Infective endocarditis of unicuspid aortic valve complicated by mitral-aortic intervalvular fibrosa pseudoaneurysm

13

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Abstract

Introduction: Pseudo aneurysm of mitral aortic inter valvular fibrosa (PA-MIVF), is a rare entity in pediatric population. It entails an avascular region between the anterior mitral leaflet and non-coronary cusp of the aortic valve with potential to become a nidus for infection with potential to compress surrounding structures.

Clinical Presentation: An 11 year old male with Osteogenesis Imperfecta and known unicuspid aortic valve with moderate stenosis and regurgitation, and progressive ascending aortic dilatation presented with weight loss, night sweats and acute onset of intermittent fever. Clinical examination revealed an unchanged systolic murmur with new onset of tender hepatosplenomegaly. Blood cultures were positive for *Streptococcus Mutans* mandating initiation of broad-spectrum antibiotics. Due to clinical suspicion of infective endocarditis an echocardiogram was performed emergently. Transthoracic echocardiogram (TTE) revealed a medium-sized, 1.2 cm x 3.0 cm, irregular, hyperechoic, mobile vegetation on the left ventricular aspect of the thickened aortic leaflets with infiltration in the plane of P-MAIVF. To and fro flow was noted from the left ventricular outflow tract into the pseudo aneurysm with LV dilation.

Discussion: P-MAIVF is a rare but fatal complication often associated with native aortic valve endocarditis. Due to the rarity of its presentation, prompt diagnosis remains challenging, as diagnosis can be confounded by presence of perivalvar abscess and sinus of Valsalva aneurysms. However, with advancement in multimodality imaging timely diagnosis and intervention has improved recognition of this rare entity. The sensitivity of TTE for this diagnosis is limited (~ 40%), hence a low threshold exists for performing TEE (greater sensitivity 90-95%) particularly in susceptible patients for prompt surgical management minimizing risks of potentially fatal complications.

Conclusion: TTE was a useful asset in determining P-MAIVF extent and its relationship with adjacent structures. Based on these findings, our patient underwent emergent aortic valve and root replacement with a 22 mm homograft with coronary artery re-implantation in a timely fashion.

Categories

2nd year Fellow: Case

Program/Institution Name

Cleveland Clinic Foundation

Non-Bacterial Thrombotic Endocarditis from Primary Anti-Phospholipid Antibody Syndrome Leading to Symptomatic Mitral Stenosis

35

Michael Biersmith

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Type of submitter

Fellow in Training

Abstract

Introduction: Antiphospholipid antibody syndrome (APS) is a multisystem autoimmune disorder characterized by arterial and venous thromboembolic events associated with circulating antiphospholipid antibodies. In many cases, APS is secondary to other autoimmune conditions such as systemic lupus erythematosus and can result in various cardiac pathologies. Primary APS is less likely to manifest valvular heart disease, usually non-specific valve thickening and mitral regurgitation. Clinically significant mitral stenosis is rare.

Case Presentation:

60 year old female with a history of APS, chronic kidney disease, subdural hemorrhage and GI bleeding while on warfarin therapy presented with recurrent dyspnea and orthopnea. Given her prior bleeding issues and labile INRs, her warfarin had previously been switched to Apixaban 2.5 mg BID. Reduced dose was used given weight parameters and renal dysfunction. Previously extensive serological evaluation did not indicate a concurrent rheumatologic diagnosis. She had no history of rheumatic fever or recurrent childhood upper respiratory infections. In a 6 month period, she had been admitted on three occasions for acute decompensated heart failure. Initial TTE and TEE showed rheumatic appearing mitral valve with mild-moderate mitral valve stenosis. After her third admission, she was referred for mitral balloon valvuloplasty for suspected rheumatic mitral stenosis after her mitral valve gradient on TTE was 20 mmHg. Intraoperative TEE showed interval development of moderately severe mitral stenosis and large thrombus burden. Comparative TEE imaging was obtained under similar physiological parameters and degree of anemia (Figure 1). All culture data was negative. Balloon valvuloplasty was aborted given extensive thrombus burden and concern for high risk of thromboembolism. Similarly, surgical valve replacement was not offered given prohibitive bleeding and thrombotic risk.

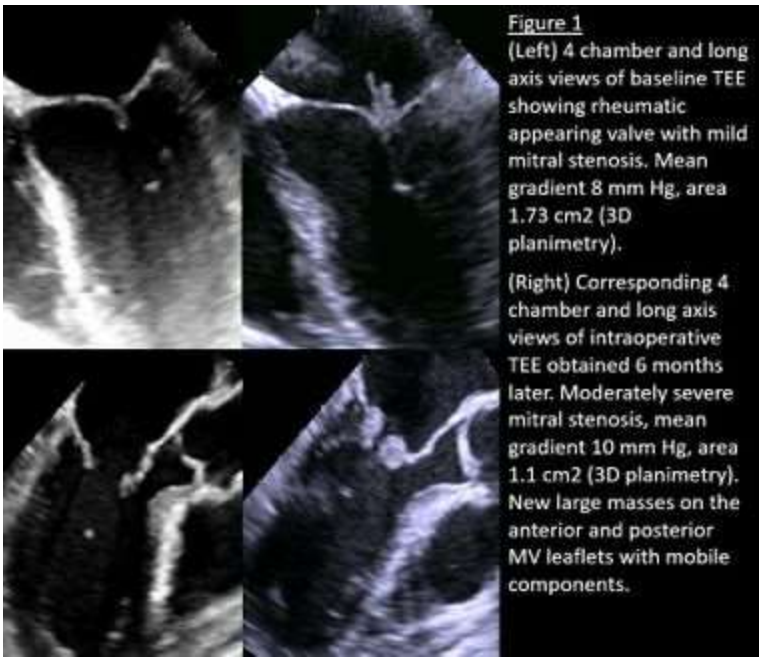

Figure 1
(Left) 4 chamber and long axis views of baseline TEE showing rheumatic appearing valve with mild mitral stenosis. Mean gradient 8 mm Hg, area 1.73 cm2 (3D planimetry).

(Right) Corresponding 4 chamber and long axis views of intraoperative TEE obtained 6 months later. Moderately severe mitral stenosis, mean gradient 10 mm Hg, area 1.1 cm2 (3D planimetry). New large masses on the anterior and posterior MV leaflets with mobile components.

Discussion: The leading diagnosis is non-bacterial thrombotic endocarditis from primary anti-phospholipid antibody syndrome leading to symptomatic mitral stenosis. The degree of stenosis rapidly progressed despite appropriate antiplatelet and anticoagulation, in line with other case report data. Though the use of Apixaban for treatment of APLA is off-label, this was felt necessary given patient's prior bleeding history with warfarin. After consultation with rheumatology, hematology, and cardiac surgery teams, a non-invasive approach including initiation of Plaquenil, Rituximab, and increasing Apixaban dosing was pursued in an attempt to control the underlying hematological drivers of her valvulopathy.

Conclusion: Primary APS is an uncommon hematologic syndrome and mitral stenosis is a rare manifestation of this disease. This case highlights how APS-related valve disease may be mistaken for rheumatic valve disease and is likely to progress despite appropriate antiplatelet and anticoagulation treatment. It further demonstrates the importance of a multidisciplinary team approach in the management of such patients, particularly given the high propensity for bleeding and thromboembolic risk.

