Unusual Presentation of an Acute Inferior Myocardial Infarction

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There is a paucity of data on pseudoaneurysms of native coronary arteries; however, several reports exist on coronary artery aneurysms, which occur in approximately 1.5% of patients studied at autopsy or during cardiac catheterization. Patients can present with a wide range of symptoms from asymptomatic to sudden death. Complications include angina, myocardial infarction, fistula formation, spontaneous rupture, and distal embolization as a result of thrombus formation within the aneurysm. Treatment options include surgical ligation with coronary artery bypass surgery and implantation of a covered stent. Coronary anomalies and nonatherosclerotic coronary artery diseases should be suspected when a young patient presents with a myocardial infarction. Additionally, coronary aneurysm or pseudoaneurysm should be considered in patients with connective tissue disorder or the suggestion of connective tissue disorder. These entities may present as masses radiographically or echocardiographically. A high clinical suspicion is required for appropriate diagnosis and treatment. [Rev Cardiovasc Med. *2002;3(3):152–156*]

Key words: Myocardial infarction • Mitral valve prolapse • Pseudoaneurysm • Connective tissue disorder • Coronary artery

SG is a 38-year-old male with a history of mitral valve prolapse and severe mitral regurgitation requiring a mitral valve repair in 1995. With failure of this repair, he underwent mitral valve replacement (MVR) of the St. Jude's valve in 1998. He had been doing well since the MVR with scrupulous attention to anticoagulation and antibiotic prophylaxis. Three years following MVR, he presented with the complaints of fatigue and decreased exercise tolerance of 6 weeks duration.

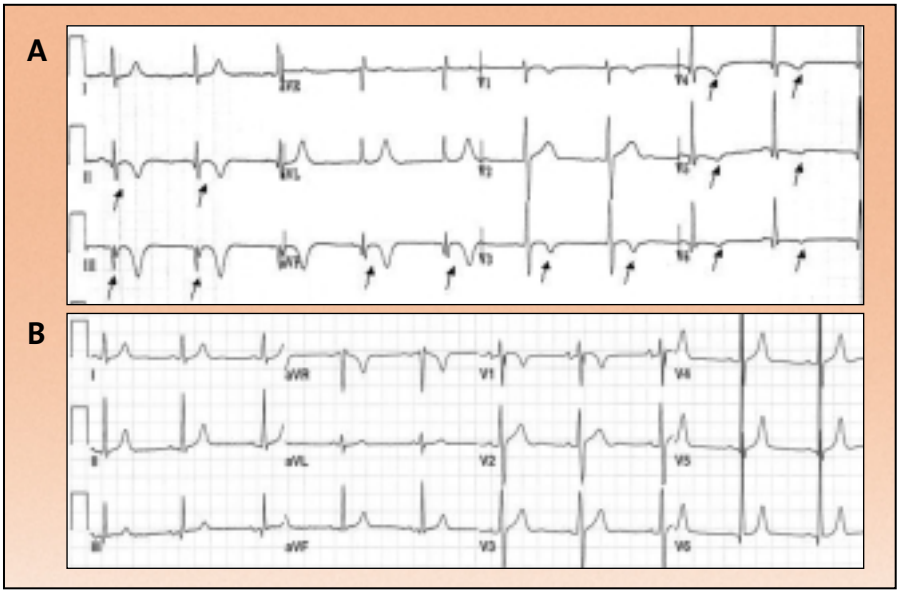


Figure 1. *Twelve-lead electrocardiogram showing new Q waves in the inferior leads and new T wave inversions in the inferior and lateral leads (black arrows) at the time of presentation* **(A)**, *which were not present on a previous EKG obtained at the time of mitral valve replacement in 1998* **(B)**.

He denied any chest pain at rest or with exertion, palpitations, syncope, or edema. There was no history of recent fevers, chills, nausea, or vomiting. He was unable to tolerate his usual regimen of aerobic exercise for 30–45 minutes 3–4 times a week. He did recall one episode of light-headedness associated with marked fatigue.

History

SG's past medical history included mitral valve prolapse diagnosed at age 12, spontaneous pneumothorax, inguinal hernias with repairs, and gastroesophageal reflux disease. His cardiac surgical history is as stated above. The intraoperative findings at the time of MVR included mediastinal adhesions, elongated chordae, and prolapsed anterior and posterior leaflets. No anomalies of the coronary arteries were noted. The leaflets were excised, and a St. Jude's mitral valve was placed without complications.

