

Caseous Mitral Annular Calcification: An Unrecognized Cause of Cardioembolic Events

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Abstract

INTRODUCTION

Caseous mitral annular Calcification (CMAC) is a rare, benign and under-recognized condition. The echocardiographic prevalence of CMAC is 0.06-0.07% of the general population and 0.64% in the patients with mitral annular calcification (MAC) whereas prevalence in necropsy series is 2.7%. CMAC is the chronic degeneration of the mitral valve fibrous ring and liquefaction necrosis of MAC. It involves mainly the posterior annulus. Here, we present a case of CMAC in a 60-year-old woman who presented with transient amnesia.

CASE PRESENTATION

A 60-year-old female presented to the emergency department after a brief period of amnesia which lasted for an hour without any other focal neurological deficit. Her medical history is significant for hypertension and mixed hyperlipidemia. Her blood pressure on arrival was elevated to 200 systolic which responded to two doses of intravenous labetalol. CT angiogram of head and neck showed calcified atherosclerosis at the origins of bilateral internal carotid artery without stenosis. MRI of the brain was negative for any acute event. EKG showed normal sinus rhythm with P-mitralae. Echocardiogram revealed severe MAC with a large echogenic mass with central echolucent area with mid left ventricle cavity obstruction. Transesophageal echocardiogram (TEE) [Image-1] confirmed 3.44 x 2.85 cm CMAC with mild mitral stenosis (MS) and mitral regurgitation (MR). After discussion at a multidisciplinary conference, the decision was made to manage the patient medically.



Image-1

DISCUSSION

CMAC is typically composed of an admixture of calcium, fatty acids, and cholesterol and has “toothpaste-like” texture. Higher prevalence of CMAC is found in patients with end-stage renal disease particularly those on hemodialysis. The observed association with hypercalcemic state suggests that there is a relationship between CMAC and altered calcium-phosphate metabolism. CMAC can cause MS, MR, left ventricular outflow obstruction, systemic embolization or conduction abnormalities. TEE is a reliable method for the diagnosis of this condition. In equivocal cases, MRI can be used. The optimal management of CMAC remains controversial. The current data suggests conservative medical management when there is no obstruction of left atrial emptying. Treatment with low calcium hemodialysis was associated with regression of the CMAC. Hence, theoretically, changes in serum calcium level may contribute to the resolution of the mass. The current indications for surgical intervention are mitral valvular dysfunction, embolic manifestations or high suspicion for tumor. Mitral valve replacement is preferred over repair. Medical management was chosen in this case because the patient presentation was thought related to her high blood pressure.

CONCLUSION

It is important for both cardiologists and cardiac sonographers to be familiar with this incidental calcific mass since it can easily be confused with a calcific peri-annular tumor, vegetation or abscess. In addition, though often a benign finding, CMAC may lead to embolic events or structural mitral valve dysfunction. Therefore, careful follow-up is necessary if they developed new symptoms or have a high propensity for tissue calcium deposition. Evaluation with TEE should be considered in all cases of suspected embolic stroke.

Categories

1st year Fellow: Case

Program/Institution Name

Summa Health System/NEOMED

Takotsubo cardiomyopathy in the setting of ST-Elevation Myocardial Infarction

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Abstract

Title: Takotsubo cardiomyopathy in the setting of ST-Elevation Myocardial Infarction

Introduction/Objective

Takotsubo cardiomyopathy (TCM), also referred to as stress-induced cardiomyopathy was first described by Japanese authors in the 1990s. The syndrome is characterized by transient left ventricular systolic dysfunction, myocardial enzyme elevation and electrocardiographic features consistent with acute myocardial infarction (AMI) but in the absence of obstructive epicardial coronary artery disease (CAD). TCM is usually distinct from CAD and AMI. However, co-existing AMI and TCM is an under-recognized entity.

Case Presentation

The patient was a 77 year old female with a past medical history of antiphospholipid antibody syndrome who presented to the emergency department with substernal chest discomfort. She was found to have lateral ST-Elevation Myocardial Infarction (STEMI) upon presentation.