Categories

2nd year Fellow: Case

Program/Institution Name

Ohio State University Hospital

Successful Treatment of Native Acquired Aortic Atresia using an Endovascular Covered Stent

76

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Type of submitter

Fellow in Training

Abstract

Introduction:

Severe aortic coarctation with progression to acquired atresia is a rare cause of hypertension in adults. The condition can lead to significant morbidity and early mortality if not treated early. Although endovascular intervention is the standard approach for native adult coarctation, the presence of aortic atresia, with need to recanalize the atretic segment, increases risk considerably and requires methodical approach and experienced operators to allow for successful outcomes.

Case:

A 25 year-old female presented with severe hypertension, refractory to treatment with amlodipine and labetalol. On examination, she had a systolic BP of 182 mmHg in the right arm, no palpable pulses of the bilateral lower extremities, and a continuous murmur at the bilateral infraclavicular areas. Computed tomography angiography for atypical chest pain showed short-segment atresia of the proximal descending aorta (Figure 1).

The patient underwent cardiac catheterization via femoral and radial approach. Simultaneous aortic angiograms above and below the obstruction demonstrated a 6 mm atretic segment. A right coronary catheter was positioned in the inferior limb and directed superiorly. The stiff end of a 0.014" wire was advanced through the atretic segment and snared. The atretic segment was ultimately crossed with a long sheath, and a 4.5 cm Covered CP Stent (NuMED) premounted on a 16 mm BIB balloon was implanted. After stent placement there was excellent flow through the recanalized aorta, no endoleak, and no vascular injury. There were no complications and she was discharged after two days. Since the procedure, she has remained normotensive off antihypertensive medications.

Discussion:

Our case demonstrates important points about caring for ACHD patients with complex and high risk anatomy. First, a high degree of suspicion for congenital etiologies should be sought when conditions present in young adults in the absence of risk factors. This patient likely had long-standing hypertension,

but despite her young age, lack of risk factors, and several pathognomonic clinical signs, a secondary etiology was never sought. Second, adult patients with congenital heart disease should be referred to dedicated ACHD programs when surgical or interventional procedures are indicated. Although, transcatheter endovascular stenting is considered first-line therapy for native severe coarctation, the optimal therapy for acquired atresia has not been demonstrated. This patient was initially referred to adult CT surgery for repair. Fortunately, her surgeon recognized the need for ACHD input and referred her to our program for evaluation. After extensive discussion at combined case conference, the transcatheter approach was recommended. The patient then went on to have a successful intervention and now is normotensive without the need for medication.

Conclusions:

Acquired aortic atresia is the severest form of aortic coarctation and invariably leads to upper extremity hypertension and significant cardiovascular complications if left untreated. Coarctation should be considered in all young patients with hypertension, especially those refractory to medications. Although both surgical and endovascular therapies carry significant risks, the endovascular approach using a covered stent can be performed safely and lead to excellent short and medium term outcomes.

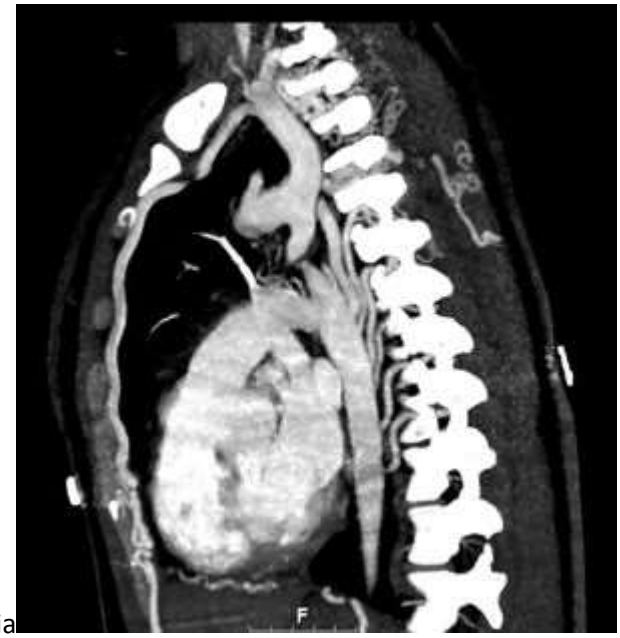

Figure 1: CTA revealing aortic atresia

Categories

2nd year Fellow: Case

Program/Institution Name

CWRU/Univ Hosps Cleveland Med Ctr/Rainbow Babies and Children's Hospital

Assessment of Exertional Ischemia in a Child with Anomalous Coronary Artery Origin and Ventricular Preexcitation using Nitrogen-13 Ammonia Positron Emission Tomography

46

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Abstract

Introduction: Anomalous origin of a coronary artery is a congenital anomaly that can result in sudden cardiac death (SCD). The 2018 ACC/AHA ACHD guidelines recommends that in patients with anomalous coronary artery origin, evidence of ischemia warrant surgical intervention. Myocardial perfusion imaging (MPI) during exercise stress testing (EST) can assess exertional ischemia. Conventionally, single-photon emission computed tomography (SPECT) has been used for this purpose. However, positron emission tomography (PET) MPI offers better spatial, temporal resolution and exposes the patient to less radiation. To our knowledge, the use of PET-MPI using ^{13}N-ammonia to evaluate ischemia in anomalous coronary artery origin has not been reported in pediatric literature.

Case: A 12-year-old previously healthy male was was found to have ventricular pre-excitation (VPE) incidentally on electrocardiogram during anesthesia monitoring during orthopedic surgery. He reported occasional exertional chest pain and shortness of breath. Echocardiogram showed anomalous origin of the right coronary artery (ARCA) from the left aortic sinus. CT angiography confirmed the diagnosis as well showed an intramural course with a slit-like ostium. A ^{13}N-ammonia PET-MPI scan was performed with EST which showed signs of ischemia involving lateral and inferior wall. During this EST, the patient also had a sudden and discrete loss of VPE suggesting low risk of rapid antegrade conduction. Patient eventually underwent surgical unroofing of the anomalous right coronary artery without complication.

Discussion: SCD in patients with ARCA is thought to be caused by ischemia due to increased myocardial demand along with compression of the ostium and intramural course during exertion. ST-segment and T-wave changes on ECG suggest ischemia during EST. However, in the presence of VPE, repolarization is altered and such findings are not a reliable assessment of ischemia. SPECT MPI can help assess for ischemia in this situation, but PET-MPI scan has been shown to have better resolution and higher diagnostic sensitivity for evaluation of ischemia in patients with coronary artery obstruction.

Conclusion: This case is the first known report of ^{13}N-ammonia PET-MPI scan to evaluate exertional ischemia in a pediatric patient with an anomalous coronary artery origin. Additionally, this case also illustrates that PET-MPI scan can be used in cases where ECG alone cannot be used due to VPE.

Categories

2nd year Fellow: Case

Program/Institution Name

CWRU/Univ Hosps Cleveland Med Ctr/Rainbow Babies and Children's Hospital

Acute coronary syndrome in a young woman with prior orthotopic heart transplant, a rare complication of cardiac allograft vasculopathy

36

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Type of submitter

Fellow in Training

Abstract

Introduction:

Cardiac allograft vasculopathy (CAV) is a significant cause of morbidity and mortality late after orthotopic heart transplantation (OHT); however, acute coronary syndrome is a rare complication of OHT. Here, we present a case of an inferior ST elevation myocardial infarction eleven years post cardiac transplant in the setting of significant CAV.

