Coronary Artery Aneurysm Caused by a Stent Fracture

47

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Type of submitter

Fellow in Training

Abstract

Introduction:

Coronary stent fracture (SF) is a rare complication of percutaneous coronary intervention (PCI). The most common adverse event caused by SF is in-stent restenosis due to neointimal overgrowth caused by an unexpected gap between stent struts. A coronary artery aneurysm (CAA) is a rare complication of SF and may lead to lethal events such as acute coronary syndrome or rupture of the CAA further leading to cardiac tamponade. We report an interesting case of a CAA caused by a SF that was treated medically.

Case Presentation:

A 69-year-old man presented with recurrent angina and borderline troponins consistent with an acute coronary syndrome. He had a history of paroxysmal atrial fibrillation and known coronary artery disease with prior PCI of the proximal, mid, and distal RCA several years ago. He then had an inferior STEMI 4 months prior due to severe mid RCA in-stent restenosis which was re-stented with a 3.5x28 mm Everolimus drug-eluting stent (DES). On his current presentation, coronary angiography revealed widely patent RCA stents, but there was angiographic and IVUS evidence of stent fracture of the recently placed stent at the mid-portion of the mid RCA DES, with evidence of a new focal coronary aneurysm at that site. By IVUS, the aneurysm measured 6x9 mm with no clear thrombus, and there was no significant flow limitation by iFR evaluation (iFR 0.94). The left coronary system had only mild disease, the LV systolic function was normal, and the left heart filling pressures were only mildly elevated. As there was no evidence of flow limitation through the RCA aneurysm, the decision was made to treat medically. Given the increased risk of thrombosis and distal embolization from coronary aneurysms, anti-coagulant therapy was recommended indefinitely in addition to his chronic anti-platelet therapy.

Discussion:

SF was first reported in 2002. The incidence varies between 0.5% and 18.6% and has declined with improvements in DES technology and design. However, SF sometimes occurs even with the second-generation DESs. The risk factors include overlapped stents, tortuous or calcified lesions, stenting in the RCA, and aggressive post-dilatation. CAA is defined as artery dilatation exceeding 50% of the reference vessel diameter. CAA has multiple etiologies and the prevalence ranges from 0.3% to 4.5%. PCI-related CAAs are rare and can be caused by mechanical factors including residual dissection and arterial wall injury due to coronary guidewire insertion into the false lumen, excessive balloon or stent dilation, and SFs. It has also been theorized that some PCI-related CAAs may actually represent pseudoaneurysms. CAA complications, such as thrombosis, distal embolization, ACS, and rupture, can be fatal. The management strategy for CAAs is controversial. Surgical approaches, percutaneous covered-stent implantation, coil embolization, and medications such as antiplatelet and anticoagulant therapy have all been utilized.

Conclusion:

CAA caused by SF is a rare complication of PCI. There are no established guidelines for the management of CAAs, particularly in the setting of SF. The management strategy remains controversial and pragmatic with limited data for guidance.

Categories

3rd year Fellow: Case

Program/Institution Name

Summa Health System/NEOMED

Anomalous RCA: Red Herring, or Culprit?

71

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Abstract

Background:

Despite coronary anomalies being among the most common congenital cardiovascular abnormalities, sufficient data is lacking regarding the implications of various anomalies and efficacy of treatment. This is especially true of RCA anomalies.

Case:

48 year old female presented with near syncope, palpitations, and worsening chest tightness. EKG was non-acute. Echo revealed akinesias of the mid-inferosteptal, mid-apical inferior, apical septal, and apical myocardium, and ejection fraction of 20%. Coronary angio demonstrated a 75% mid RCA lesion, with dominant RCA coursing between the aorta and pulmonary artery.

Discussion:

CTA confirmed an anomalous RCA. Per 2018 ACHD Guidelines, anomalous coronaries from the left or right sinus with symptoms or evidence of ischemia attributable to the anomalous coronary is a class I indication for surgery. Of question was whether patient's cardiomyopathy could be attributed to RCA lesion, anomalous RCA, or alternate etiology. Intravascular ultrasound of the RCA was performed and demonstrated an RCA originating from the left coronary cusp with an oval shaped ostium and no significant compression. Circumferential 75% stenosis, believed to be the culprit, was stented. Patient was discharged on optimal therapy with plan for echo in 3 months.