Patient was taken for percutaneous coronary intervention and was found to have severe single vessel disease. There was a 100% occluded mid RCA which was subsequently treated with drug-eluting stent. Ventriculogram was completed and was consistent with a stress-induced cardiomyopathy pattern along with a focal inferior wall motion abnormality likely related to the AMI. Her left ventricular function was previously normal. Moderately elevated left ventricular end-diastolic pressure was also noted. The patient's transthoracic echocardiogram revealed akinesis of the mid-distal anterior wall, septum, and apex consistent with stress cardiomyopathy. The inferior infarct alone does not explain the electrocardiographic features or the wall motion abnormalities. Cardiac MRI was done to further differentiate whether this was stress-induced cardiomyopathy in the LAD territory. The MRI revealed Diminished left ventricular systolic function with akinesis of the mid to distal anterior wall septum and apex, thinning of the distal anterior septal wall and apex of delayed myocardial enhancement most compatible with transmural LAD distribution infarct

Discussion

In the present case the patient did not report about emotional stress preceding chest pain. However, STEMI is frequently associated with severe emotional stress. The pathogenesis of TCM is not entirely known, but is believed to be related to the sudden release of stress hormones, such as catecholamines, causing cardiac stunning and subsequent changes in the cardiac myocytes and coronary perfusion. A previous case report has reported simultaneous AMI and TCM. However very little is known about the association between AMI and TCM.

Conclusion

This case adds to the limited literature of TCM in association with AMI. These two distinct condition can occur simultaneously. Although causality cannot be determined, it is believed that the stress associated with AMI may have caused TCM in this patient.



Categories

1st year Fellow: Case

Program/Institution Name

University of Toledo

Beyond Scimitar Syndrome: A Unique Combination of Complex Congenital Heart Disease

77

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Abstract

Background: Scimitar Syndrome is a rare entity within congenital heart disease. The clinical presentation may be insidious and associated with a number of cardiopulmonary anomalies.

Case: A 47 year old male presented with progressive exertional dyspnea, palpitations, and a new systolic murmur in the setting of known dextrocardia. He underwent TTE which showed mildly reduced biventricular function, dilated RV, and moderate TR.

Decision-Making: Given the suspicion for underlying congenital heart disease, the patient underwent cardiac MRI, X-ray, and CTA (Figure 1) which confirmed dextrocardia with evidence of Scimitar syndrome with all right sided pulmonary veins draining into the IVC, a Qp/Qs of 1.6:1, and concomitant dilation of the RA, RV, and PA. Additional findings included an anomalous single coronary artery originating from the right coronary cusp, aortic root dilation (5 cm) and moderate TR. Surgical repair was performed with in-situ pericardial repair of scimitar vein routing to the LA, repair of the TV, and repair of the noncoronary aortic sinus with CardioCel. On follow up, the patient reported resolution of his dyspnea on exertion and improved functional capacity.

Conclusion: This unique case highlights dextrocardia with Scimitar Syndrome, aortopathy, and anomalous single coronary artery. This complex congenital case went undiagnosed for nearly five decades with only mild symptom burden. Multimodality cardiac imaging played a crucial role in the diagnosis and surgical planning of this case.

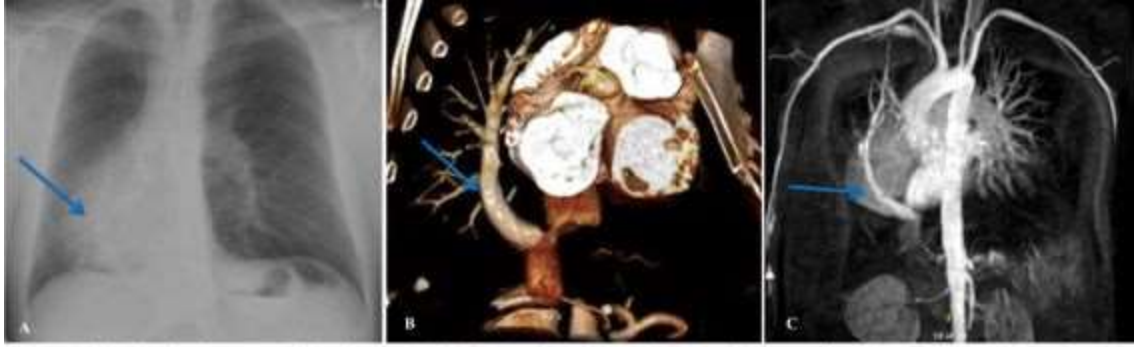


Figure 1: Scimitar Syndrome (A) Chest X-ray and (B) CTA showing the scimitar vein and dextrocardia, and (C) Cardiac MRI showing the scimitar vein with drainage to the IVC

Categories

1st year Fellow: Case

Program/Institution Name

Ohio State University Hospital

A Ghost Left Behind After Transvenous Lead Extraction: A Finding To Be Feared.