Case Presentation:

The patient is a seventeen-year-old female with a history of congenital dextro transposition of the great arteries that was repaired with arterial switch at two weeks of age. She required urgent OHT at six months of age due to complications and a second OHT at five years of age due to CAV. Serial left and right heart catheterizations with biopsies post-transplant were unremarkable. Eleven years post-transplant, the patient developed sudden onset left shoulder pain, nausea, and dizziness. On presentation, she was found to have inferior ST elevations on electrocardiogram. She underwent emergent coronary angiography that revealed an acute, complete occlusion of the distal right coronary artery. She underwent successful percutaneous intervention and was found to have moderate vasculopathy of the terminal right and posterior descending coronary arteries. Additionally, there was diffuse 30-40% stenosis and distal pruning of the left anterior descending and moderate to severe disease in the left circumflex consistent with CAV.

Discussion:

Cardiac allograft vasculopathy is the result of circumferential intimal thickening due to accelerated smooth muscle proliferation, accumulation of inflammatory cells, and deposition of lipids.[1]CAV is clinically relevant as it is responsible for one in eight deaths beyond one year post OHT.[2]Patients afflicted by CAV most often present with arrhythmia, sudden death, or heart failure; presentation as an acute coronary syndrome is atypical and considered extremely rare. It was interesting that our patient presented with acute anginal symptoms. Due to denervation of the allograph with transplantation, patients may not experience the classical symptoms associated with acute coronary syndrome.

Therefore, there must be a high index of suspicion for transplant patients presenting with vague symptoms, such as dyspnea or fatigue. Our patient did not possess any significant risk factors for atherosclerosis and screening angiography roughly six months prior to her infarct was unremarkable, further making this presentation unusual and unique. In fact, there are few case reports of similar presentations in the published literature.[3-6]The management of CAV includes further optimization of immunosuppression, such as the use of a mammalian target of rapamycin inhibitor (mTORi). mTORi's have been shown to be beneficial when initiated early transplant period.[1]Revascularization, either percutaneously or surgically, is an option for some patients with chronic CAV, however, is not universally associated with improved outcomes.[1,7]Retransplantation can also be considered in highly selected patients.[8]

Conclusions:

CAV is a significant cause of morbidity and mortality late after orthotopic heart transplantation. Acute coronary syndrome is a rare complication of CAV. Patients may not experience the classical symptoms associated with ACS due to allograft denervation. Therefore, there must be a high index of suspicion of ACS in patients with prior transplantation presenting with vague symptoms.

Categories

2nd year Fellow: Case

Program/Institution Name

Ohio State University Hospital

Exploring Cardiac Tumors: A rare case of a hemodynamically significant spindle cell lipoma arising from the pulmonary valve

10

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Type of submitter

Fellow in Training

Abstract

Title

Exploring Cardiac Tumors: A rare case of a hemodynamically significant spindle cell lipoma arising from the pulmonary valve

Introduction

We present a 52-year-old male who with a long-standing history of a pulmonary valve mass which ultimately required resection due to increased growth, symptoms, and new biventricular systolic dysfunction. Our objective is to explore the demographics, proposed etiologies, evaluation, and management for benign cardiac neoplasms with a focus on cardiac lipomas. Due to the extreme rarity of physiologically significant cardiac lipomas the optimal management strategy remains unknown.

Case Presentation

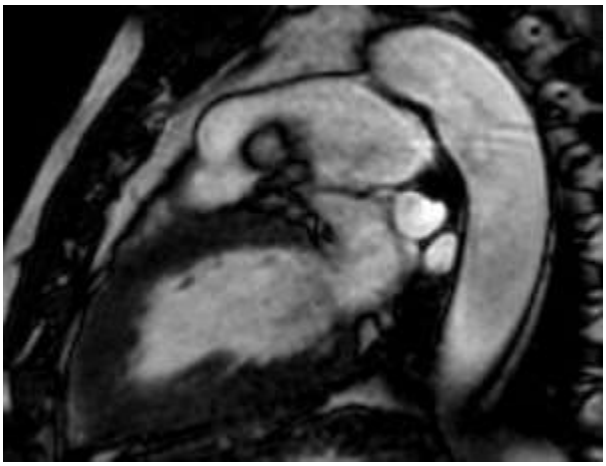
A 52-year-old male was admitted to the hospital with symptoms of light headedness after a routine echocardiogram revealed an enlarging, cystic, mobile mass attached to the pulmonic valve with new biventricular systolic dysfunction. The mass had enlarged from initial discovery a decade prior with prolapse through the annulus. Cardiac MRI further characterized it as a hypermobile 28x32x25 mm growth attached to the posterior leaflet. Cardiac function by MRI was depressed with both a right and left ventricular ejection fraction of 38% decreased from previously normal function on echocardiography. Given interval growth, biventricular systolic dysfunction, and new symptoms surgical intervention was deemed appropriate. The pulmonic valve and associated neoplasm were resected and replaced with a 29 mm St. Jude Trifecta bovine pericardial valve. Pathologic analysis was consistent with a spindle cell lipoma. The patient had an uneventful recovery and follow up cardiac MRI demonstrated improvement in both left and right ventricular systolic function to 56% and 52% respectively with resolution of symptoms.

Discussion

Cardiac tumors remain a very rare clinical entity with an autopsy frequency of 0.001% to 0.030%. The majority of primary cardiac neoplasms are benign. Around 50% are cardiac myxomas, the rest are papillary fibroelastomas, rhabdomyomas, and lipomas. Little is known about the etiology of cardiac lipomas with some studies suggesting a strong genetic component. Valvular involvement is exceedingly rare. Most cardiac lipomas are clinically silent and found incidentally on imaging or at autopsy. True lipomas of the heart account for less than 0.5% of excised tumors. Histologic findings are classic and similar to those for lipomas found elsewhere in the body. Prognosis is typically good with very rare cases associated with hemodynamic compromise or arrhythmia if there is involvement of valvular structures or the conduction system. Complete radiographic evaluation including echocardiography, magnetic resonance imaging, and computed tomography are warranted for non-invasive evaluation.

Conclusion

Benign tumors of the heart including lipomas, especially those with valvular involvement, represent an especially rare and challenging scenario. Every effort should be made to appropriately characterize the lesion in question with both echocardiography and cardiac MRI for a complete functional evaluation. This includes doppler interrogation of any valve in question as well as tissue characterization with contrast enhancement. Because these tumors represent such a small population optimal management remains unknown. In general, benign neoplasms of the heart should be observed conservatively unless implicated in symptoms or enlarge to a point of threatening complications. Treatment is generally surgical excision with preservation of normal cardiac structures.



Categories

2nd year Fellow: Case

Program/Institution Name

University of Toledo

Transcatheter Intervention of an Obstructed Common Right Pulmonary Vein in Repaired Scimitar Syndrome

58

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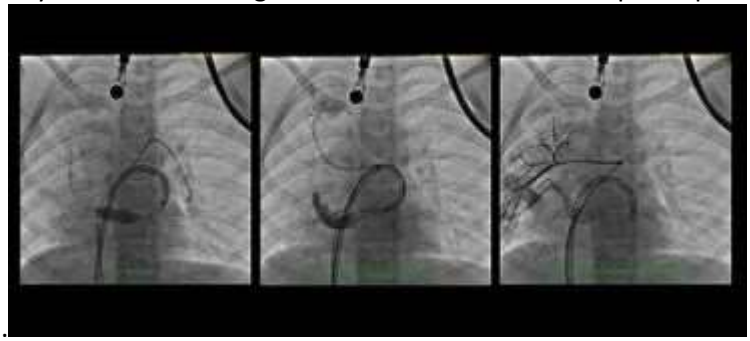
Type of submitter

Fellow in Training

Abstract

Scimitar syndrome is a rare congenital heart defect characterized by right-sided partial anomalous pulmonary venous return (PAPVR) to the inferior vena cava. The surgical repair for Scimitar syndrome and variants of PAPVR vary, but carry a risk of post-operative pulmonary venous obstruction. We report a 6-year-old patient with scimitar syndrome who underwent surgical repair via direct anastomosis of right pulmonary vein to the left atrium. Due to social reasons, she had poor follow up after surgical repair and was seen 14 months post-surgery. Transthoracic echocardiography revealed a mean gradient of 3-6 mmHg across the scimitar vein at the anastomosis site. Cardiac CT scan revealed the lesion measured 1.3 x 1 mm in diameter and the distal vein measured 10 x 10 mm. The patient was referred to interventional cardiology for hemodynamic assessment and treatment of her stenosis. The lesion was accessed using a steerable Agilis™ transseptal sheath (Abbott, Abbott Park, IL) which was curved 180 degrees within the left atrium. She was found to have a mean gradient of 9 mmHg across the lesion and then underwent balloon sizing to understand the compliance of the lesion. A 16 mm Mega LD stent (Medtronic, Dublin, Ireland) mounted on a 12 mm balloon was implanted across the lesion resulting in a residual gradient of 3 mmHg. The patient was placed on aspirin and at one month follow up had a mean gradient of 1mmHg on transthoracic echocardiogram assessment.

