

It Is Not Just Mouth Ulcers; Dilated Cardiomyopathy As a Rare Presenting Symptom For Behcet's Disease

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Type of submitter

Fellow in Training

Abstract



Background:

Behcet's disease is a relatively rare entity, affecting 1 per 15,000 to 500,000 in developed countries. It is most commonly known to cause recurrent oral and genital ulcers alongside a wide variety of systemic

presentation, mostly related to generalized vasculitis. Here we present a case of orthopnea and intractable headache which was later attributed to severe heart failure with reduced ejection fraction as well as superior vena cava thrombosis as an initial presentation of Behcet's disease.

Case Presentation:

A 32-year-old gentleman with a past medical history of asthma presenting with generalized fatigue, orthopnea with intermittent fevers as well as recurrent sore throat over the course of 9 months that triggered multiple emergency room visits and for which he received the diagnosis of URTI. The patient, however, started to develop intractable headaches with facial and chest wall swelling for 1 week prior to his admission to our hospital. Patient's physical exam showed a positive Pemberton sign. He subsequently underwent CT angiogram that confirmed the presence of SVC thrombosis. The patient continued to have orthopnea despite initiation of heparin infusion and improvement of his SVC thrombosis symptoms for which he underwent transthoracic echocardiogram that revealed global hypokinesia with EF of 20-25%.

Upon further detailed history taking, the patient reported having mouth ulcers that used to erupt whenever he had a sore throat, he also reports unusual pimples in his legs that matched the description of pseudofolliculitis. He also reported a family history of Behcet disease in one of his distant family members. A diagnosis of Behcet was made and the patient was started on steroids.

The patient was also started on Sacubitril/Valsartan, Carvedilol, Aspirin, and Atorvastatin. He later underwent Myocardium Perfusion imaging that showed normal EF with no reversible ischemia. He eventually had a Cardiac MRI that showed Improvement of his EF to a low-normal with no evidence of myocardial scarring or fibrosis.

Discussion:

Behcet disease is a rare disease especially outside of the ancient Silkroad area. It is mostly diagnosed clinically, and that why a thorough history and physical is needed to uncover it. A diagnosis is made when a patient is found to have recurrent oral ulcers alongside two of the following; Recurrent genital aphthae, Eye lesions like anterior or posterior uveitis, Skin lesions like pseudofolliculitis, or a positive pathergy test

Cardiac involvement in Behcet's disease is rare but reported in the medical literature. The cardiac presentation can vary from inflammation of one or all of cardiac layers, endomyocardial fibrosis, coronary arteritis, intracardiac thrombus, conduction system disturbances, and valvular disease. Treatment of Behcet varies depending on the type of organs affected, ranging from topical steroids to Colchicine to systemic steroids and immunosuppressants. In our case patient was started early on steroids with a fast and rapid recovery of the myocardium.

Conclusion:

Recognizing the rare, but possibly grave, cardiac manifestation of Behcet's disease including dilated cardiomyopathy is essential as it might aid in making the diagnosis and avoid the burden of increased medical visits and over-testing.

Categories

Advanced Fellow: Case

Program/Institution Name

University of Toledo

Pause and Consider the Late Complications of Heart Transplantation

29

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Type of submitter

Fellow in Training

Abstract

Introduction

Heart Transplantation is the gold standard treatment for patients with stage D heart failure. Survival after transplantation continues to improve; however, chronic complications impact outcomes and survival after transplantation.

Case

We present a 75-year-old female, 23 years status post orthotopic heart transplant presented with nausea, vomiting and an unexplained fall. She was followed routinely in the transplant clinic and maintained on Prednisone 2.5mg daily and Myfortic 720mg BID.

On hospital day two, she experienced sinus arrest lasting 14 seconds. A permanent pacemaker was emergently placed. An echocardiogram revealed normal graft function and workup for rejection was negative. She began complaining of worsening headaches, nausea, left arm numbness and right sided hearing loss. CT scan of her brain revealed a 2.7 cm ring enhancing lesion in the cerebellum; the mass was surgically resected. Pathology was consistent with a lambda restricted B-cell lymphoma favoring post-transplant lymphoproliferative disease (PTLD). Bone marrow biopsy showed no involvement and viral serologies including EBV were negative. A PET scan only localized disease to the cerebellum. Immunosuppression was reduced to Myfortic 360 mg BID and prednisone 5 mg. She was started a chemotherapy regimen of dexamethasone, rituximab, zidovudine and ganciclovir.