Conclusion:

This case highlights potential complexities in diagnosing/managing anomalous coronaries, in light of limited data, and highlights the importance of managing these patients on a case to case basis.

Categories

3rd year Fellow: Case

Program/Institution Name

Kettering Health Network

Atypical Hemolytic Uremic Syndrome: A Rare Cause of Reversible Cardiomyopathy

17

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Type of submitter

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Abstract

Introduction: Atypical hemolytic–uremic syndrome (aHUS) is a life-threatening, genetic disease of complement-mediated thrombotic microangiopathy that typically presents as anemia, thrombocytopenia, and renal failure. Cardiomyopathy is seen in up to 10% of aHUS cases, but the etiology is not well understood.

Case: A 63-year-old male with a history of renal adenocarcinoma status post nephrectomy, paroxysmal atrial flutter, obstructive sleep apnea and scleroderma recently was diagnosed with aHUS by renal biopsy after presentation with acute renal failure requiring hemodialysis. He was started on therapy with complement inhibitor, eculizumab. Six weeks after diagnosis, he presented with progressive dyspnea on exertion and chest pain. Initial evaluation included an ECG without acute ischemic changes, CT scan without evidence of pulmonary embolism, and chest x-ray with pleural effusions. Troponin level peaked at 0.21 ng/ml. An echocardiogram demonstrated an acute drop in left ventricular ejection fraction to 20-25% (prior 60-65%) with global hypokinesis. Left heart catheterization showed moderate, non-obstructive coronary artery disease. Cardiac MRI demonstrated diffuse myocardial edema. Cardiac biopsy revealed an arteriole with obliterative changes, a few possible fragmented red blood cells suggestive of thrombotic microangiopathy, and showed no evidence of immune complex deposition or myocarditis. Electron microscopy demonstrated myocyte vacuolization. For heart failure, he was started on carvedilol and lisinopril. Lisinopril was discontinued within one week due to symptomatic hypotension. The patient was continued on eculizumab. On repeat echocardiogram three months later, patient had complete recovery of his ejection fraction (60-65%) with no regional wall motion abnormalities.

Conclusion: In this case report, we describe complete recovery of aHUS-associated heart failure with eculizumab therapy and for the first time that the etiology of aHUS-associated heart failure is likely due to acute thrombotic microangiopathy as demonstrated by cardiac biopsy.

Categories

3rd year Fellow: Case

Program/Institution Name

Ohio State University Hospital

Left Ventricular Hypertrophy with progressive conduction disease and worsening renal function: Make the Diagnosis!

11

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Abstract

Introduction

Fabry disease is an X-chromosome-linked lysosomal storage disease which is caused by a deficiency or lack of enzyme α-galactosidase A (a-Gal A) leading to systemic accumulation of globotriaosylceramide (Gb3). This lysosomal storage disease is exceedingly rare and progression of the disease commonly occurs in adulthood. The systemic accumulation of Gb3 leads to eventual cardiovascular dysfunction, renal failure, and neuropathy. We present a case of Fabry Disease which was diagnosed after the patient was found to have classic ECG findings and progressive conduction abnormalities.

Case Presentation

A 68 year old male with a medical history significant for supraventricular tachycardia (SVT), hypertension, non-obstructive coronary artery disease, hyperlipidemia and chronic kidney disease presents to the cardiology clinic for further evaluation of his abnormal ECG and lightheadedness. Patient stated that he was initiated on beta blocker therapy due to his history of SVT which he had not been able to tolerate due to progressive lightheadedness. In regards to his abnormal ECG, the patient stated that he had an abnormal ECG in 1995 (Figure 1) which revealed a short PR interval and marked left ventricular hypertrophy (LVH). He underwent a subsequent ECG in his primary care doctor's office which revealed a new first degree AV Block with a wide right bundle branch block and continued LVH (Figure 2). Upon evaluation of his labwork, the patient was also found to have progressively worsening renal function with his BUN now up to 74 and Creatinine up to 4.59. Due to the concern for infiltrative cardiomyopathy given the progressive conduction disease coinciding with LVH on ECG and worsening renal, echocardiogram was ordered which revealed severely increased LV wall thickness , moderately increased RV wall thickness, severely dilated atria, and apical sparing pattern on strain analysis. Given all of these echocardiographic findings, patient was sent to his Nephrologist for kidney biopsy which revealed podocyte inclusion bodies consistent with Fabry Disease. Patient was then started on replacement enzyme therapy with a-Gal A supplementation with Fabrazyme (agalsidase beta). Patient was eventually initiated on hemodialysis due to progression to end stage renal disease and has done well with his replacement enzyme therapy.