This unique case demonstrates the ability to address post-surgical pulmonary venous obstruction in a pediatric patient with Scimitar syndrome in the catheterization laboratory in lieu of the operating room. This procedure was facilitated by the use of a steerable sheath which allowed improved access to the lesion within a relatively small left atrium. To our knowledge, this is the first documented pediatric patient with scimitar syndrome to undergo transcatheter treatment of post-operative pulmonary venous obstruction.



Categories

2nd year Fellow: Case

Program/Institution Name

Nationwide Children's Hospital/Ohio State University

Right Heart Thrombus: An Under-recognized Cause of Sudden Death

31

Semaan Kobrossi, Phoo Nandar, Joseph Pietrolungo, Otto Costantini

Summa Health System

Type of submitter

Fellow in Training

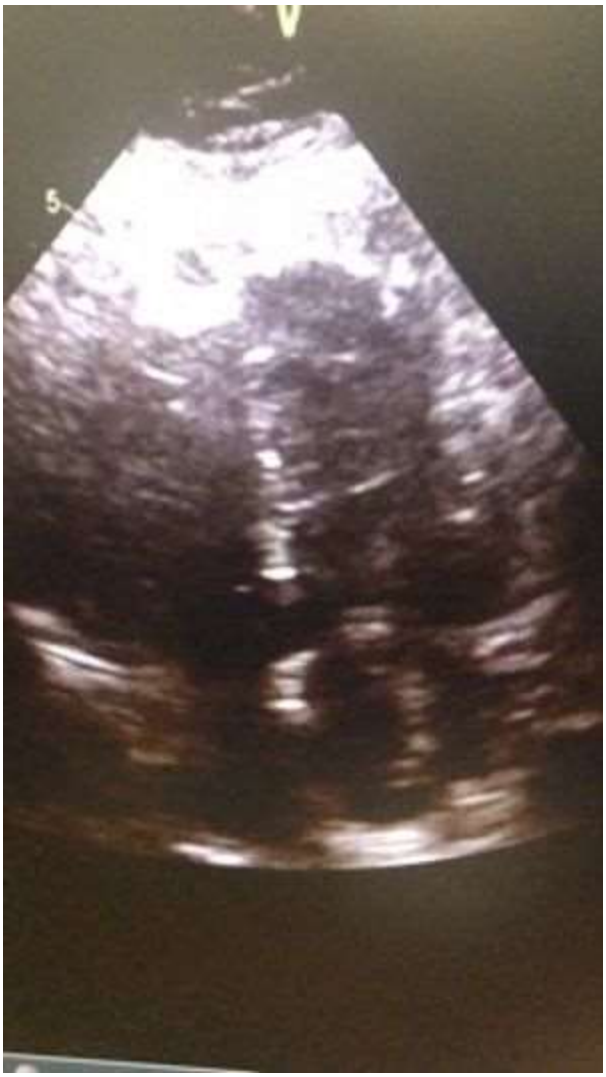
Abstract

Introduction

Right heart thrombi (RHT) are DVTs in transit, temporarily lodged in the RA or RV. They account for 4 % of pulmonary emboli and are associated with increased mortality of 28-40 %. Mortality rates in untreated patients are 80-100 %. Optimal management remains controversial because of the challenges of prospective randomized trials. We present a 60 year old woman who presented with a right heart thrombus and treatment options available.

Case presentation

60 year old woman with hypertension and hyperlipidemia presented to the Emergency Department with 2 days of chest pain and dyspnea on exertion. She had no leg swelling, pain or erythema. She had no risk factors for pulmonary emboli. Patient was chest pain free on presentation. Her blood pressure was 119/96 mmHg, HR 101 bpm and oxygen saturation 98% on room air. EKG showed sinus tachycardia and CXR was clear. Creatinine was elevated at 1.77, Troponin I was elevated at 0.19 and ProBNP was elevated at 13931. Patient was started on Heparin drip for a presumable acute coronary syndrome and admitted to the telemetry floor. She was assessed by the Cardiology fellow overnight. A bedside echocardiogram showed normal LV function, severely dilated RV, RV pressure and volume overload, 2+ TR and a large mobile mass in the RA prolapsing into the RV. Heparin drip was switched to high dose and patient was transferred to the CCU for closer monitoring. Cardiothoracic surgery was consulted and recommended further evaluation. Lower extremities Doppler showed extensive acute left leg DVT. Shortly after arriving to the CCU, she suffered a cardiac arrest. Resuscitative efforts including thrombolysis were not successful at restoring spontaneous circulation. Autopsy showed an extensive thrombus obstructing the RVOT.



Discussion

RHT can be fatal if not recognized in a timely fashion. Treatment options include anticoagulation with Heparin, thrombolysis (systemic or catheter-directed) and surgery. There are no randomized trials comparing the treatment options due to the challenges in randomizing such patients. A systemic review in 2002 by Rose, on 177 patients diagnosed with RHT showed mortality rates of 28.6, 23.8 and 11.3 % with anticoagulation, surgery, and thrombolysis respectively. In another review by Attapan, on 328 patients, the mortality associated with anticoagulation alone was significantly higher than surgical embolectomy or thrombolysis (37.1% vs 18.3% vs 13.7%, respectively). Another report based on a Spanish registry of 325 patients from 2000-2015 showed no difference in mortality between anticoagulation and reperfusion, the latter consisting of surgery or thrombolysis.

Conclusion

RHT is a potentially deadly condition that must be considered in patients with acute pulmonary emboli and diagnosed promptly. The presence of RHT significantly increases mortality. The management remains controversial and a multidisciplinary approach is needed. Some data suggest better outcomes with thrombolysis or surgery vs anticoagulation alone. While treatment should be individualized, aggressive strategies to remove or lyse the thrombus should be paramount.

Categories

2nd year Fellow: Case

Program/Institution Name

Summa Health System/NEOMED

Multiple Floating Thrombi in Aortic Arch Leading to Acute Stroke in a Young Adult: A Case Report and Review of Management

56

Mobasser Mahmood, Hemindermeet Singh, Sindhu Avula, Dilnoor Patti, Syed Sohail Ali

Mercy St Vincent Medical Center

Type of submitter

Fellow in Training

Abstract

Introduction

15% of acute cerebrovascular events occur in young adults with age less than 40 years. Trans-esophageal echocardiogram (TEE) is routinely performed to rule out any cardio-embolic source in patients with ischemic stroke. TEE has shown to significantly change management strategy in up to 16.7% of stroke cases. We report a case of recurrent stroke in a young female who was found to have multiple mobile thrombi in aortic arch on TEE. We also review literature for similar cases to highlight the management strategies.