Discussion

PTLD is a heterogenous group of disorders presenting in immunosuppressed patients after organ transplantation. The most common sites of PTLD initial involvement were the gut, hilar lymph nodes and lung. The central nervous system (CNS), as in our patient, was a rare site of initial involvement. An analysis identified three cases of primary CNS-PTLD in 1674 heart and lung transplants with an incidence of 0.18% and all were driven by EBV. Our patient had no evidence of EBV in her tumor, bone marrow biopsy or serum.

The late onset nature of our patient's PTLD is unique. PTLD has a bimodal distribution with the majority of cases occurring in the first year after transplantation. The average time to diagnosis after heart transplantation has been reported as 5.5 years. Prior to this report, only two cases of PTLD were described twenty years post transplantation.

Beyond the characteristics of this patient's PTLD, the other novel aspect of this case is the bradycardia and asystole potentially of a neurologic etiology induced by the brain lesion. Although sympathetic reinnervation of the heart is accepted, parasympathetic reinnervation of the heart is controversial. Canine models have shown evidence of parasympathetic reinnervation as early as 6-12 months after transplantation. Human models, however, have not consistently shown parasympathetic control of donor hearts. .

Conclusion

Our patient presenting with an EBV negative primary CNS-PTLD 23 years after heart transplantation is a rare presentation of this immune system driven malignancy. Although conduction system delays in heart transplants are typically due to acute rejection, surgical anastomosis failure or chronic graft vasculopathy, there is data to suggest that reinnervation of the autonomic system the further a patient is from initial transplantation. We recommend pausing and considering neurally mediated etiologies, when evaluating asystole as a late complication of transplantation.

Categories

Advanced Fellow: Case

Program/Institution Name

Ohio State University Hospital

Reverse Takotsubo from Iatrogenic Stress Induced Cardiomyopathy

64

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Type of submitter

Fellow in Training

Abstract

Introduction: Reverse Takotsubo is a version of stress cardiomyopathy in which the base and mid-ventricular segments of the left ventricle are akinetic. This is opposite of the classically described Takotsubo cardiomyopathy in which there is apical ballooning from systolic dysfunction of the apex.

Case: A 63 year old woman with no previous cardiac history presents after suffering a fracture. Her course was complicated by septic shock requiring hemodynamic support. Shortly after starting norepinephrine, she developed tachycardia, severe chest pain and dyspnea. An EKG showed atrial flutter with RVR thus she was started on IV Amiodarone. Troponin-T was elevated and EKG was remarkable for new T-wave inversions. During preparation for cardioversion, an error in the norepinephrine pump was noticed and thus stopped. The patient was successfully cardioverted into sinus rhythm. Her chest pain and dyspnea subsided. A prior echocardiogram showed normal wall motion in all territories and ejection fraction of 55%. The echo done during this episode demonstrated normal function of the apex but hypokinesis of the base. One week later, repeat echocardiogram was remarkable for complete resolution of the basal hypokinesis.

Discussion: Due to an error in pump setup, the patient received 10 mcg/kg/min (700 mcg/min) rather than 10 mcg/min of norepinephrine. This case brings up many important learning points including the management of iatrogenic stress induced cardiomyopathy. The patient received 70 times more norepinephrine than intended resulting in cardiac complications. These symptoms and laboratory findings were all a result of the iatrogenic stress induced cardiomyopathy. Stopping norepinephrine infusion was essential. The error was correctly recognized in a timely manner, preventing the need for unnecessary and potentially harmful procedures.

Conclusion: Reverse Takotsubo is thought to result from the relative density of adrenergic receptors at the apex compared to the base of the heart. The reverse pattern typically causes more myocyte damage and higher troponins. Cardiac adrenergic receptors cause a negative inotropic effect when stimulated by excessive amounts of catecholamines.

Categories

Advanced Fellow: Case

Program/Institution Name

Ohio State University Hospital

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

Electrophysiology Fellowship

Heart University: knowledge-sharing through a web-based learning management system

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Theresa Faulkner, Justin Tretter, Michelle Hudgens

Cincinnati Children's Hospital Medical Center

Type of submitter

CV Team Member (RN, NP, PA, PharmD, etc.)

Abstract

INTRODUCTION: Advances in pediatric and adult cardiology testing and management related to congenital heart disease (CHD) has led to an increase in survival and quality of life for the CHD population in some areas of the world. Both children and adults with complex CHD bring unique challenges and needs to the cardiology community. Unlike acquired heart disease, the relative increased heterogeneity of CHD is not as straightforward in description or treatment. There are over 11,000 Pediatric Cardiac Care Consortium (PCCC) codes, CHD diagnoses that define the unique anatomy and surgical interventions related to CHD. Focused CHD training and education are key elements to improved patient survival and quality of life. The global imbalance in quality training for CHD providers creates a unique opportunity for knowledge-sharing within the CHD community through a web-based learning center.