Discussion

The most common driver of mortality in patients with Fabry Disease is cardiomyopathy as the disease has a propensity to infiltrate into the cardiomyocytes and cardiac conduction tissue causing irreversible cardiac damage. Common ECG findings associated with the disease include short p wave duration, PQ interval prolongation, repolarization abnormalities, and AV Block.

Conclusion

We present an interesting case of Fabry Disease in which the patient was initially found to have progressive conduction abnormalities and upon further evaluation was found to have significant LVH with worsening renal dysfunction. These findings led to the physician team to consider an infiltrative cardiomyopathy and this case highlights the utility of the ECG in the diagnosis of Fabry Disease.

Categories

3rd year Fellow: Case

Program/Institution Name

Summa Health System/NEOMED

Percutaneous Pulmonary Valve Implantation (PPVI) of the Melody™ Transcatheter Pulmonary Valve Using the Gore DrySeal Sheath: A Modified Technique

72

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Abstract

Introduction: Patients with right ventricular (RV) outflow tract obstruction often require surgical correction. This frequently includes placement of an RV to pulmonary artery (PA) conduit or augmentation of the outflow tract. Percutaneous pulmonary valve implantation (PPVI) is an attractive option for patients who have failure of these conduits, allowing them to forgo or postpone surgical valve replacement. The Melody Transcatheter Pulmonary Valve (TPV) (Medtronic, Dublin, Ireland) is typically implanted using the Ensemble Delivery System (Medtronic, Dublin, Ireland) with good results, though alternative delivery methods have been described in select cases.

Case presentation: An 11 year-old, 32.5kg male with double outlet RV, uncommitted ventricular septal defect, malposition of the great arteries, and pulmonary valve stenosis had undergone initial neonatal palliation with a modified Blalock-Taussig shunt. At 20 months old, he underwent complete surgical repair via Rastelli operation with an RV to PA conduit. Four years later, he required a conduit replacement with an 18 mm Hancock valved conduit. Over the next 5 years, he developed severe RV hypertension from moderate RV to PA conduit stenosis and was referred to the catheterization laboratory for intervention. Baseline hemodynamics revealed the RV pressures that were 73% of systemic arterial pressure secondary to a peak systolic gradient (PSG) of 43 mmHg across the proximal conduit. Given the patient's small size, the intervention was performed from the right internal jugular vein. The proximal conduit was stented using two Palmaz 3110 stents (Cordis, Hialeah, FL) dilated to a diameter of 18 mm. The right internal jugular would not accept the Ensemble II Delivery System nor a 22 Fr Gore Dry-Seal sheath (Gore, Newark, DE). The Melody TPV was crimped onto an 18 mm BiB balloon (NuMed Inc., Cross Roads, TX) and front loaded within an 18 Fr Gore Dry-Seal sheath. The sheath and valve were advanced through the venotomy site over the interventional wire to the conduit where it was successfully deployed. The resultant RV pressure was 50% of systemic arterial pressure with no valvar or perivalvar regurgitation by intracardiac echocardiography.

Discussion: PPVI generally utilizes the implantation system designed for the specific transcatheter valve. It has been shown that an internal jugular venous approach can be beneficial in smaller/younger patients and those with higher RV pressures. In this case, the Ensemble system proved to be too large for the internal jugular vein and the Melody TPV was successfully delivered via a modified technique

using an 18 Fr Gore Dry-Seal sheath. This modified approach may be useful in smaller patients who may not be able to accommodate traditional TPV delivery systems.

Conclusion: The Gore Dry-Seal sheath may be used as a modified delivery system for TPV implantation in small patients.