Case Presentation

A 38-year-old female presented with one-week history of right upper and lower extremity paresthesia along with headache. Physical examination was unremarkable for any focal neurological deficits at the time of initial evaluation. She had pertinent history of acute stroke two years ago associated with non-occlusive left common carotid artery thrombus for which she was previously on anticoagulation with rivaroxaban. The anticoagulation, however, was stopped five months ago after repeat imaging revealed complete resolution of thrombus. Electrocardiogram showed normal sinus rhythm without any other significant abnormality. CT head showed no acute bleeding or infarct. MRI brain showed scattered infarcts in right cerebral hemisphere and a larger area of infarct in the left cerebellar hemisphere. CT angiography of head and neck showed multiple small nodular and linear pedunculated thrombi in distal arch of aorta (see Figure 2). TEE was then performed which confirmed two pedunculated and mobile echogenic masses, largest measuring 0.9 x 0.6 cm, in the distal aortic arch (see Figure 1). TEE did not show intracardiac source of embolism. Laboratory work up showed normal blood counts and normal renal function. Testing for thrombophilia was negative for Factor V and Prothrombin gene mutation and heterozygous positive for Methylenetetrahydrofolate reductase (MTHFR)-677T gene. She was also found to have elevated homocysteine levels. She was restarted on anticoagulation with rivaroxaban.

Discussion and Conclusion

Young patients with stroke should undergo detailed investigation to evaluate for hypercoagulable pathology and cardiovascular embolic sources. This should also include multimodality imaging including TEE in the selected patients. During TEE examination, a particular attention should be paid for evaluation of aortic source of thrombo-embolism. Our patient was heterozygous for MTHFR-66T gene which is associated with decreased activity of MTHFR by 35 % with elevated homocysteine levels. Treatment of floating aortic thrombus is controversial. Anticoagulation is suggested as primary modality by multiple authors who reported complete resolution of thrombus. Other option includes surgical thrombectomy. Our patient was treated with anticoagulation alone due to hypercoagulable state and small size of thrombi.

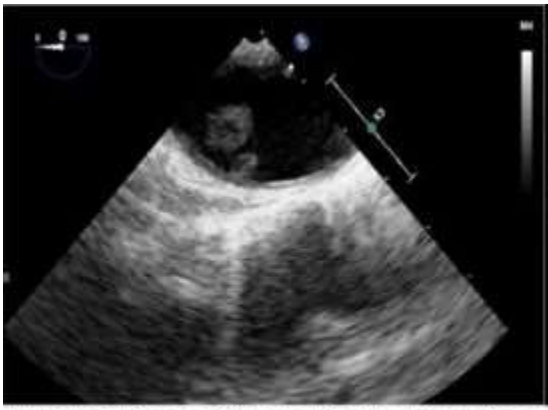


Figure 1: TEE image of distal aortic arch showing two mobile pedunculated masses suggestive of thrombi

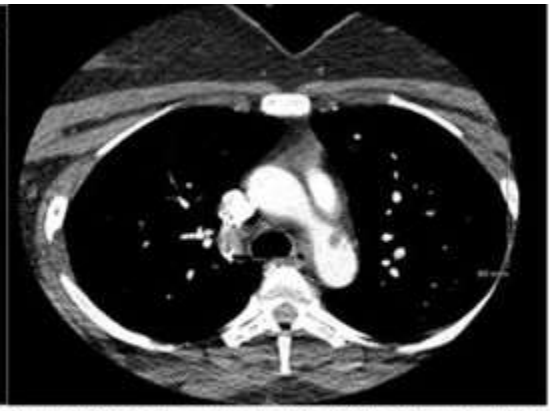


Figure 2 CT angiography showing filling defect in distal aortic arch suggestive of thrombus

Categories

2nd year Fellow: Case

Program/Institution Name

Mercy St Vincent Medical Center

Acute coronary syndrome in a patient with a single coronary artery arising from the right coronary sinus

75

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KETTERING MEDICAL CENTER

Type of submitter

Fellow in Training

Abstract

Introduction:

Coronary artery anomalies are typically discovered as coincidental findings during coronary angiography or post-mortem evaluation. A single coronary artery (SCA) is a rare anomaly of the coronaries. We describe a case of a 78-year-old male who presented with chest pain for several days. Cardiac catheterization showed the entire coronary system originating from the right coronary sinus and dividing into a right coronary artery (RCA) and left main coronary artery (LMCA). The left anterior descending (LAD) and left circumflex (LCx) took separate and unusual courses. A SCA poses diagnostic and therapeutic challenges especially in the setting of acute coronary syndrome (ACS). We present an interesting case of ACS in a patient with one of the rarest forms of a SCA which was successfully treated with angioplasty.

Case presentation:

A 78-year-old male with a history of hypertension, hyperlipidemia presented to the emergency department with the chief complaint of substernal chest pain for several days accompanied by dyspnea. Troponin was initially 0.112 ng/mL and trended up to 7.04 ng/mL. Electrocardiogram demonstrated normal sinus rhythm with non-specific ST-T changes. Cardiac catheterization demonstrated the absence of a left coronary system with the entire coronary system originating from the right sinus of Valsalva as a short common trunk dividing into a RCA and LMCA. The LMCA gave off a hypoplastic LAD coarsing posterior to the pulmonary artery and a LCx which coursed anterior to the pulmonary artery. There were lesions identified in the RCA which were the cause of the patient's presentation. Due to an unacceptably high surgical risk he underwent rotational atherectomy of the RCA followed by the placement of two drug eluting stents with an excellent angiographic and clinical result.

Discussion:

Coronary artery anomalies are rarely seen during routine coronary angiography with an incidence of 0.2-1.3% and 0.3% during autopsy. A SCA is an extremely rare anomaly of the coronaries, occurring in approximately 0.019 to 0.024% of the population. Patients can present with arrhythmias, syncope, ACS

or sudden death. In this case, the SCA originates from the right coronary sinus of Valsalva and gives rise to an RCA and a LMCA. The LMCA gave off an LAD which coursed between the pulmonary trunk and aorta in what is referred to as a malignant variant. The left circumflex coursed anteriorly to the pulmonary artery. The course of the anomalous arteries can have significant clinical and prognostic implications. An intervention in the setting of a SCA is technically difficult and potential complications can be catastrophic. In this case, the culprit vessel was the RCA which was successfully treated with rotational atherectomy and placement of two drug eluting stents.

Conclusion:

Single coronary arteries are extremely rare and pose significant diagnostic and therapeutic challenges to an operator. Treatment of ACS in such patients should be approached cautiously and surgery should be considered especially if the anomalous vessels take an interarterial course. Angioplasty can be considered if the surgical risk is considered to be high.

Categories

2nd year Fellow: Case

Program/Institution Name

Kettering Health Network

Kissing into third degree heart block

67

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[1]University of Cincinnati. [2]Robert Wood Johnson University Hospital

Type of submitter

Fellow in Training

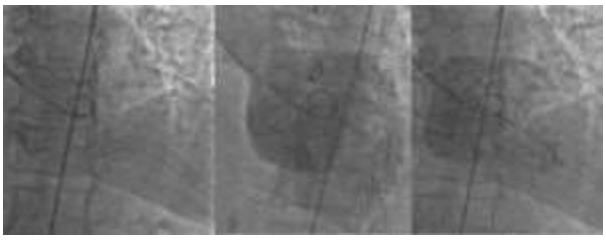
Abstract

Introduction

Chagas disease is the third most common parasitic disease encountered world-wide. Chagas cardiomyopathy is one of the leading causes of cardiovascular morbidity and mortality in Latin America. However, in the United States, Chagas cardiomyopathy is an exceptionally rare cause of heart failure and remains a challenging diagnosis. Clinical suspicion and a meticulous travel history is critical in making this diagnosis.

