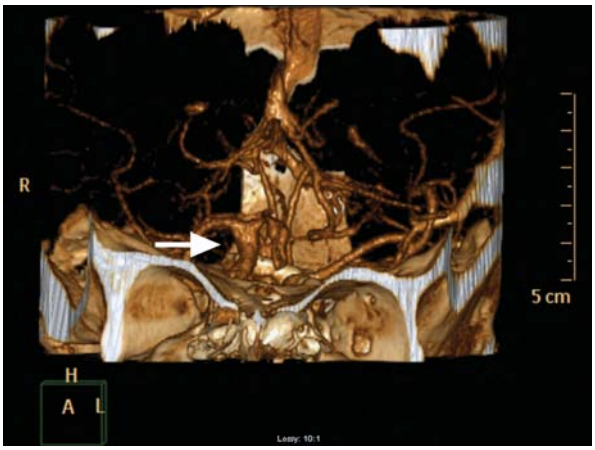

Fig. 6 *Computed tomographic angiogram (3-dimensional reconstruction) of the head and neck vessels shows the aneurysm of the distal right internal carotid artery and middle cerebral artery (arrow).*

monitor the patient by means of annual serial imaging. The patient was discharged from the hospital on postoperative day 6 in good condition, on a regimen of aspirin, clopidogrel, and metoprolol. The patient continued to do well in an outpatient clinic.

Discussion

Coronary artery aneurysms have been defined as a dilation in a coronary artery segment to more than 1.5 times the diameter of adjacent normal coronary segments.[1] The prevalence of coronary artery aneurysms ranges widely from 0.3% to 5.3%,[2] on the basis of several early angiographic studies. Markis and colleagues[3] devised a classification system in a study of 30 patients with coronary artery ectasia: diffuse ectasia of 2 or 3 vessels was classified as type I, diffuse disease of 1 vessel and localized disease in another vessel as type II, diffuse ectasia of 1 vessel only as type III, and localized or segmental ectasia as type IV.

The causes of coronary artery aneurysms are multifactorial: approximately 50% of coronary artery aneurysms are thought to be caused by atherosclerosis, another 20% to 30% are thought to be due to congenital causes, and 10% to 20% are due to inflammatory or connective-tissue disorders, such as Kawasaki disease, Takayasu arteritis, lupus, or rheumatoid arthritis.[4] In younger patients, atherosclerotic disease is less likely a contributing factor, and congenital and acquired causes from childhood illnesses such as Kawasaki disease are more likely. A review of 50 reported cases[5] of adult patients who had acute coronary syndromes due to known or suspected Kawasaki disease reported a median age of 28 years (range, 18–69 yr); 54% presented with LAD occlusion. The most common type of lesion consisted of giant calcified aneurysms in the proximal portion of the coronary arteries—43% of patients had giant aneurysms with coronary calcification, and 38% had only giant aneurysms. Only 3 patients had no aneurysm or coronary artery calcification. Six percent of the 50 patients died.[5] A retrospective analysis of 60 Kawasaki patients who had a myocardial infarction[6] (median age, 2 yr; range, 3 mo–33 yr) showed a 30-year survival rate of 62.7% and a 25-year ventricular tachycardia-free survival rate after myocardial infarction of 28.5%. The survival rate after myocardial infarction was poor for patients who had a postinfarction left ventricular ejection fraction of less than 0.45.[6] Although extracardiac aneurysms are not frequently associated with Kawasaki disease and there are no current indications to screen for them,[7] they are reported to have been found in neurovascular and large vessel territories.[8]

In addition, the increased use of angioplasty and stents in the modern interventional period has given rise to an iatrogenic form of postintervention coronary artery aneurysm.[9] The underlying mechanism behind the development of coronary artery aneurysms is not clear, although on a molecular level, matrix metalloproteinases (MMPs) might contribute to the development of aneurysms through increased proteolysis of extracellular matrix proteins. Lamblin and colleagues[10] showed that the MMP-3A allele was associated with the occurrence of coronary artery aneurysm in an older population (average age, 62 years).

Overall, the most studied coronary artery aneurysms are atherosclerotic in origin, and in older patients.[10-12] Postmortem histologic evaluation of these patients with coronary artery aneurysms revealed underlying changes that were similar to those in patients with atherosclerotic lesions—diffuse hyalinization, together with intimal and medial damage—which suggested an overlapping pathophysiologic mechanism.[2,9] Risk-factor studies that have compared patients with and without aneurysmal disease have uncovered varying prevalence of hypertension, diabetes mellitus, dyslipidemia, and smoking.[2,11,12] In a cohort of coronary artery aneurysm patients at Emory University Hospitals,[12] investigators found coronary artery aneurysm to be an independent predictor of death, with overall 5-year survival of coronary artery aneurysm patients at 71%. Of the patients in the study, 84% were over the age of 50 years, and 71% of both aneurysmal and control patients had concurrent obstructive coronary artery disease. A more stringent criterion was used to define coronary artery aneurysm: an aneurysmal segment had to be at least 2 times the diameter of the adjacent normal artery or had to have an absolute ectactic diameter of 8 mm or more. There was no statistically significant difference between survival curves of aneurysmal patients with or without obstructive coronary artery disease.[12]

In case reports, there has been a rare association between coronary artery aneurysm and intracranial aneurysm—particularly of the basilar artery. Other vascular

regions, including the abdominal aorta and varicose veins, have been involved as well.[13-15] Aneurysms of the aorta and its branches are most commonly thought to be of atherosclerotic origin in older patients and of congenital origin (possible defects in the vascular wall) in younger patients. Given the lack of studies in which patients with coronary artery ectasia have been screened for extracardiac aneurysms, the prevalence, cause, prognosis, and treatment options for patients with this association are unknown; in fact, descriptions of these unusual findings have been mostly confined to case reports. In a small prospective trial, Lamblin and colleagues[16] screened coronary artery aneurysm patients for abdominal aortic aneurysms and found a significant association: more than 20% of these patients had asymptomatic abdominal aortic aneurysms, versus 5% of patients in a control group. Notably, this study was done in an older population (average age in the coronary artery aneurysm group, 65 years).

In summary, we have presented the case of a young man with no known medical history who presented with acute thrombotic occlusion of an aneurysmal LAD, together with extracardiac manifestations in the form of intracranial aneurysms—due either to congenital causes or to Kawasaki disease. This association between coronary and extracardiac aneurysms is a phenomenon that warrants further study to determine its incidence and possible causes. The results of such studies could influence recommendations for further screening of patients diagnosed with coronary aneurysmal disease.

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