Categories

3rd year Fellow: Case

Program/Institution Name

Nationwide Children's Hospital/Ohio State University

Platypnea Orthodeoxia Syndrome: A Story of Weight Gain Unmasking Congenital Heart Disease

8

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Abstract

Introduction: Platypnea orthodeoxia syndrome (POS) describes a rare condition of dyspnea and hypoxemia induced by upright posture that resolves when supine. The majority of cases are caused by right to left shunting through a patent foramen ovale (PFO) or atrial septal defect (ASD). Two conditions must coexist for this syndrome to occur: an interatrial communication and a functional component that redirects shunt flow in the upright position.

Case: A 65 year old lady with recent 70-pound weight gain was transferred to our institution with severe hypoxemia not corrected with supplemental oxygen. Outside hospital work-up was unrevealing for an etiology for her dyspnea and hypoxemia. CT chest was negative for pulmonary embolism or lung parenchymal disease. Liver ultrasound showed no cirrhosis. Transthoracic echocardiogram was unrevealing due to poor windows. Right heart catheterization showed normal right atrial and pulmonary pressures and no evidence of intracardiac shunting while supine. Cardiac MRI showed normal pulmonary vein anatomy and Qp:Qs of 1.16 while supine.

Discussion: Upon arrival at our institution, physical examination of our patient demonstrated abrupt hypoxia and dyspnea while standing and sitting up, both relieved by recumbency, pathognomonic for platypnea-orthodeoxia syndrome. Supine transesophageal echocardiogram (TEE) showed large atrial septal aneurysm (ASA) and secundum ASD with small bidirectional shunt. TEE was then performed in the upright position. Sitting upright, she desaturated to 80% and the ASA bulged continuously to the left, resulting in a marked right to left shunt with saline contrast (see attached Figure). She underwent successful transcatheter ASD closure with an 18 mm Amplatzer septal occluder device with complete resolution of symptoms and hypoxemia.

Conclusion: Our patient developed platypnea-orthodeoxia syndrome from a previously unrecognized ASD. Significant weight gain caused diaphragmatic compression of the right atrium, resulting in marked leftward displacement of the atrial septum when upright. This promotes streaming of blood from the IVC directly across the septal defect into the left atrium. POS is a rare cause of dyspnea requiring a high index of suspicion to recognize, as symptoms occur opposite to heart failure and other cardiopulmonary conditions. POS should be considered in patients with positional dyspnea and refractory hypoxemia,

especially in the absence of pulmonary parenchymal disease. Due to the positional nature of POS, it is imperative to document saturation levels and to evaluate shunting in both supine and seated positions. Treatment is percutaneous or surgical closure of atrial septal defect, which causes quick relief of symptoms.

Fig 1. Supine TEE showing large, redundant atrial septal aneurysm

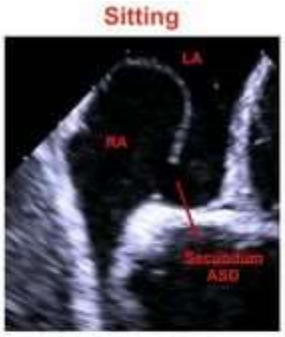

Fig 2. Upright TEE: ASA bulges to left, 7 mm secundum ASD visible

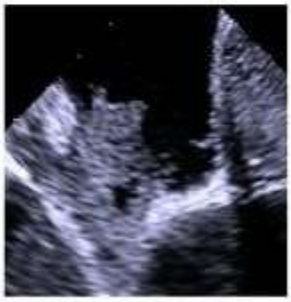

Fig 3. Supine TEE with saline contrast showing ASA with small right to left shunt

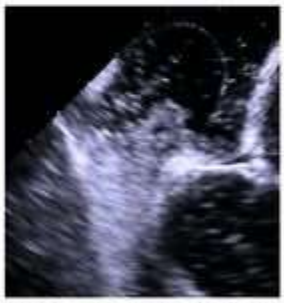

Fig 4. Upright TEE with saline contrast shows more pronounced right to left shunt

Categories

3rd year Fellow: Case

Program/Institution Name

Ohio State University Hospital

When the Gold Standard Fails: An Interesting Case of CPVT

22

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Cleveland Clinic Children's

Type of submitter

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Abstract

Introduction:

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare ion channelopathy that results in sudden death, mostly in adolescents. The gold standard of diagnosis is an exercise treadmill stress test that elicits the hallmark rhythm of CPVT: Bidirectional ventricular tachycardia. Polymorphic ventricular ectopy can also be seen. Two separate gene mutations, specifically in *RYR2* and *CASQ2,* have been demonstrated to cause CPVT with incomplete penetrance. We present a case of a de novo mutation of CPVT in an adolescent that sustained multiple cardiac arrests while playing sports despite having multiple exercise stress tests that failed to demonstrate ventricular ectopy.