Case presentation

A 75-year-old Haitian woman with hypertension and epilepsy presented to the emergency department complaining of “feeling unwell”. She reported a two week history of non-productive cough and exertional dyspnea associated with lightheadedness. The remainder of her review of systems was non-contributory. Her only medication was over the counter guaifenesin as needed for cough. Her exam was unrevealing except for bradycardia. Electrocardiography revealed a complete heart block. Chest radiograph had no acute cardiopulmonary findings. Her cardiac biomarkers, thyroid function studies and electrolytes were within normal limits. She was admitted to Coronary Care Unit for intensive monitoring. Transthoracic echocardiography revealed akinesis and aneurysm of the basal inferoseptal and basal inferior walls, with no evidence of intracardiac thrombus. Coronary angiography revealed non-obstructive coronary artery disease. Left heart catheterization with ventriculography revealed a calcified, aneurysmal inferior wall. This with the patient’s country of origin, travel history, and left ventricular aneurysm in absence of coronary artery disease prompted a suspicion of Chagas disease. A thorough workup included titers for Chagas disease through the Center for Disease Control. IgG antibody for Trypanosoma cruzi was positive, suggesting Chagas cardiomyopathy. A permanent pacemaker was implanted. She remained hemodynamically stable throughout the remainder of her hospitalization and was discharged home with outpatient follow up.



Discussion

Chagas disease is caused by the protozoan Trypanosoma cruzi. Without treatment, Chagas disease can lead to long term complications, including Chagas cardiomyopathy. In Central and South America, Chagas cardiomyopathy is the leading cause of non-ischemic cardiomyopathy. Complications include heart failure, arrhythmias and sudden cardiac death. Though rarely seen in the United States, the prevalence is increasing and should be considered in patients with relevant risk factors. Chagas disease is growing in prevalence in non-endemic areas and now affects greater than 300,000 patients in the United States alone.

Conclusion

Recognition of Chagas cardiomyopathy remains challenging for practitioners in the United States due to it relatively low prevalence. However, the global burden of disease is high, particularly in the Central and South America. Additional research and treatment guidelines are critical for the early recognition and treatment of this devastating cardiovascular disease.

Categories

2nd year Fellow: Case

Program/Institution Name

University of Cincinnati

Three For One Deal: Uterine Embolization, Pulmonary Embolism with Cor Pulmonale and Ischemic Stroke

69

David Pratt, Mahyar Pourriahi

University of Cincinnati

Type of submitter

Fellow in Training

Abstract

Introduction

Uterine fibroid embolization (UFE) is considered a safe treatment option for patients with symptomatic leiomyomas. Here, we present a rare case of pulmonary embolism with cor pulmonale and ischemic stroke shortly after a successful uterine artery embolization.

Case Presentation

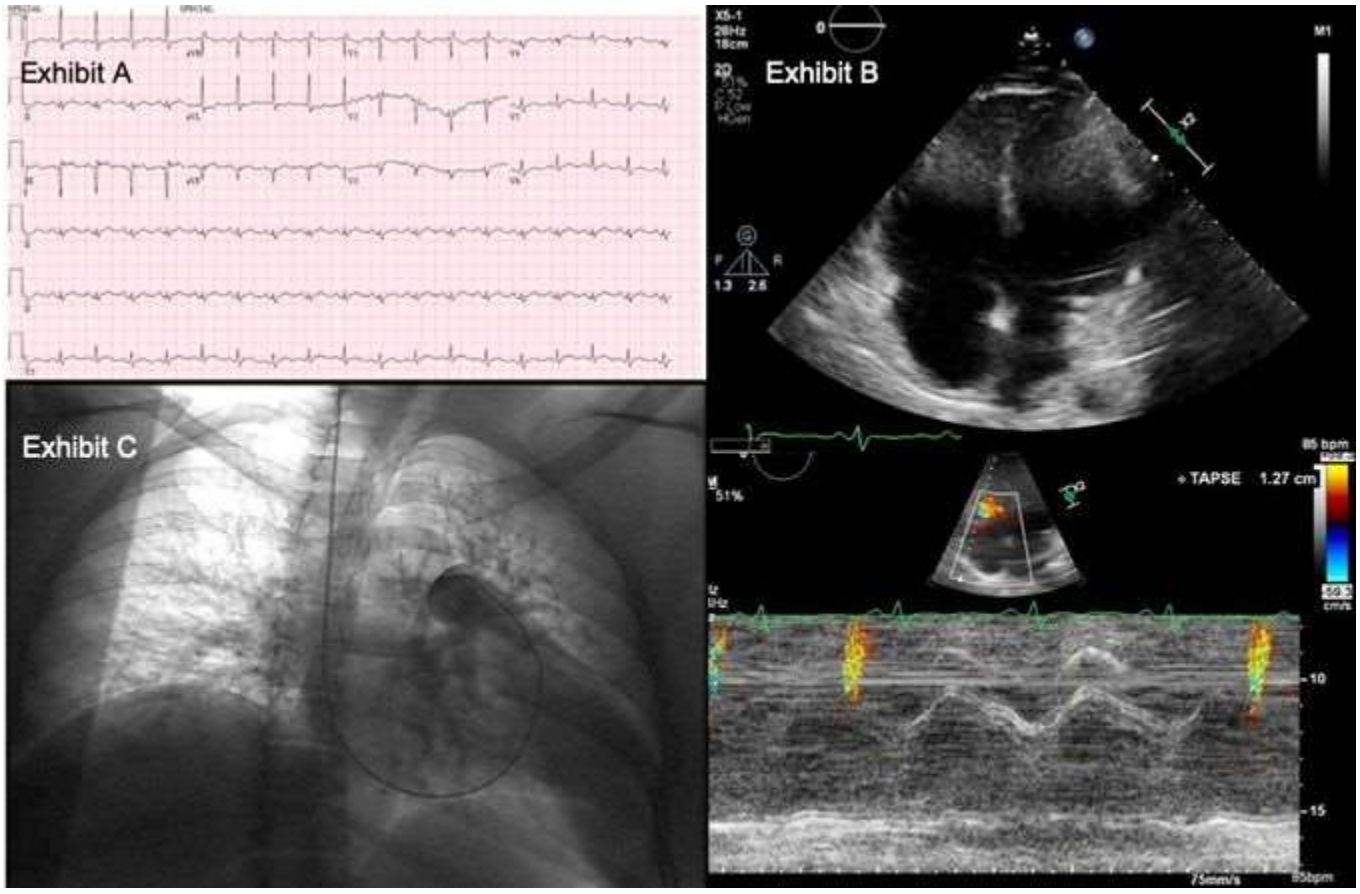
A 48-year-old female with a past medical history of asthma, hypertension and severe symptomatic leiomyomas presented to the outpatient surgical center for elective uterine fibroid embolization. Her procedure was successful with no immediate complications. While in the post-anesthesia care unit, she became acutely tachycardic, dyspneic, and hypoxic requiring a non-rebreather to maintain adequate oxygenation. She was subsequently admitted to the surgical intensive care unit. Computer tomography with contrast of the thorax was limited by motion artifact, but showed evidence of right heart strain. Electrocardiography showed sinus tachycardia with an incomplete right bundle branch block (Exhibit A). Transthoracic echocardiogram was significant for moderate pulmonary hypertension with moderate right ventricular enlargement and systolic dysfunction. She also had a positive bubble study at rest and with Valsalva (Exhibit B). Ventilation-perfusion imaging of the lungs showed numerous subsegmental perfusion defects bilaterally. Pulmonary angiography showed a filling defect in the right superior pulmonary lobar arteries (Exhibit C). Lower extremity doppler ultrasounds showed no evidence of a superficial or deep venous thrombus. She was initiated on a continuous heparin infusion. Two days after her presentation, she developed severe headache, left sided weakness and vision changes. Computed tomography of her head showed a large right parietal infarct with hemorrhagic conversion. Anticoagulation was discontinued and serial head imaging showed stability of her intracranial hemorrhage. Follow up ventilation-perfusion imaging showed resolution of her perfusion defects. Transthoracic echocardiography showed resolution of her right ventricular function and dilation.