The postoperative transesophageal echocardiogram revealed a normally functioning mitral prosthesis and normal left ventricular wall motion. The pathology revealed fibrosis and myxoid degeneration, and blood cultures were negative. The postoperative course was uneventful except for transient atrial fibrillation, and the patient was discharged on metoprolol, aspirin, and coumadin.

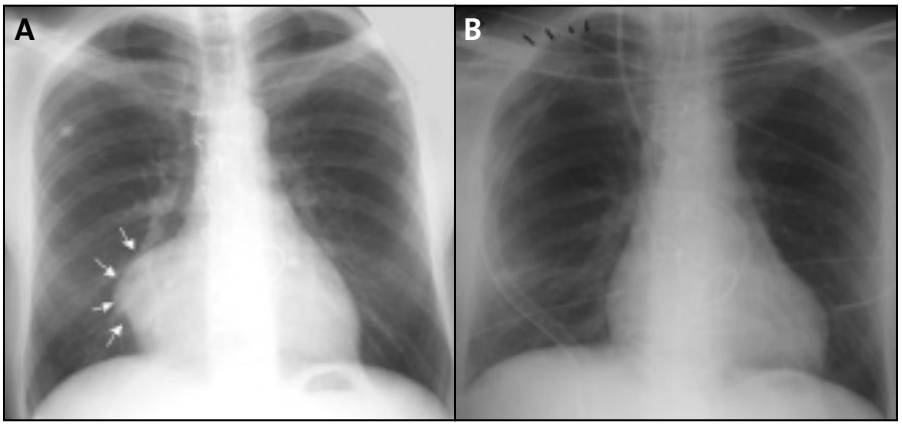
His family history was significant for mitral valve prolapse, along with an inguinal hernia in his father and hypertension in his mother. There was no family history of premature coronary disease. SG denied any current or prior tobacco, alcohol, or drug use.

Examination

Examination revealed a thin, tall male, lying flat in bed in no apparent distress. He was afebrile, heart rate was 56 beats per minute, blood pressure was 110/56 mm Hg, and respiratory rate was 18 breaths per minute. His head and neck examination was unremarkable. His sclerae were anicteric and no ectopia lentis was noted. There was no jugular venous distension. Precordial examination revealed a nondisplaced point of maximal impulse and no heaves. There was a mechanical S1 and a normal S2. No murmurs, gallops, or rubs were appreciated. His lungs were clear to auscultation. The abdomen was benign. There was no cyanosis, clubbing, or edema noted. The neu-

A St. Jude's mitral valve was placed without complications.

Figure 2. *PA chest x-ray showing a large density (white arrows) along the right heart border* **(A)** *that was not present on the chest x-ray following the mitral valve replacement in 1998* **(B)**.



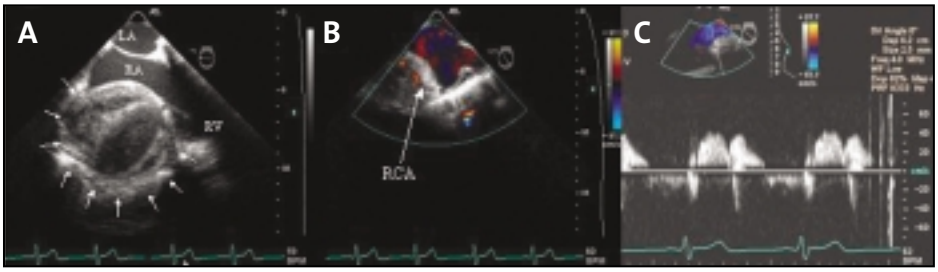


Figure 3. *Photographs of transesophageal echocardiographic images showing a large coronary artery pseudoaneurysm compressing the right atrium and right ventricle* **(A)** *and predominantly systolic flow into the pseudoaneurysm on color flow* **(B)** *and pulsed-wave Doppler* **(C)**.

rological examination was without focal deficits.

Laboratory data revealed a basic metabolic panel and a complete blood count within the normal ranges. The total cholesterol was 165 mg/dL, low-density lipoprotein cholesterol was 71 mg/dL, high-density lipoprotein cholesterol was 39 mg/dL, and triglycerides were 61 mg/dL.