Case presentation:

Our patient was a 15-year-old male who suddenly collapsed while playing lacrosse. He required CPR per bystanders at the scene. His initial echocardiogram and ECG were negative for abnormalities as was his exercise treadmill stress test. His family history was negative for dysrhythmias and channelopathies. No definitive cause of his syncope was diagnosed during this encounter. He continued to play competitive sports without recurrence. Three years from the initial event, he sustained a cardiac arrest while swimming that required CPR and an AED shock for ventricular fibrillation. On hospital admission, his MRI was suggestive for subtle findings of myopericarditis. Again, his exercise stress test was negative for dysrhythmia. He was started on nadolol and underwent implanted defibrillator placement for multiple episodes of cardiac arrest. Genetic testing for channelopathies was performed which demonstrated that he was heterozygous for RYR2 c.1258C>T (p.Arg420Trp), exon 14, a known pathological variant of CPVT. His first degree relatives were found to be genotype and phenotype negative for any RYR2 mutations. He has since self-restricted himself from intense exercise and competitive sports, though remains engaged in regular exercise, without recurrence of cardiac arrest or ICD shocks. This year, after an extensive discussion with the patient and his family, he opted to have the generator of his ICD replaced.

Conclusion:

The gold standard of diagnosis for CPVT is the exercise treadmill stress test which elicits symptoms in 80-100% of cases, though there is a small subset of patients that do not demonstrate symptoms on exercise stress testing. Genetic analysis in these scenarios, particularly when a deleterious variant in a CPVT gene is found, can be critical for proper diagnosis and treatment of this channelopathy.

Categories

3rd year Fellow: Case

Program/Institution Name

Cleveland Clinic Foundation

A Case of Takotsubo Cardiomyopathy after Cardioversion for Atrial Fibrillation

52

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Type of submitter

Fellow in Training

Abstract

Introduction: Takotsubo cardiomyopathy is an acute transient form of nonischemic cardiomyopathy that occurs after an intense emotional or physical stress. We report a previously unrecognized, novel cause of stress induced cardiomyopathy after transesophageal guided direct current cardioversion for atrial fibrillation.

Case: An 83-year-old woman presented to the office with shortness of breath and fatigue and was found to be in persistent atrial tachycardia with a 2:1 rapid ventricular response at approximately 120 bpm. Her ECG showed a QTc of approximately 450 ms and no ST/T wave abnormalities. She has a pertinent cardiac history significant for paroxysmal non-valvular atrial fibrillation, coronary artery disease s/p percutaneous coronary interventions to the right coronary artery and left circumflex arteries. Because she was not fully anticoagulated, she underwent transesophageal (TEE) guided direct current cardioversion. At the time of TEE, her left ventricular ejection fraction (LVEF) was measured at 45-50%. She was discharged home on anticoagulation and a beta blocker, but no antiarrhythmic drugs. One day after the procedure she presented to the emergency department with acutely worsening shortness of breath and lower extremity edema. She was admitted to the hospital for treatment of acute decompensated heart failure. On presentation, her electrocardiogram (ECG) showed sinus rhythm with deep, diffuse T wave inversions with a markedly prolonged QTc of 600 ms. On telemetry, she developed non-sustained, 5-10 beat runs of polymorphic wide complex tachycardia consistent with Torsades de Pointes ventricular tachycardia. A transthoracic echocardiogram demonstrated an LVEF of 20%. She underwent a left heart catheterization that demonstrated patent stents and no coronary artery disease that would require revascularization. It was unchanged from a prior left heart catheterization in 2018. The left ventriculogram showed evidence of a Takotsubo cardiomyopathy pattern. She was optimized from a volume standpoint and was started on guideline directed medical therapy. An ECG one week later showed normalization of her ST/T changes and QTc interval. An echocardiogram 3 weeks later showed a LVEF of 45%.