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Abstract

Introduction

Cardiac device-related infective endocarditis (CDRIE) necessitates a transvenous lead extraction (TLE) procedure for removal of all hardware. Following TLE, there may be a persistent intra-cardiac remnant sheath referred to as a “ghost” (8-14%). First reported in 2008, a ghost remains a novel entity understudied with no guideline-directed management.

Case Presentation

We present a case of a 72 year old male who was found to have a ghost post-TLE and who underwent leadless pacemaker (LP) placement as well as subcutaneous implantable cardioverter defibrillator implantation (SICD). He presented after a mechanical fall with generalized malaise and was found to have methicillin-resistant staphylococcus aureus (MRSA) bacteremia. After echocardiographic evidence of endocarditis, he underwent a TLE to remove a cardiac resynchronization therapy defibrillator (CRT-D) device. Due to a previous atrioventricular (AV) node ablation for drug-refractory atrial fibrillation, he was pacemaker-dependent and thus underwent placement of a temporary transvenous wire. After culture negativity, an LP device was placed in the right ventricular apex and transvenous pacemaker was removed. Following 6 weeks of parenteral antibiotic therapy, a transesophageal echocardiogram revealed a 1.3 cm tubular and mobile ghost in the right atrium, which preceded SICD placement. He was subsequently discharged with plan for close monitoring.

Discussion

When pathologically examined, ghosts consist of infected fibrous sheaths mixed with vegetation; In multiple prospective studies, they are associated with increased mortality and CDRIE relapse/reoccurrence. However, according to expert consensus, no specific therapy is indicated for

patients with ghosts. The infectivity of ghosts raises concerns for proper management, antibiotic therapy, as well as the role of LP in this setting. Also, pertinent questions remain regarding the role of serial imaging, risk factor stratification, and predisposing conditions.

Conclusion

Further description of the role of ghosts toward clinical outcomes may avert the residual risk as a large body of evidence confirms that ghosts are to be feared rather than ignored.

Categories

1st year Fellow: Case

Program/Institution Name

Doctors Hospital/OhioHealth

Complete heart block- A manifestation of aortic root abscess in a patient with bioprosthetic aortic valve and bacteremia.

26

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Abstract

INTRO

Infective endocarditis (IE) is a life-threatening complication in patients with prosthetic heart valves. Early diagnosis and prompt management are necessary to prevent significant morbidity and mortality. We present an interesting case of IE involving a bioprosthetic aortic valve, aortic root abscess, and complete heart block requiring emergent surgical intervention. Emphasis must be given for early recognition of clinical signs of IE so as to prevent its complex course.

CASE

A 78-year-old diabetic male patient with a history of bioprosthetic aortic valve replacement in 2014 (23mm mosaic porcine valve) for severe calcified aortic valve stenosis.

In March 2019, the patient presented to an urgent care for intermittent fevers and cough. He was medically treated with steroids and antibiotics for suspected bronchitis. However symptoms persisted so that he presented to the emergency department(ED) 2 months later in May 2019. He was again sent home with a course of antibiotics for a suspected upper respiratory tract infection.

Three months later in June, he presented again to the ED with fever, night sweats, and shortness of breath. He was admitted and treated for new onset congestive heart failure and possible pneumonia. Blood cultures grew pan-sensitive *Enterococcus faecalis* and IV antibiotics were started. A transthoracic echocardiogram was of poor quality, and a transesophageal echocardiogram showed perivalvular aortic root abscess with valve dehiscence (Figure- 1) and mild to moderate aortic regurgitation.