Other studies, including an electrocardiogram (EKG), showed new Q waves and T wave inversions in the inferior leads compared to a prior EKG in 1998 (Figure 1). A chest x-ray showed a density along the right heart border that was new compared to a previous chest x-ray from 1998 (Figure 2). A transthoracic echocardiogram revealed a normally functioning mitral prosthesis with physiologic mitral regurgitation. The new findings compared to previous echocardiograms included akinesis of the inferoposterior walls with a left ventricular ejection fraction of 40%, hypokinesis of the right ventricle, and a large mass outside the right atrium and basal right ventricle.

A transesophageal echocardiogram confirmed the presence of a 6.5 × 6.5 cm mass compressing the right atrium and basal right ventricle. The mass appeared to be cystic and lined with thrombus (Figure 3A). A dilated and tortuous vessel with predominantly systolic flow was seen coursing through the mass (Figures 3B and 3C). In addition, a patent foramen ovale with bidirectional flow and a dilated aortic root (4.2 cm) were seen.

A resting thallium study revealed a large, partially reversible, resting perfusion defect in the inferior and inferolateral walls, with severe hypokinesis in the inferior and septal walls and a reduced ejection fraction of 45% (Figure 4).

He denied any chest pain at rest or with exertion, palpitations, syncope, or edema.

Cardiac catheterization revealed normal left main, left anterior descending, and circumflex arteries. The right coronary artery was dominant and was notable for a large pseudoaneurysm (PA) around 6 cm in its mid-portion, after the takeoff of the right ventricular branch (Figure 5). Severe hypokinesis of the distal inferior and apical walls and hypokinesis of the posterolateral walls were noted, with an estimated ejection fraction of 45%–50%.

Hospital Course

After cardiac catheterization, the patient developed a femoral PA of 2.6 × 1.5 cm, which was repaired using ultrasound-guided thrombin injection.

The options of using a covered stent or coronary artery bypass surgery with ligation of the PA were discussed in length. Favoring stent application in this case was the avoidance of a third open-chest operation in a patient with a dilated aortic root who may ultimately require operative repair. However, anatomic considerations, particularly the severe tortuosity of the right coronary artery, made the likelihood of safe implantation of a relatively inflexible covered stent small.

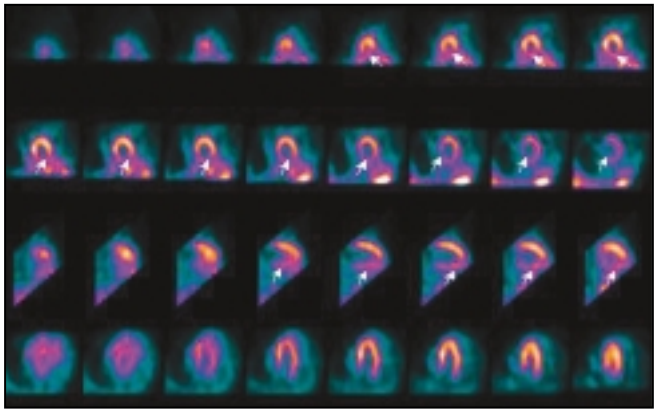


Figure 4. *Resting thallium study reveals a large inferior and inferolateral defect on thallium uptake (white arrows).*

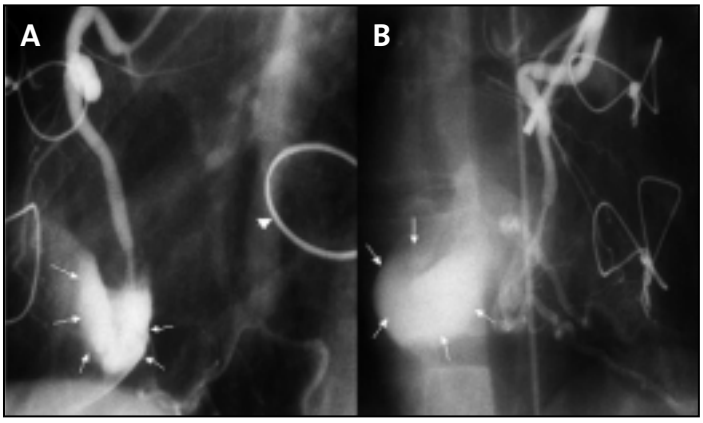


Figure 5. *Cardiac catheterization reveals an ectatic, tortuous right coronary artery and a large pseudoaneurysm in its mid-portion (white arrows) in* **(A)** *anteroposterior and* **(B)** *lateral views. White arrow head points to the St Jude's mitral valve ring.*