Discussion

Thromboembolism is a rare complication from uterine fibroid embolization. There are only a few reported cases in the literature, and only one reported case of an ischemic stroke following uterine embolization. Two pathophysiologic mechanisms have been proposed. The first proposed mechanism is an acquired hypercoagulable state following UFE due to the induction of procoagulants from tissue injury. The second proposed mechanism described is from shunting of the embolization substrate across uterine arteriovenous malformations. In patients with intracardiac shunts, systemic embolization is possible. Post-mortem studies of patients who have suffered from thromboembolic event following UFE would be helpful to further characterize the nature of this phenomenon.

Conclusion

Ischemic stroke and pulmonary emboli with cor pulmonale are rare complications of uterine fibroid embolization. The mechanism and best treatment of thromboembolic events is not well understood. Further research is needed to better counsel our patient is needed. Additionally, preoperative risk stratification may be helpful to prevent this rare but devastating phenomenon.



Categories

2nd year Fellow: Case

Program/Institution Name

University of Cincinnati

Suck it out! Successful Removal of Infected Thrombus from the Superior Vena Cava Using the Percutaneous AngioVac System:

37

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Fellow in Training

Abstract

Background:

AngioVac (AngioDynamics Inc., Latham, NY, USA) is a vacuum-assisted thrombectomy device, which was approved in 2014 by FDA for removal of large thrombi and inappropriate material from body and its successful use for debulking large right-sided vegetations has been reported. We present a case of persistent bacteremia in a severely ill patient from infected thrombus, who underwent thrombus extraction with the use of AngioVac system.

Case:

A 77-year-old male patient, with recent history of necrotizing fasciitis of the right foot, complicated with acute renal failure required temporary hemodialysis with a tunneled catheter that was placed in the right internal jugular vein. He presented with fever and was found to be septic with methicillin-resistant staphylococcus aureus bacteremia. There was erythema and discharge noted from the skin surrounding the dialysis catheter, so the catheter was removed and was started on broad-spectrum antibiotics with Vancomycin and Piperacillin-Tazobactam. Extensive work up including CT chest/abdomen, Echocardiogram (TTE) and MRI spine and lower extremity were negative for potential source of bacteremia. He continued to be septic with positive blood cultures despite of appropriate antibiotics. Transesophageal echocardiogram (TEE) revealed a, 1.0 x 1.3 cm echo dense filling defect in the SVC suspicious for infected thrombus. Patient was deemed to be a high risk for surgical intervention, so he underwent extraction of the thrombus using AngioVac. Resolution of bacteremia was noted on follow up blood cultures and pathology report confirmed infected thrombus. The patient recovered well with intravenous antibiotics.

Discussion:

AngioVac is vacuum based device, which was approved for removal large thrombi and inappropriate material from body. There have been many case reports showing successful extraction of right-sided vegetations especially of infected thrombus associated with cardiac implantable devices using AngioVac device. Surgical extraction of septic emboli used to be the mainstay of therapy for persistent

bacteremia, but patients tend to be high risk given acuity and other co-morbidities. With the use of AngioVac suction device system, the patient had successful eradication of the source of infection and a good clinical outcome.

Conclusion:

This case highlights a promising role of the AngioVac system for successful aspiration of infected thrombus as an alternative to surgical intervention, especially in high risk, critically ill patients.

Categories

2nd year Fellow: Case

Program/Institution Name

Kettering Health Network

Correlation of exercise right heart catheterization hemodynamics in a patient with an implantable hemodynamic monitor.

16

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Type of submitter

Fellow in Training

Abstract

Introduction

In patients with chronic heart failure (HF), the use of the implantable pulmonary artery (PA) sensors, such as the CardioMEMS, are associated with a reduction in HF hospitalizations[i]. Previous studies have demonstrated excellent correlation between resting PA pressure sensors and right heart catheterization (RHC) pressures[ii]. However, it is not known whether implantable hemodynamic monitors correlate with RHC during exercise. We report a pilot case of an exercise RHC with simultaneous readings from a PA sensor.

Case Presentation

A 56 year old female with a history of hypertension, chronic obstructive pulmonary disease (COPD), tobacco use, and diastolic heart failure with previous PA monitor implant was evaluated for several months of worsening exertional dyspnea and exercise intolerance. Sensor-derived PA pressures readings demonstrated normal pressures at rest. Given her profound symptoms and concomitant respiratory and cardiac disease, an exercise RHC was performed using supine ergometer to assess for exercise induced diastolic dysfunction. Pressures from her PA sensor were obtained simultaneously with invasive RHC measurements both at rest and at peak exercise.

At rest, pulmonary artery pressure by RHC and cardioMEMS 23/10 mmHg (mean 16 mmHg) and 19/10 mmHg (mean 12 mmHg) respectively (Figure). At 20 Watts (55 RPM), the patient experienced exercise-limiting dyspnea. At peak exercise, pressures recorded by RHC and cardioMEMS were 36/10 mmHg (mean 27 mmHg) and 40/6 mmHg (mean 22 mmHg) respectively. [SE1] Based on her low filling pressures with exercise, exercise induced diastolic dysfunction was ruled out. Ultimately, COPD was thought to be responsible for her dyspnea and she was referred back to her pulmonologist to escalate her therapy.

Conclusion

In our patient, PA sensor measurements obtained during exercise appeared to be similar to those obtained by invasive RHC values. Further evaluation with an expanded patient cohort is needed to validate this observation. If true, the use of implantable PA pressures sensors could offer a non-invasive means of assessing exercise hemodynamics in patients with a device. This could also potentially be expanded to treadmill exercise testing, which is more physiologic than cycling ergometer and currently not possible with RHC. With emerging therapies for patients with exercise induced diastolic dysfunction and pulmonary hypertension, this information could potentially be used to target future therapeutics as well.

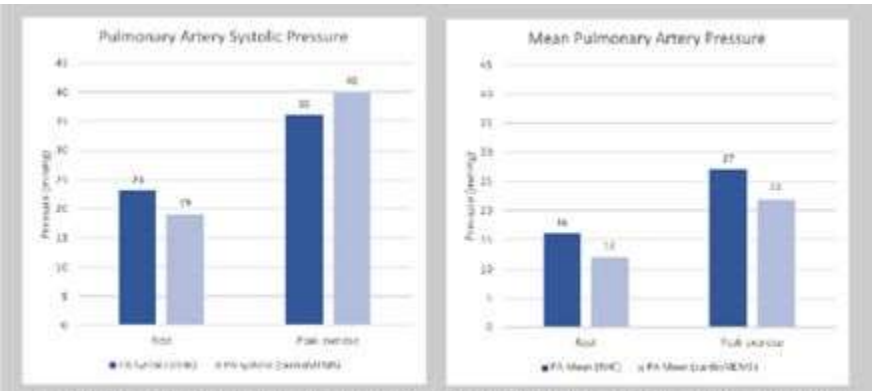

Figure: results of systolic (left panel) and mean pulmonary artery pressures (right panel) at rest and with exercise using right heart catheterization (dark blue) and with the implantable hemodynamic monitor (light blue).