Discussion: The etiology of takotsubo cardiomyopathy is unclear, but it is thought to be due to a catecholamine surge and vasospasm as a result of a stressful event. In the case of this patient we believe that the procedure was an overwhelming stressful event and caused an increase in endogenous catecholamines and subsequent cardiomyopathy from which she acutely decompensated. Although many causes of both physical and emotional stress have been reported to cause Takotsubo

cardiomyopathy, electrical cardioversion is a previously unrecognized cause. Fortunately, stress-induced cardiomyopathy is usually reversible with an overall good prognosis.

Conclusion: This case represents a rare cause of takotsubo cardiomyopathy.

Categories

3rd year Fellow: Case

Program/Institution Name

Summa Health System/NEOMED

Don't Myx Tamponade, Restriction and Constriction

49

Anezi Uzendu, Aarthi Sabanayagam

The Ohio State University

Type of submitter

Fellow in Training

Abstract

Introduction:

The cardiologist's ability to differentiate the wide array of myocardial and pericardial pathologies in integral to excellent patient care.

Case Presentation:

A 44-year-old female with a history of hypertrophic cardiomyopathy s/p primary prevention ICD presents in shock. Multiple days of progressive dyspnea, and fatigue led to her being found gasping for air by a family member. En-route she was intubated for hypoxia and was started on epinephrine due to hypotension. On arrival, she was hypothermic, heart rate 130 bpm, BP 84/48, Oxygen saturation 82% on 100% FiO2 and 15 of PEEP. The jugular veins were distended to the earlobes at 30 degrees. There are diffuse crackles over both lung fields, muffled heart sounds with a systolic murmur best heard over the left sternal border and 1+ lower extremity edema. Labs were notable for a troponin of 17 ng/mL (ref range <0.1) which eventually peaked at 353 ng/mL six hours later, and a TSH 62 IU/mL. EKG showed incomplete LBBB. Chest X-ray shows a large cardiac silhouette and pulmonary edema. Device interrogation was negative. Her echocardiogram demonstrated severe left ventricular hypertrophy with small LV cavity, EF 60%, and moderate MR without LVOT gradient or SAM. There was a (3 cm) circumferential effusion with a dilated non-collapsing IVC, RVSP 60, lateral and septal mitral annular velocities of 0.02m/s and 0.03m/s respectively, E/E' of 22, no mitral inflow variation, no diastolic chamber collapse, nor expiratory septal bounce. Right heart catheterization demonstrated: RA 23, PA 55/29, PCWP 24, Fick CI 1.8 (equalization of diastolic filling pressures).

Discussion:

Our patient presented with a myxedema crisis in the setting of a restrictive cardiomyopathy. Although tamponade presents with a dilated IVC, large effusion, and diastolic equalization of pressures, her echo

parameters told another story. The lack of enhanced ventricular interdependence evidenced by absent septal bounce, and absent inflow variability make tamponade less likely, despite having a 3 cm effusion. Restriction is characterized by severely reduced tissue doppler velocities, large E/A ratio and E/E' >14 which our patient had. These elevated filling pressures were actually protective against tamponade, and when her hemodynamic instability persisted after draining, we received confirmation the effusion did not indeed drive her clinical picture. Remember equalization of diastolic filling pressures, and a dilated non-collapsing IVC can be seen in tamponade, restriction and constriction.

On that background of restrictive cardiomyopathy myxedema crisis tipped her over the edge. Myxedema patients usually present as she did with a subacute nonspecific syndrome of lethargy, confusion, that can eventually lead to respiratory distress, and cardiogenic shock. The volume retention and cardiovascular suppression from her myxedema created a hemodynamic storm her myocardium could not handle. We treated her myxedema, and optimized her left ventricular filling/ stroke volume by draining her effusion and starting phenylephrine and milrinone

Conclusion

This case highlights the importance of a clinical cardiology fellow's ability to systematically assess patients using both invasive and noninvasive studies to differentiate disease states with similar and sometimes overlapping pathophysiology.

Categories

3rd year Fellow: Case

Program/Institution Name

Ohio State University Hospital