Meanwhile, the patient also developed asymptomatic complete heart block on EKG. He was referred for emergent surgical intervention. The operative findings included severely infected aortic valve with gross purulence from the aortic root to the left main coronary artery. The valve was able to be removed almost without cutting any sutures due to extensive root abscess. Aortic root was successfully repaired with a 21mm Homograft with reimplantation of coronary arteries. Post-operative recovery was uneventful, the patient regained normal sinus rhythm, left ventricular ejection fraction was normal at 55%. He was discharged to a nursing facility with a 6 week course of intravenous antibiotics.

DISCUSSION

To prevent morbidity and mortality in patients with prosthetic heart valves, it is extremely important to recognize the early signs of IE so that prompt and appropriate treatment can be started. New EKG changes i.e. first degree AV block, bundle branch block, or complete heart block may be indicative of underlying aortic root abscess. This is due to the fact that the conduction system is contained within the membranous interventricular septum which is in close proximity with the right- and non-coronary aortic cusps. This case highlights the importance of considering IE as a differential diagnosis when assessing patients with prosthetic heart valves, fever, and heart failure in the ER.



Figure 1: TEE Short axis view showing dehiscence aortic valve

Categories

1st year Fellow: Case

Program/Institution Name

Kettering Health Network

Program/Institution Name If NOT listed in the prior question.

Dayton Veterans Affairs Medical Center

An Uncommon Cause of ST Elevation Myocardial Infarction

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Abstract

Introduction:

Acute aortic dissection is a potential life-threatening cause of chest pain in the emergency department. However, it is an uncommon cause of ST elevation on electrocardiogram. We present a case of Inferior STEMI elevation MI as a consequence of type A aortic dissection that was confirmed using aortography.

Case:

A 52-year-old gentleman with unknown past medical history came to the hospital with ongoing shortness of breath and chest pain for one day. Upon further questioning he did admit to discomfort in his thoracic back that was pleuritic in nature. He was visibly uncomfortable in bed, diaphoretic and writhing in pain. He clinically presented in cardiogenic shock with blood pressure 80/40 and heart rate of 110. His initial troponin level was only 0.22 ng/mL. Initial chest x-ray showed mild cardiomegaly. ST changes involving the inferior leads were noted on the EKG and the STEMI alert was called. Patient was taken urgently to the cardiac catheterization laboratory for emergent catheterization. Angiography was performed and showed no obstructive coronary disease; however, aortogram revealed aortic root dilatation with a type A aortic dissection extending from the right coronary cusp all the way up to the right innominate artery. The patient was managed aggressively with intravenous fluids which maintained blood pressure. A tertiary care center was immediately contacted and emergent transfer was initiated for definitive surgery.

Discussion:

The diagnosis of aortic dissection is often difficult and mimics many other causes of life-threatening cardiovascular and pulmonary emergencies that present as chest pain. The incidence of aortic dissection is far less common. The availability bias plays a major role in this delayed diagnosis. This propagates a problem in prognosis since each hour without treatment significantly increases mortality. Since the incidence of aortic dissection is not going to dramatically increase this bias will always be a major deterrent in timely diagnosis. Physician awareness can overcome many of these medical diagnostic biases.

Conclusion:

This was a 52-year-old gentleman who presented to the emergency department with atypical chest pain initially believed to have acute coronary syndrome from the electrocardiogram and laboratory data but quickly diagnosed with acute aortic dissection after aortogram. Despite his atypical symptoms he was misdiagnosed. Biases in the medical diagnosis have been talked about for decades but we must continue to raise awareness for our practices and our patients.



Categories

1st year Fellow: Case

Program/Institution Name

Canton Medical Education Foundation/Aultman Hospital/NEOMED

ORAL PRESENTATION ABSTRACTS

Ventricular Arrhythmia Prevalence and Characteristics for HIV+ Persons and Matched Uninfected Controls

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Abstract

Introduction:

Sudden cardiac death and myocardial fibrosis are common in HIV. No studies to our knowledge have examined the prevalence and morphology of ventricular ectopy or arrhythmia (VEA) for HIV+ versus uninfected persons.

Methods:

We screened 5,041 HIV+ persons and 10,121 uninfected controls (matched 1:2 on demographics and location) at an urban medical center between 2000 and 2016 for VEA using administrative codes. We then reviewed electrocardiographic data to determine (1) whether VEA were present, and (2) VEA morphology (left or right bundle and inferior or superior axis). Prevalence and morphology of VEA were compared by HIV status and markers of HIV severity.