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OBJECTIVE: To create a global CHD web-based learning center that will be utilized for CHD training and education. The focus of this site will be to make accessible a comprehensive online library of high

quality, evidence-based educational material created and edited by global experts in all subspecialties related to CHD testing, surgery and management.

METHOD: Develop an internationally endorsed Learning Management System (LMS) that will be a portal to access a comprehensive collection of high quality evidence-based educational material on all aspects of CHD. A pediatric and adult CHD editorial board of experts has been established in all the subspecialty areas to collect, create and review materials to be used on the site. This material may include identified high-quality existing material linked to the site, content created by experts specifically for the site, or recordings from global symposia in CHD. The curriculum geared towards fellow training will be guided by current board certification guidelines.

RESULTS: Quality of site and content will be collected monthly through site utilization reports, user content value rating system and discussion boards. Editorial boards will review user recommendations and needs requests quarterly, to update and improve learning material hosted on the website.

CONCLUSION: Creating an international educational website for both pediatric and adult CHD trainees and providers will decrease the imbalance in education and improve the current quality of care which is globally disproportionate.

Categories

CV Team Case

Program/Institution Name

Cincinnati Children's Hospital Medical Center

Device related thrombus following left atrial appendage closure with WATCHMAN®

45

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OhioHealth

Type of submitter

CV Team Member (RN, NP, PA, PharmD, etc.)

Abstract

Introduction

In patients with non-valvular atrial fibrillation (NVAf) who are deemed to be at increased risk for bleeding events, left atrial appendage closure (LAAC) has been proven to be a feasible alternative to long term oral anticoagulation therapy (OAC). Device-related thrombus (DRT) is an under recognized sequelae of implant. We present a case of DRT following LAAC with a WATCHMAN® closure device.

Case Report

An 85 year old female with paroxysmal atrial fibrillation, CHADS₂VASC 6 and extensive cardiovascular history notable for a recovered mixed cardiomyopathy was referred for LAAC secondary to serial episodes of gastrointestinal bleeding related to duodenal angiodysplasias while taking either warfarin or apixaban. LAAC with WATCHMAN was recommended. The patient was instructed to start OAC for 3 weeks prior to implant per our institutional protocol. In this case we initiated apixaban 5 mg twice daily. Pre procedural CT imaging was performed after the patient had been on anticoagulation for 17 days. The left atrial appendage (LAA) was 27 mm in length with an ostial dimension of 23 x 22 mm, with possible distal thrombus. Using conscious sedation, 23 days after initiating anticoagulation, intracardiac echocardiography (ICE), verified the absence of LAA thrombus. The maximum LAA ostial dimension was measured to be 22 mm by ICE evaluation and a 27mm WATCHMAN® device was implanted without a peri-device leak. The device was positioned at the ostium of the appendage meeting PASS criteria. The patient was discharged home with recommendations to continue apixaban 5 mg twice daily and aspirin 81 mg daily for 45 days.

At 45 day follow up, the patient admitted to discontinuing the apixaban one week following the procedure due to concerns for bleeding. Transesophageal echocardiography demonstrated a large mobile DRT adhered to the anchor pin of the WATCHMAN device, measured at 1.4 cm in height. Weight based enoxaparin was initiated and the patient was transitioned to apixaban 5 mg bid after 7 days. She

returned 4 weeks later for repeat imaging and TEE demonstrated complete resolution of the DRT. She was then transitioned to dual antiplatelet therapy for 6 months, and then aspirin (81 mg) alone. In follow-up there have been no bleeding or thromboembolic complications since

Discussion

The incidence of DRT following LAAC has been reported to occur in 3-7% of patients¹⁻⁵ with the incidence of stroke or thromboembolism reported as four times that of those patients without DRT². Several risk factors have been identified and include elevated CHADs2VASC score, large LAA diameter, permanent atrial fibrillation, vascular disease, prior stroke or TIA, and reduced left ventricular systolic function. Despite the increased risk of stroke, the majority of patients with DRT do not incur thromboembolic sequelae². Nonetheless, prompt initiation of anticoagulation should be considered and in the majority of cases extended anticoagulation can resolve the DRT.

Conclusion

DRT may be an under-recognized complication of LAAC. Optimal screening and management recommendations for this population are lacking. This case highlights the potential of early DRT in the setting of patient non-compliance with anticoagulation, although DRT can occur any time following LAAC.



Categories

CV Team Case

Program/Institution Name

Other

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

OhioHealth Riverside Methodist Hospital

ORAL PRESENTATION ABSTRACTS

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

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Type of submitter

Fellow in Training

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

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Cleveland Clinic

Type of submitter

Fellow in Training

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