[i] Givertz, Michael M *et al*. "Pulmonary Artery Pressure-Guided Management of Patients With Heart Failure and Reduced Ejection Fraction." *Journal of the American College of Cardiology* 70, no. 15 (October 10, 2017): 1875–86.

[ii] Abraham, William T. *et al*. "Safety and Accuracy of a Wireless Pulmonary Artery Pressure Monitoring System in Patients with Heart Failure." *American Heart Journal* 161, no. 3 (March 2011): 558–66.

Categories

2nd year Fellow: Case

Program/Institution Name

Ohio State University Hospital

A Marginal STEMi

65

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Type of submitter

Fellow in Training

Abstract

Introduction

The ECG remains one of the best bedside predictors of clinically significant coronary artery disease(CAD). ST elevation(STe) in V1-V5 usually indicates anteroseptal infarction caused by acute/subacute occlusion of the left anterior descending(LAD) or one of its branches. We describe a case of STe in the precordial leads in a patient with no involvement of the LAD or its branches.

Case presentation

A 67 year old male with history of LAD and diagonal drug eluting stents (DES) presented for staged right coronary artery(RCA) intervention. He was treated with 2 over lapping DES (24 and 38 mm). The second stent resulted in jailing of right marginal branch. Few hours later the patient had a ventricular fibrillation arrest. The patient achieved return of spontaneous circulation after 3 minutes of advanced cardiac life support.

Clinical decision making:

EKG post arrest showed STe in the precordial leads. Immediate angiography showed patent stents in LAD and RCA. Ostial Left main & LAD were also evaluated with IVUS without any significant disease. Balloon pump was inserted and the patient was admitted to the ICU . His EKG changes resolved and he was later discharged home in stable condition.

conclusion:

Isolated RV branch occlusion following angioplasty may be accompanied by STe in the precordial leads. These changes were replicated experimentally with balloon occlusion of the RV branch. Prominent anterior forces of RV infarction are postulated to be responsible for the STe in the precordial leads without the typical changes in the inferior leads. It is important that physicians recognize this entity and protect patients from unnecessary interventions.

Categories

2nd year Fellow: Case

Program/Institution Name

University of Cincinnati

Never Too Late: Stroke and Aortic Thrombosis Post Remote Splenectomy

23

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[1]Summa Health Heart and Vascular Institute. [2]Summa Health Department of Internal Medicine

Type of submitter

Fellow in Training

Abstract

Background:

Arterial thrombosis post splenectomy is underappreciated. Arterial events may present as an acute MI, stroke, pulmonary or peripheral artery embolism and can be devastating.

Case Presentation:

A 50-year old asplenic woman smoker with sleep apnea, Hodgkin lymphoma (in remission), stage 1B invasive ductal carcinoma of the left breast status post lumpectomy with adjuvant radiation and anti-estrogen therapy recently presented with headache, confusion and expressive aphasia. Multifocal left hemispheric strokes were found on brain MRI. A TEE showed a large protruding thrombus with mobile pieces in the distal aortic arch and descending aorta. An MRA and CTA confirmed this finding. Her symptoms resolved within four days. Hypercoagulability work up was negative. She was discharged on anticoagulation that was discontinued after a repeat TEE showed marked reduction of thrombus size at forty-five days post hospitalization. She was maintained on aspirin and clopidogrel thereafter. A repeat MRA was performed two months after. It showed no signs of inflammation, scar or fibrosis and almost complete resolution of the thrombus in the descending aorta.

Discussion:

Hypercoagulability in malignancy is certainly a differential. However, work up was unrevealing of any specific disorder other than persistent reactive thrombocytosis. Radiation injury to the aorta was ruled out after a thorough review of her recent adjuvant therapy dosimetry plan. She was on anastrozole even before her hospitalization, ruling out estrogen excess as cause. Her platelets have consistently been elevated since her remote splenectomy. Review of literature, albeit inconsistent, showed that there is a significant, long-lasting, increased risk of arterial thromboembolic events after splenectomy done for any indication. Risk is especially high for patients with an underlying hematologic disorder. Her continued heavy smoking likely contributed to her overall risk.

Conclusion:

Unexplained stroke-like symptoms in an asplenic patient should raise concern for arterial thromboembolism and other undiagnosed hematologic disorders. There is evidence that post splenectomy patients maintain a relative hypercoagulable state indefinitely due to multiple mechanisms. Although reactive thrombocytosis should not cause thrombosis by itself, empiric antiplatelet therapy in post-splenectomy patients with multiple risk factors must be considered.

Categories

2nd year Fellow: Case

Program/Institution Name

Summa Health System/NEOMED

Endocarditis: Classic presentation of a Classic Condition with a very Unclassical Etiology

78

Navdeep Tandon, Chris Tanayan, William Bauman

Summa

Type of submitter

Fellow in Training

Abstract

Introduction:

Intravenous drug abuse has unfortunately become a national crisis. Infective endocarditis incidence parallels the growth of this epidemic. Cases now present in a wide variety of ways, including with rare organisms that are more aggressive and damaging to the valves. Although there are many known classical physical exam findings associated with endocarditis, they are not commonly seen. This case presents a patient who presented with multiple classical physical exam findings of endocarditis, however, whose sonographic images and bacterial etiology are extremely uncommon.

Case:

A 28-year-old woman intravenous drug abuser with hepatitis C presented with a three-week history of fevers, fatigue, palpitations, arthralagias and a change in personality. On presentation she was found to be in septic shock and was confused. She had classic physical exam findings of infective endocarditis and a swollen right knee and ankle. She was anemic, thrombocytopenia and had an elevated INR. An echocardiogram showed a 2.4 cm x 1.7 cm mobile vegetation on the posterior leaflet of the mitral valve with moderate regurgitation. Her blood culture and urine culture grew Serratia Marcescens and was then started on appropriate antibiotics. A MRI was ordered due to her personality changes and showed bilateral infarcts in the parietal and the right thalamus and frontal lobe. A right knee arthrotomy was done which also grew Serratia and an MRI of the right ankle showed evidence of osteomyelitis. She developed acute respiratory failure due to worsening mitral valve regurgitation prompting urgent valve replacement. She was eventually discharged to a facility for rehab and long term antibiotics.

Discussion:

Infective endocarditis typically involves the tricuspid valve and is most commonly caused by Staphylococcus Aureus. Serratia Marcescens is an aerobic gram negative bacillus and was first reported to cause endocarditis in 1951. Since 1951 there has only been several additional cases of Serratia induced endocarditis reported. The International Collaboration on Endocarditis prospective cohort, which consisted of almost 2800 patients over a 5 year period showed that Serratia to be the cause of

endocarditis in only 0.14% of that cohort. Serratia has a propensity to infect the mitral valve compared to the right sided valves and has been shown to cause a larger sized vegetation, resulting in greater valve destruction and more frequent embolization than other bacteria. Thus, a higher morbidity and mortality is associated with this bacteria necessitating a more prompt intervention and aggressive treatment.

Conclusion:

This case demonstrates a classic infective endocarditis presentation with a rare organism and its more aggressive progression. If Serratia marcescens is cultured, one should consider an earlier surgical treatment and forego conservative measures.

Categories

2nd year Fellow: Case

Program/Institution Name

Summa Health System/NEOMED