Results:

Of 5041 HIV+ persons, 139 (2.8%) had VEA vs. 165 out of 10121 (1.6%) for controls ($p < 0.001$). This association persisted after adjustment for demographics (Odds Ratio [OR] 1.53, 95% Confidence Interval [CI] 1.21-1.94) but was attenuated to non-significance after adjustment for diabetes and hypertension. Compared with HIV+ persons with nadir $CD4 \geq 200$ cells/mm³, those with nadir $CD4 < 200$ cells/mm³ had significantly elevated odds of VEA after adjustment for demographics, diabetes, and hypertension (OR 1.65, 95% CI 1.12-2.31). Likewise, each log₁₀ higher peak HIV viral load was associated with a significantly elevated odds of VEA (OR 1.24, 95% CI = 1.07-1.44) after adjustment for demographics, hypertension, and diabetes. Right bundle, superior axis morphology was somewhat more common among HIV+ versus uninfected persons, but this did not reach statistical significance ($p = 0.092$).

Conclusions:

VEA is more common among HIV+ persons but this was attenuated after adjustment for CVD risk factors. Greater HIV viremia and immunosuppression are associated with greater odds of VEA. Compared with uninfected persons, HIV+ persons may more commonly have VEA originating from the left ventricular myocardium, suggesting abnormal myocardial substrate rather than idiopathic outflow tract arrhythmia.

Categories

1st year Fellow: Research

Program/Institution Name

Ohio State University Hospital

Virtual Visits in Cardiac Electrophysiology: Patient and Physician Preference

55

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Abstract

Background: Cardiologists have long utilized devices to follow patients with arrhythmias in order to guide management. Virtual visits have been adopted as one modality to follow-up established patients with arrhythmias. Factors contributing to patient and physician preferences with virtual visits are unknown. To our knowledge, there are no prior studies that have collected objective feedback from patients and physicians after virtual visits.

Objectives: To determine patient and physician experience with virtual visits in Cardiac

Electrophysiology.

Methods: We performed a prospective survey of patients and physicians who participated in a virtual visit in the Department of Cardiac Electrophysiology at the Cleveland Clinic from December, 2018 and July, 2019. All established patients in the Department of Cardiac Electrophysiology at the Cleveland Clinic who had a virtual visit were invited to partake in our survey. A constructed, standardized phone script and patient survey questionnaire of 15 questions was implemented for each patient. In addition, for each virtual visit encounter the cardiac electrophysiologist who performed the virtual visit was also invited to participate in a separate physician survey.

Results: 100 patient and physician virtual visit encounters were included. The average age of patients who participated in a virtual visit was 65 years old. 70% were male and 30% were female. The average distance patients participated in their virtual visit was 656 miles. Of the 100 patients who participated in a virtual visit, 64 elected to complete a survey, 10 patients declined, 17 patients were unable to be reached on follow-up, and 9 patients were not included due to technical difficulties. Of those who responded, 51 patients participated in their first virtual visit, 4 participated in their second virtual visit, and 8 participated in their third or more virtual visit. 38/64 (59.4%) of patients preferred a virtual visit for their next visit, 12/64 (18.8%) preferred an in office visit, 13/64 (20.3%) responded that their decision for a virtual or office visit depended on their specific needs, 1/64 (1.6%) did not have a preference. A total of 14 cardiac electrophysiologists participated in 100 virtual visits. 9/100 visits were not included due to technical error and inability to complete the virtual visit. Of the 91 virtual visits by physicians, 62/91(68.1%) preferred a virtual visit for their next visit, 7/91 (7.7%) preferred an in office visit, 10/91 (11.0%) responded that their decision for a virtual or office visit depended on the indication

for follow-up, 6/91 (6.6%) did not have a preference, and 6/91 (6.6%) did not indicate their preference for their next visit.

Conclusions: Both patients and physicians showed favorable responses to virtual visits, with a majority of patients and physicians preferring a virtual visit over an in-office visit for their next encounter. Factors such as convenience, cost, feasibility, and reason for follow-up were important determinants that affected both patient and physician preference.

Categories

3rd year Fellow: Research

Program/Institution Name

Cleveland Clinic Foundation