A constellation of findings, including mitral valve prolapse, dilated aortic root, patent foramen ovale, spontaneous pneumothorax, inguinal hernias, and the recently diagnosed coronary PA and postprocedural femoral PA were highly suggestive of a connective tissue disorder (CTD), such as Marfan syndrome, Ehlers-Danlos syndrome, or osteogenesis imperfecta. Given this possibility, he was considered to be at increased risk of an iatrogenic coronary dissection with stent placement. Upon review of the literature, no data were found comparing covered stents to bypass surgery in patients with CTD. Although a recent report demonstrated success of transcatheter closure of coronary artery fistulae comparable to that of historical surgical closure,[1] there are no data currently available comparing covered stents to bypass surgery in the setting of native coronary PA.

The patient eventually underwent an open-chest operation. Intraoperative findings included a PA filled with thrombus with antegrade blood flow from the right coronary artery through a fistula. The PA was ligated and bypass of the distal right coronary artery was performed using a left radial conduit. There were no complications following surgery. The patient was discharged home in stable condition and remains stable at last follow-up.

Discussion

An underlying CTD is a distinct possibility given the constellation of findings of mitral valve prolapse, dilated aortic root, patent foramen ovale, right coronary PA, inguinal hernias, spontaneous pneumothorax, postprocedural femoral artery PA, and a family history of mitral valve prolapse and inguinal hernias. The etiology of the PA remains unclear. It may have been spontaneous and related to an underlying CTD or to a traumatic event from one of his previous mitral valve surgeries. A spontaneous coronary dissection resulting in PA formation may explain the patient's presentation.

The PA, which was lined with thrombus, communicated with an abnormal tortuous right coronary artery. The development of an inferior infarction could have been related to distal embolization of thrombus from the PA into the distal right coronary bed territory or arterial occlusion resulting from a spontaneous dissection of the right coronary artery with subsequent PA formation.

There is a paucity of data on PA of native coronary arteries. This patient is a unique case of an inferior infarction that developed as a result of a spontaneous or iatrogenic PA in the setting of a possible underlying CTD and that was successfully treated surgically.

As opposed to coronary PA, several reports exist on coronary artery aneurysms. Coronary artery aneurysms occur in approximately 1.5% of patients studied at autopsy or during cardiac catheterization, although a higher incidence (4.9%) was reported in the Coronary Artery Surgery Study (CASS) registry.[2] The causes of coronary aneurysms are varied, including atherosclerosis (50%),[3] trauma,[4] prior coronary interventions, syphilis, Kawasaki's disease,[5] systemic lupus erythematosus,[6] spontaneous or secondary dissections, and CTDs including Marfan syndrome,[7] Ehlers-Danlos syndrome,[8,9] and osteo-

Main Points

- Coronary artery aneurysms occur in approximately 1.5% of patients studied at autopsy or during cardiac catheterization.
- Coronary aneurysm or pseudoaneurysm should be considered in young patients with connective tissue disorder presenting with myocardial infarction.
- Coronary aneurysms and pseudoaneurysms may present as masses radiographically or echocardiographically.
- Treatment options include surgical ligation with coronary artery bypass surgery and implantation of a covered stent.

genesis imperfecta.[10] Patients can present with a wide range of symptoms from asymptomatic to sudden death. Other symptoms include dyspnea, fatigue, chest discomfort, palpitations, and syncope.[3] Complications include angina, myocardial infarction,[11] fistula formation,[12] spontaneous rupture,[13] and distal embolization as a result of thrombus formation within the aneurysm.

Treatment options include surgical ligation with coronary artery bypass surgery and implantation of a covered stent. There have been several case reports of successful bypass surgery to repair coronary aneurysms. Additionally, successful bypass surgery has been described in patients with CTD, including Marfan syndrome[7] and osteogenesis imperfecta.[10] Covered stent use has also been successful in isolating coronary aneurysms[14,15] and coronary artery fistulae,[1] though not in patients with CTD.

In summary, coronary anomalies and nonatherosclerotic coronary artery diseases should be suspected when a young patient presents with a myocardial infarction. Additionally, coronary aneurysm or PA should be considered in patients with CTD or the suggestion of CTD. These entities may present as masses radiographically or echocardiographically. A high clinical suspicion is required for appropriate diagnosis and treatment. As in our patient, coronary aneurysms or PA can be successfully repaired with bypass surgery. ■

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